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Case Reports – Special Supplement

The first special supplement of case reports of the year 2018 is presented here.

The Journal of College of Physicians and Surgeons Pakistan is continuously receiving a large number of case reports of clinical importance for publication in JCPSP. As per JCPSP editorial policy, it cannot accommodate more than four case reports per issue, giving way for other categories of manuscripts.

We hope medical professionals and readers would benefit from this compilation of interesting case reports.

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Abdominal Supernumerary Testis Complicated by Yolk Sac Tumor

Safdar Shah[1], Muhammad Danial Ali Shah[2], Abeer Fatima[3], Fawad Nasrullah[4], Wajid Ali[1] and Irfan Ahmed Gorya[1]

ABSTRACT

We describe a case of a 26-year man with complaints of suprapubic pain and burning micturition for two weeks and intermittent hematuria for two months. On physical examination, there was palpable mobile pelvic mass measuring 10x10 cm. Both testes were palpable in the scrotum. CT scan abdomen revealed well-defined, soft tissue mass, about 11x10 cm between rectum and urinary bladder. Mass showed internal necrotic changes and enhancement along the walls. No calcification was seen. Exploratory laparotomy was done. Tumour mass was nodular tissue weighing 194 gm. Diagnosis was confirmed histologically showing yolk sac tumor. Postoperatively, tumour markers were normal. MRI pelvis revealed no residual tumor.

Key Words: *Polyorchidism. Supernumerary testis. Yolk sac tumor.*

INTRODUCTION

Polyorchidism is a rare congenital defect of the genitourinary tract. It is defined as presence of more than two histologically proven testes. Until now, 200 cases of this defect have been reported in the literature.[1] This usually causes no impairments. It is usually associated with maldescended testis, inguinal hernia, and testicular torsion. There are chances of associated testicular malignancy.[2] Until now, seven cases of malignancy associated with polyorchidism have been reported in various studies. Three patients had teratoma, two seminoma, one each a para-testicular rhabdomyosarcoma and embryonal carcinoma.[3-6] This shows that there is a high risk of testicular cancer in patients with a supernumerary testis.[7]

We present a case of yolk sac tumor in association with polyorchidism where we found supernumerary testicle within abdomen, which has not been previously reported.

CASE REPORT

A 26-year man was admitted in General Surgery Department of Lahore General Hospital, Lahore, Pakistan, with the complaints of suprapubic pain and burning micturation for the last two weeks and painful intermittent haematuria for two months. There was no history of weight loss, fever and generalized weakness. On physical examination, the patient was haemodynamically stable. Abdominal examination revealed palpable, mobile, pelvic mass measuring 10x10 cm. Bowel sounds and digital rectal examination were normal. On inguinoscrotal examination phallus was normal and both testes were palpable within scrotum.

On investigations, complete blood count (CBC), renal profile, liver function tests (LFTs) and bleeding profile were within normal limits. Abdominopelvic ultrasound revealed a large pelvic mass measuring 10x10 cm and bilateral renal cortical cysts. Other visceras were unremarkable. Inguinoscrotal ultrasound revealed both testes of normal size. Scrotal Doppler ultrasound showed both testes of normal size and echogenicity. Right testis measured 3.6x1.7 cm and left 3.34x1.7 cm. Doppler showed normal flow pattern of both scrotal testes.

CT-scan abdomen showed a well-defined, soft tissue mass, about 11x10 cm between rectum and urinary bladder. Mass displaced the urinary bladder and compressed it anteriorly. Mass showed internal necrotic changes and enhancement along the walls. No calcification was seen (Figure 1).

Due to haematuria, Surgical Department shifted this patient to Urology Department, Lahore General Hospital, Lahore, Pakistan.

After preoperative measures, on bimanual examination, pelvic mass was palpable, which was hard and mobile. Cystourethroscopy was done. Urinary bladder was

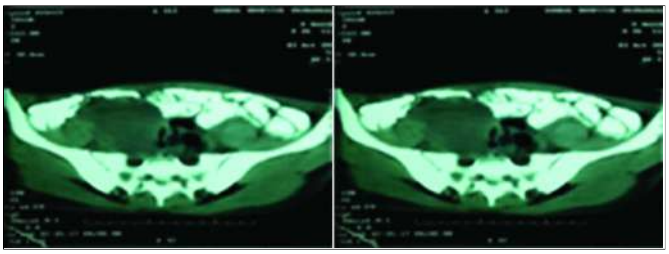

Figure 1: CT-scan showing pelvic mass.

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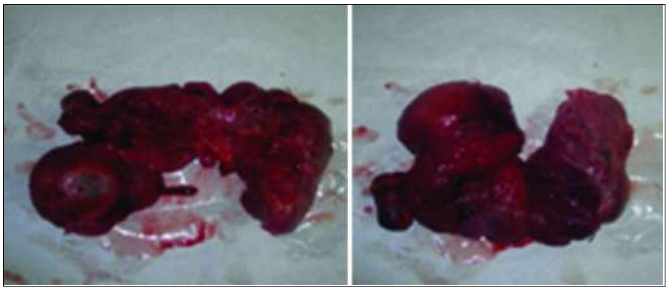

Figure 2: Excised pelvic mass.

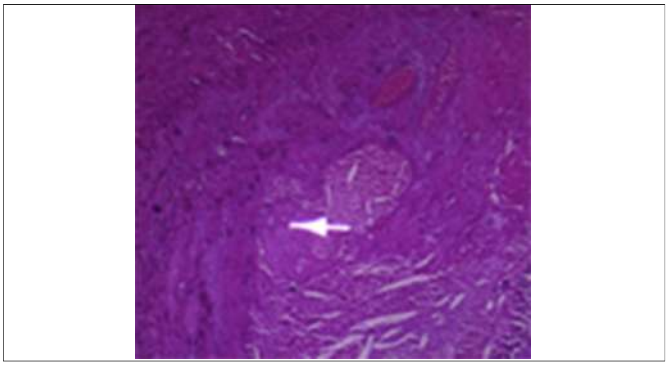

Figure 3: Histological examination shows myxoid stroma and Schiller-Duval bodies.

normal but showed compression from outside. No active bleeding was found. Urethra was unremarkable. After preoperative measures, exploration of mass was done by lower midline incision with the help of general surgeon.

On gross examination, tumor compressed of a large nodular mass and cord like structure. This nodular tissue weighed 194 and measured 7.5x6.5x4.0 cm (Figure 2). It seemed to be testis and had intact capsule. Attached tubular structure seemed to be spermatic cord and measured 5.5 cm in length and 0.5 cm in diameter. Diagnosis was confirmed histologically of germ cell tumor composed of microcystic and solid patterns. Multiple Schiller-Duval bodies were identified. There was loose and myxoid stroma (Figure 3). Hyaline lobules were identified; yolk sac tumor. Spermatic cord resection margin was free of tumor. All morphological features were consistent and the diagnosis of lymphovascular invasion was seen.

Postoperatively, tumor markers were carried out. Alpha fetoprotein (AFP) was 0.646 IU/mL, human chorionic gonadotropin (HCG) <2 ng/ml, lactate dehydrogenase (LDH) 629 U/L and testosterone 166 ng/ml.

MRI abdominopelvis was reported unremarkable, no residual recurrent tumor in pelvis was seen. The supernumerary testicle was intraoperatively found in retro-peritoneal cavity and completely developed with separate cord structure and epididymis. This case of polyorchidism, with abdominal supernumerary testicle having yolk sac tumor, has not been previously reported.

DISCUSSION

The exact mechanism for the presence of polyorchidism is still unknown. There are various views regarding its etiology, including foldings of peritoneum, gonads segmentation, and division of the genital ridge.[4]

In fetal life, at about 6 weeks of gestation, the primordial testis develops from the primitive genital ridge medial to the mesonephric ducts.[5] On the basis of embryologic development, Leung classified polyorchidism into four types.[6]

In our study, the patient was a 26-year man and presented with suprapubic pain. Bergholz and colleagues reported polyorchidism in 187 cases; in 140 cases, histology was confirmed.[1] Average age was 17 years and most patients were in the age of 11-25 years, and majority of patients had left sided supernumerary testis. Commonest site was within left scrotum.[3]

In our case, supernumerary testis had yolk sac tumor without any associated abnormality and was found incidentally within the abdomen, which is different from other cases.

Inguinal hernia, cryptorchidism and testicular torsion have incidence of 24%, 22% and 15%, respectively in association with polyorchidism.[1] Amodio and colleagues reported a case of triorchidism with co-existant micro-lithiasis.[8] Hydrocele, infertility, varicocele, retractile testis, hypospadias and epididymitis also have association with polyorchidism.[9] It has been reported in literature that there is increased risk of testicular malignancy with polyorchidism.[1,2] Malignant transformation may occur regardless of the location of the supernumerary testis. Commonest malignancies are germ cell tumors, such as embryonal carcinomas and seminomas.[1,2,10] Rhabdo-myosarcomas of extra testicular tissue and rete testis adenoma is also reported in supernumerary testis.[1]

In uncomplicated cases of polyorchidism, MRI does not provide any additional data as given by sonography, but only plays a confirmatory role. However, MRI plays an important role in complicated cases, such as those associated with cryptorchidism and tumours.

Jatkar *et al*. reported a similar case with intra-abdominal swelling as supernumerary testis which was located intra-peritoneally and was completely developed and had epididymis and separate cord.[12]

In the setting of an uncomplicated polyorchidism, the current treatment is conservative, including a close sonographic observation, with a biopsy of the supernumerary testicle for diagnosis or follow-up being unnecessary.[4,7] On the other hand, in the presence of coexisting conditions, such as cryptorchidism, torsion or malignancy, surgical treatment is indicated.

Polyorchidism is uncommon anomaly. In most cases, sonography is diagnostic choice but MRI may provide

further details. The treatment of choice is a conservative management but surgical treatment is indicated in cryptorchidism, torsion, and malignancy in polyorchidim.

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Life-Threatening Hypokalemic Quadriplegia in a Postoperative Patient

Manoj Kamal[1], Geeta Singariya[2], Om Prakash Suthar[2] and Ashwini S.[2]

ABSTRACT

Acute hypokalemic paralysis is a reversible but potentially lethal clinical condition. We report a case, who developed rapid-onset quadriparesis in immediate postoperative period after undergoing right percutaneous nephrolithotomy for bilateral renal stones. On evaluation, she was found to have hypernatremic, hyperchloremic, hypokalemic acidosis. This severe hypokalemia-induced quadriparesis was precipitated by repeated furosemide injections, use of potassium-free fluid as maintenance, intracellular shift due to free water administration in this patient, who had pre-existing distal renal tubular acidosis.

Key Words: *Hypokalemia. Paralysis. Renal tubular acidosis.*

INTRODUCTION

Hypokalaemic paralysis can be caused or precipitated by hereditary disorders like hypokalemic periodic paralysis; due to nutritional deficiency, drugs or iatrogenic factors or due to gastrointestinal losses or renal wasting in renal tubular acidosis. Secondary causes of hypokalemic paralysis include thyrotoxicosis, dengue viral infection, Gitelman syndrome, and Conn's syndrome.[1,2]

Distal renal tubular acidosis (dRTA) is caused by failure of acid secretion in the urine by the alpha intercalated cells of the cortical collecting duct of the distal nephron.[3] This leads to an inability to acidify urine to a pH of less than 5.5. The clinical manifestations of dRTA are normal anion gap metabolic acidosis, hypokalemia, urinary tract stone formation, nephrocalcinosis, and bone demineralisation causing rickets in children and osteomalacia in adults. The symptoms and sequelae of dRTA are variable, ranging from being completely asymptomatic, being compensated by high rate of NH_4^+ excretion, loin pain and haematuria due to kidney stones, failure to thrive and severe rickets as well as possible renal failure and even death. The most common cause of death in RTA type I is hypokalemia-induced cardiac dysrhythmia. dRTA leads to sodium loss and volume contraction, which further cause a compensatory increase in the blood level of aldosterone. Aldosterone causes increased resorption of sodium and loss of potassium in the collecting duct of the kidney. Increased aldosterone levels cause the hypokalemia, which is a common symptom of dRTA.[4]

There are many reports of hypokalemia manifesting as quadriparesis in the literature in the presence of autoimmune diseases or other underlying systemic illnesses.[5-7] We report a case of quadriparesis in a known case of dRTA, which was precipitated by concurrent iatrogenic factors.

CASE REPORT

This case was a 23-year female, who presented with complaints of recurrent episodes of abdominal pain radiating to back and haematuria. Her ultrasound (USG) abdomen revealed right sided lower and upper ureteric calculi of sizes 21.7 and 15.8 mm, respectively with associated hydronephrosis. Left kidney had nephrocalcinosis with normal ureter and no hydronephrosis. She was a diagnosed case of dRTA since last 2 years, and was on oral soda-bicarbonate replacement therapy. She had previous history of left upper ureteric calculus removal; frequent hospital admissions for percutaneous nephrostomy tube insertion; and urine and blood stream infection requiring antibiotic therapy. The patient had history of generalized weakness in the past, but there was no history of breathing difficulty or limb weakness. Patient had no history of diabetes, thyroid disease, or any autoimmune disease like systemic lupus erythematosus (SLE) or Sjögren's syndrome (SS).

She underwent ureteroscopic lithotripsy with double J stenting on right side and percutaneous nephrolithotomy on left side under general anaesthesia. Her intra-operative and immediate postoperative course was uneventful. During intraoperative period, she was administered furosemide injection on surgeon's demand for constant flush and prevention of hematoma formation in the surgical area, which was repeated in the post-operative period, too.

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Ten hours after the surgery, patient developed rapid, shallow breathing pattern and extreme muscle weakness mimicking flaccid paralysis. Patient was conscious, opening her eyes spontaneously, but there was no power (0/5) in upper and lower limbs. Deep tendon reflex was sluggish with mute plantar response. The sensory functions were normal. Arterial blood gas (ABG) analysis done at that time showed pH of 6.851, PO_2 150.3 mmHg, PCO_2 119.9 mmHg, base deficit of 15, bicarbonate levels of 10 mEq, sodium 159 mEq/L, chloride 110 mEq/L, and potassium 1.7 mEq/L. The blood glucose was within normal limit. The patient was intubated in view of the grossly inadequate respiratory efforts and ABG findings and subsequently shifted to surgical intensive care unit (SICU), where she was electively ventilated on assist-volume controlled mode.

In SICU, electrocardiogram (ECG), USG abdomen, X-ray cervical spine and chest x-ray were done, which revealed no abnormality except for the hypokalemic changes in ECG. MRI study of brain, spine and abdomen was also normal.

Initially, in view of hypernatremia, she was given intravenous 5% dextrose along with potassium chloride and free water by nasogastric tube. This, further led to worsening of hypokalemia to 1.4 mEq/L.

For rapid potassium correction and also to know the fluid status of the patient and cause of hypernatremia, central line was then inserted in the right internal jugular vein under USG guidance. Central venous pressure value at that time was 6 mmHg. With correction of relative volume contraction state with Ringer lactate and correction of hypokalemia to potassium level >3.5 meq/L with potassium chloride infusion through central line, patient showed dramatic recovery. Her respiratory muscles and limbs muscle power recovered to near normal in next 48 hours and we were able to wean her off ventilator, extubate her and also mobilize out of bed.

DISCUSSION

The cause of severe hypokalemia leading to transient quadriplegia in this patient was multifactorial: 1. presence of dRTA in the patient, which is known to be associated with chronic hypokalemia. Further, this patient was not receiving any potassium supplementation in the preoperative period; 2. Repeated use of furosemide in the perioperative period, which increases urinary excretion of potassium, and 3. Postoperative use of potassium-poor intravenous fluids because of presence of hypernatremia.

dRTA, as was present in our patient, can be: 1. Primary, which is inherited as autosomal dominant or autosomal recessive form; can be persistent or transient; can be associated with sensorineural deafness or bicarbonate wasting; 2. Secondary to autoimmune diseases such as SLE, SS, chronic active hepatitis, primary biliary cirrhosis, thyroiditis, fibrosing alveolitis, rheumatoid arthritis; 3. Calcium disorders (primary hyperparathyroidism, hypercalcemic hyperthyroidism, vitamin D intoxication, idiopathic hypercalciuria with nephrocalcinosis, 4. Drug induced, such as amphotericin B, lithium, analgesic abuse, trimethoprim etc. or 5. As a consequence of hypergammaglobulinemia, chronic renal allograft rejection etc.[4]

The cornerstone of medical treatment for dRTA consists of replenishing potassium with intravenous and oral potassium chloride or potassium citrate, and compensating for the lost bicarbonate with alkali in the form of oral sodium bicarbonate (1-2 mEq/kg/day), which may be required for lifetime in primary cases. Alkali therapy restores growth in children and prevents the progression of nephrocalcinosis at all ages. Late diagnosis or delaying the therapy to late childhood or adulthood may lead to end-stage renal insufficiency.[4]

Perioperative management of dRTA patients should focus on evaluation of the causes, preoperative optimization of acid base and electrolyte status, perioperative blood gas and electrolytes monitoring, avoidance of drugs/factors precipitating hypokalemia/acidosis, intravenous administration of balanced potassium-containing fluids, monitoring of ECG changes for hypokalemia; neuromuscular monitoring guided use of muscle relaxants for general anaesthesia and local anaesthetic agents for regional blockade and maintaining a high degree of vigilance.

Early diagnosis is imperative for prompt and optimal management of this life-threatening condition.

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Complete Heart Block due to Occlusion of Jailed Septal Perforator after Percutaneous Coronay Intervention of Proximal Left Anterior Descending Artery

Ghazanfar Ali Shah[1], Jawaid Akbar Sial[2] and Tahir Saghir[2]

ABSTRACT

A 70-year diabetic and hypertensive lady admitted with acute coronary syndrome (ACS) underwent coronary angiography, which showed severe diffuse disease of proximal left anterior descending (LAD). She underwent percutaneous coronay intervention (PCI) of proximal LAD artery that resulted in occlusion of jailed first septal perforator. She remained stable and asymptomatic and was shifted to Coronary Care Unit (CCU) after successful procedure. Approximately five hours later, patient developed complete heart block (CHB) and became hemodynamically unstable. Temporary pacemaker (TPM) was implanted and relook angiogram was performed, which showed patent stent. Patient remained dependent on TPM. After one week, permanent pacemaker was implanted and patient discharged in stable condition.

Key Words: *Percutaneous coronery intervention. Septal perforator. Complete heart block.*

INTRODUCTION

Percutaneous coronay intervention (PCI) is a novel approach for the treatment of critical coronary artery stenosis. Side branch occlusion is not an uncommon complication of PCI occurring in up to 18% of patients.[1] Atheromatous plaque shift or the "snow plough" effect into the side branch is reported to be the responsible mechanism.[2] Documentation of transient complete heart block (CHB) due to jailing of the first septal perforator during PCI of LAD is not infrequent. However, occurrence of irreversible or delayed CHB following occlusion of septal perforator after PCI of left anterior descending (LAD) is an extremely rare complication. Our literature review found only 4 published reports of delayed CHB following jailing of septal perforator with only two cases of permanent delayed CHB requiring implantation of permanent pacemaker (PPM).[3] To our knowledge, this is third case of delayed CHB, where PPM implantation was required secondary to occlusion of jailed septal perforator following PCI to proximal LAD.

This case demonstrates the importance of electro-cardiograph (ECG) monitoring, if procedure gets complicated with occlusion of jailed first septal perforator, which may require consideration of necessary steps.

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CASE REPORT

A 70-year lady, known case of diabetes mellitus (DM) and hypertension (HTN), admitted in hospital with non-ST elevation myocardial infarction (NSTEMI). She underwent coronary angiography, which showed severe diffuse disease in proximal LAD (Figure 1). Other vessels were not obstructed. Mild left ventricular systolic dysfunction on left ventricular angiogram was noted.

PCI was performed through right radial access using extra backup guiding catheter. Balanced medium weight (BMW) wire was used to cross the lesion and predilated it with 2.0x20 mm splinter semi compliant balloon. A 3.5x30 zotarolimus eluting stent (resolute integrity) was deployed successfully with TIMI 3 flow. Jailed side branch (septal perforator) was occluded. Stent was postdilated with 3.75x12 mm non-compliant balloon. Procedure was completed without complication except occlusion of septal perforator (Figure 2). Patient was shifted to coronary care unit (CCU) in stable condition.

After five hours, patient became unstable as her rate dropped to 30 beats per minute and systolic blood pressure to 70 mm Hg. Electrocardiogram showed complete heart block (Figure 3).

Patient was shifted to Cath Lab, where temporary pacemaker (TPM) was implanted and relook angiogram was performed, which showed patent stent. Patient's condition stabilized after putting pacemaker. She again shifted to CCU. She was observed for seven days, but she remained dependent on TPM. Finally, dual chamber permanent pacemaker (PPM) was implanted and patient was discharged next day in stable condition.

DISCUSSION

The first septal perforator supplies the superior and anterior portion of the interventricular septum; its

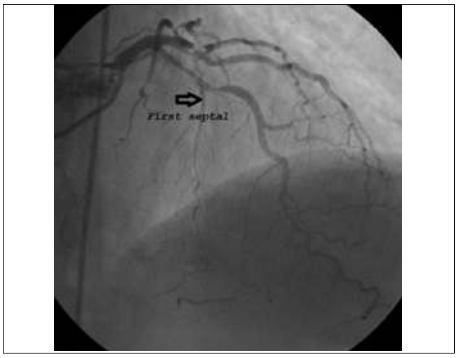

Figure 1: AP cranial projection showing severe disease in proximal LAD, septal perforator arising from diseased segment of LAD.

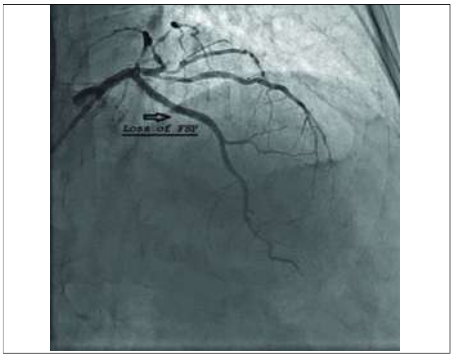

Figure 2: AP cranial projection showing successful deployment of stent at proximal LAD, occlusion of jailed septal perforator is also evident.

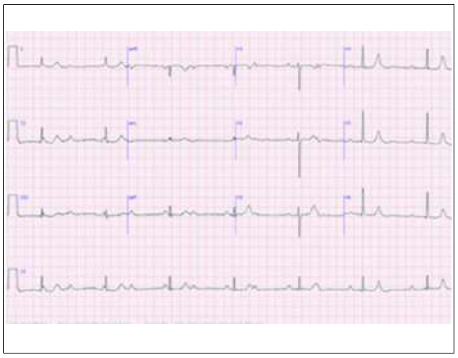

Figure 3: Electrocardiogram showing complete heart block.

occlusion following PCI of LAD can cause infarction, conduction disturbances, giving rise to symptoms of angina, arrhythmias, and heart failure. ECG changes vary from sinus bradycardia, first-degree AV block, new-onset right bundle branch block (RBBB), polymorphic ventricular tachycardia, to complicating acute myocardial infarction (MI).[4] In our patient, development of CHB after occlusion of the first septal perforator was consistent with damage to the conduction system caused by hypoperfusion and necrosis, which ultimately required PPM implantation.

In the case reported by Pillai *et al*, spontaneous reversion to sinus rhythm with normal PR interval and QRS complexes was observed 12 hours after the onset of delayed CHB.[1] Kireyev *et al*. performed successful intervention of first septal perforator (FSP) in their patient with delayed CHB with a gradual return of AV conduction via the left bundle branch within 48 hours. Although the septal artery angioplasty is relatively safe and not associated with aortic dissection, MI, or death, we decided not to reattempt wiring of FSP as CHB associated with anterior MI is usually considered to be irreversible. Sadiq *et al*. reported an observation of delayed CHB due to jailing of FSP following PCI to LAD, on basis of which they recommended extension of hospital stay for continuous ECG monitoring or prophylactic temporary transvenous pacing.[5]

In our patient, development of CHB was after five hours of procedure and was not preceded by fascicular blocks. This finding is not consistent with all previously reported cases.

Our case demonstrates a very rare complication of PCI where loss of septal perforator resulted in CHB. Putting an extra wire to side branch (septal perforator) and maintaining its patency, it can prevent this very rare and grave complication in selected cases.

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Treatment of a Rare Vascular Complication of Coronary Stenting in an Octagenarian

Hunaina Shahab, Bilal Hussain, Nasir Rahman and Fateh Ali Tipoo Sultan

ABSTRACT

The ability of drug-eluting stent (DES) to inhibit intimal proliferation has resulted in a massive increase in their usage over the years. However, it is known that the application of DES can alter the normal cascade of vascular healing, resulting in delayed endothelialisation with risk of vascular complications. Coronary artery aneurysms (CAN) are defined as more than 50% dilatation of the coronary artery compared to the reference vessel diameter with the reported incidence after percutaneous intervention (PCI) being only around 0.35 to 6.0%. Previously, CAN had been reported with the use of bare metal stent secondary to stretch, stent fracture and dissection. However, recently, increasing number of cases have been reported describing CAN after DES implantation. To the best of the authors' knowledge, they present the first case from Pakistan of a left anterior descending coronary artery aneurysm after DES implantation treated successfully with stenting under intravascular ultrasound guidance.

Key Words: *Coronary vessels. Aneurysm. Stents.*

INTRODUCTION

Coronary artery aneurysms (CAN) are defined as more than 50% dilatation of coronary arteries compared to reference vessels diameter,[1] with incidence of 0.35 to 6.0%.[2,3] Previously, CAN had been reported with bare metal stents (BMS) secondary to stretch, stent fracture and dissection.[2] Recently, increasing number of cases report CAN after drug-eluting stent (DES) implantation.[4,5]

To the best of the authors' knowledge, the first case presented from Pakistan of a left anterior descending (LAD) CAN after DES implantation treated successfully with stenting.

CASE REPORT

An 85-year lady with a history of percutaneous intervention (PCI) to left anterior descending LAD with CypherTM stent (DES) in 2007, presented to the Emergency Department of The Aga Khan University Hospital with increasing restlessness and cough. She had a pulse of 110 beats/minute, respiratory rate of 40 breaths/minute and blood pressure of 90/60 mmHg. She had bibasilar crepitations on auscultation and troponin-I of 10.5 ng/ml. Her electrocardiogram (ECG) showed old left bundle branch block. Transthoracic echocardiogram showed an estimated ejection fraction of 25-30% with akinetic apex, septum, mid anterior segments along with mild-moderate aortic stenosis and moderate mitral regurgitation.

She underwent a left heart catheterisation via right femoral artery 6F sheath (Cordis®). Left coronary system was visualised using JL4 6F` catheter showing short left main vessel bifurcating into LAD and left circumflex vessels (LCx). LAD had tight in-stent re-stenosis (ISR) in the proximal part of previous stent (Cypher 2.5x23 mm), followed by CAN (Figures 1A and 1B). LCx had mild plaquing. Right coronary artery was engaged using JR4 6F` diagnostic catheter, showing mild plaquing.

The left main artery was engaged with VL3 (Cordis®) guiding catheter and LAD wired with BMW (Abbott®)

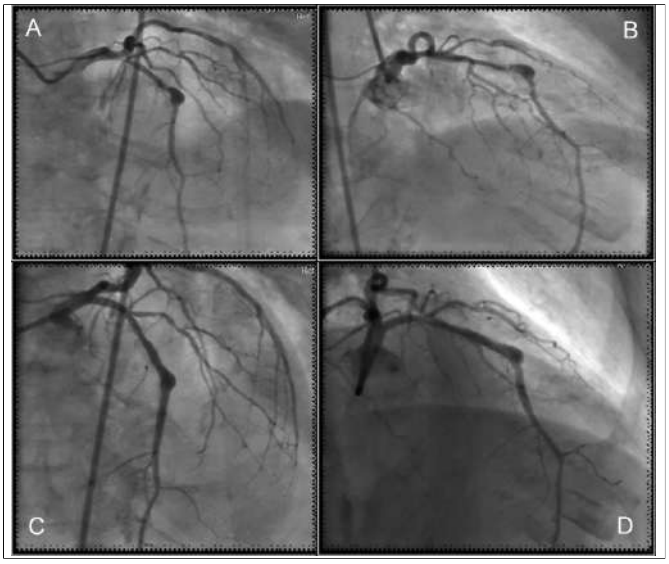

Figure 1: (A, B): Coronary angiogram showing tight in stent re-stenosis (ISR) in Left Anterior Descending (LAD) stent, followed by coronary artery aneurysm in the prior stent. (C, D) Angiogram after stenting of prior mid Left Anterior Descending (LAD) stent and coronary aneurysm. Post PCI TIMI III flow was achieved in LAD.

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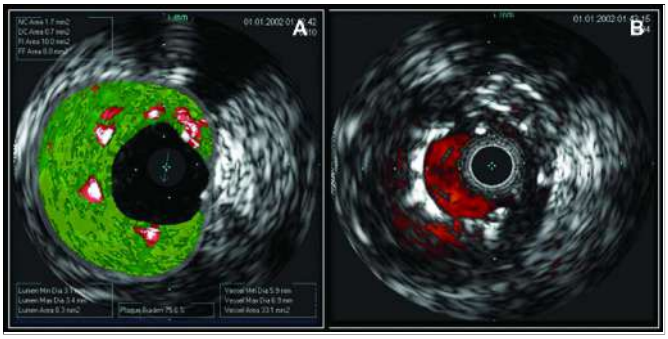

Figure 2: (A) Intravascular ultrasound (IVUS) showing ISR along with aneurysm in prior mid Left Anterior Descending (LAD) stent. (B) Well deployed and apposed stent with minimal gap in between two stent struts visualised.

0.014 inch wire. Mid-LAD ISR was predilated with Saphire II (OrbusneichTM) 2.0x10 mm at 6 atm (Figures 1C and 1D). Intravascular ultrasound (IVUS) revealed ISR in proximal part of mid-LAD stent with aneurysm (Figure 2A). Xience xpedition (Abbott®) 2.5x33 mm (DES) was deployed into mid-LAD stent including proximal and distal segments to it at 10 atm. Stent was post-dilated sequentially with NC TREK (Abbott®) 2.5x15 mm at 14-20 atm and NC TREK (Abbott®) 3.0x8 mm at 10-16 atm. At the end of procedure, IVUS revealed well-deployed and apposed stent with minimal gap in between two stent struts (Figure 2B). TIMI-III flow was achieved in LAD.

The patient was kept on dual antiplatelets (DAPT) and glycoprotein IIb/IIIa inhibitor (GpIIb/IIIa) infusion. She improved over the course of admission.

DISCUSSION

DES are impregnated with drugs which interfere with inflammatory pathways and neo-intimal proliferation, however, these local effects induce delayed re-endothelialisation, hypersensitivity reactions, and inflammatory changes of vessel wall, leading to CAN formation.[6,7] Coronary angiography is gold standard for diagnosing CAN, yet it only gives information regarding lumen of the arteries.[2] IVUS allows detailed characterisation of the aneurysms, as it visualises layers of the coronary arteries and differentiates between true and pseudo-aneurysms.[8] Aoki *et al*. have classified CAN into 3 types.[2] Type-1 occurs within 1 month secondary to mechanical injury to the arterial wall. Type-2 occurs after 6 months of stent placement as an arterial wall response to DES. Type-3 are infectious mycotic aneurysms.

There are no documented guidelines for management of stent-related CAN. The only available data are isolated management protocols from individual case reports. These include observation, antiplatelet treatment, coiling or surgical excision.[9] Aoki *et al* proposed a management plan.[2] They suggest percutaneous treatment with stent graft, bavemetel stent (BMS) or surgery for large (more than 2 times the size of reference vessel) or symptomatic pseudo-aneurysms and large and/or symptomatic true aneurysms. For small or asymptomatic true and pseudoaneurysms, followup angiography at 3 to 6 months is suggested. For small true aneurysms and those large asymptomatic with no change in follow-up angiography, careful observation and DAPT is suggested. We used IVUS, as it aptly detects mal-apposition of DES and vessel wall and also defines the size of aneurysms especially in cases of DES thrombosis.[10] There is a lack of guidelines on the choice of antiplatelet therapy and duration of antiplatelets in cases of CAN. We decided to keep our patient post-PCI on DAPT and GpIIb/IIIa infusion for optimal antiplatelet effect.

CANs are rare vascular complications of DES. Their optimum treatment is still debatable, yet the application of DES under IVUS guidance can result in successful treatment.

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Use of TAP Technique in Percutaneous Coronary Intervention, for Stenting Critical Osteal Disease in an Old Patient with Compromised Left Ventricular Function

Uzma Zahid and Asad Islam

ABSTRACT

With recent advancements in techniques and technologies in the field of interventional cardiology, Percutaneous Coronary Intervention (PCI) is preferred over Coronary Artery Bypass Grafting (CABG), provided patient selection is kept in mind, and all the technical facilities are available to ensure successful end results without any immediate or late cardiac complications. However, dealing with Left Main Coronary Artery (LMCA) osteal disease can be challenging. We report a case of T and Protrusion (TAP) technique performed at Hearts International Hospital, Rawalpindi, Pakistan. The patient was old and had compromised Left Ventricular Ejection Fraction (LVEF).The results were excellent with no complications.

Key Words: *Interventional cardiology. Percutaneous coronary intervention. Left main coronary artery osteal disease. T and protrusion technique.*

INTRODUCTION

Almost 15-20% of all Percutaneous Coronary Interventions (PCIs) are done for bifurcating lesions.[1] It has been challenging to deal with such lesions as far as procedural success rate and long term cardiac events are concerned.[2] T and protrusion (TAP) technique, if done by expert clinical hands, shows excellent results in dealing with osteal disease. Reported here is a case of TAP technique performed successfully at Hearts International Hospital, Rawalpindi. The patient was old and his left ventricular (LV) function was compromised.

CASE REPORT

A 70-year male patient underwent Percutaneous Transluminal Coronary Angioplasty (PTCA) and stenting to Left Anterior Descending (LAD) artery in 2004. Since then, he has been on dual antiplatelet therapy and lipid lowering drugs. One year later, he presented in outpatient department (OPD) with history of intermittent chest pain. Coronary angiography was recommended by cardiologist, but refused by the patient.

In 2015, he presented in our hospital with complaint of on and off chest pain. This time, he agreed to coronary angiography, but refused to stenting. Coronary angiography showed double vessel coronary artery disease with distal tapering in Left Main Coronary Artery (LMCA), mild late loss of stents of LAD, severe 99% osteal disease with mild disease in Circumflex Artery (Cx) and mild disease in proximal segment of Right Coronary Artery (RCA). Plain Old Balloon Angioplasty (POBA) to Cx was performed using 2.75 × 8.0 non-compliant balloon at 10 ATMS (atmospheric pressure). The procedure had excellent end results with reduction of stenosis up to 40%, causing good distal coronary bed flow. He was discharged from hospital with added nitrates and non-dihydropyridine calcium channel blocker to previous medications.

In July 2017, he presented to local hospital with severe shortness of breath and orthopnea; he was managed there as a case of Left Ventricular failure (LVF) and was shifted to Hearts International Hospital, Rawalpindi, where echocardiogram showed Left Ventricular Ejection Fraction (LVEF) of 40%. LV was hypokinetic, and there was grade II Mitral Regurgitation (MR). There was added moderate osteal disease noted in LAD, in addition to previous coronary angiography findings. Stenting to LAD, Left Main Stem (LMS) and Cx was done after patient's informed consent. Six French (Fr) guiding catheter was used. Balloon dilatation of Cx and LAD was done using 2.0×10 balloon catheter, followed by LMS/LAD stenting with Drug Eluting Stent (DES) 3.5×24 at 14 ATM. Cx was rewired and pre-dilated with 2.5×12 Balloon at 16 ATM. DES did not cross, so Cx artery was stented with 2.5×10 Bare Metal Stent (BMS). TAP technique to LAD/Cx with 3.5×15 and 2.5×10 Balloon was performed. Proximal Optimisation Technique (POT) to LMS was done with 4.0×10 non-compliant Balloon at 14 ATM. The procedure had excellent end results (Figures 1 and 2). He was discharged from hospital on diuretics added to previous medications. His symptoms have much improved on follow-up.

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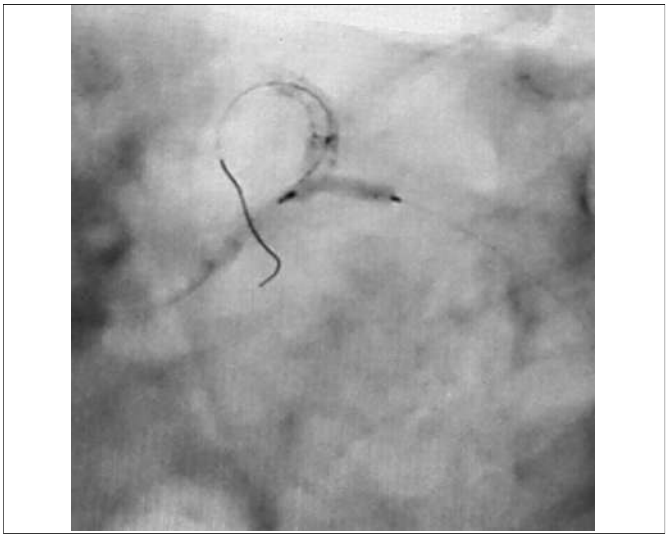

Figure 1: Stents in LMS/LAD and Cx, covering the osteal lesion.

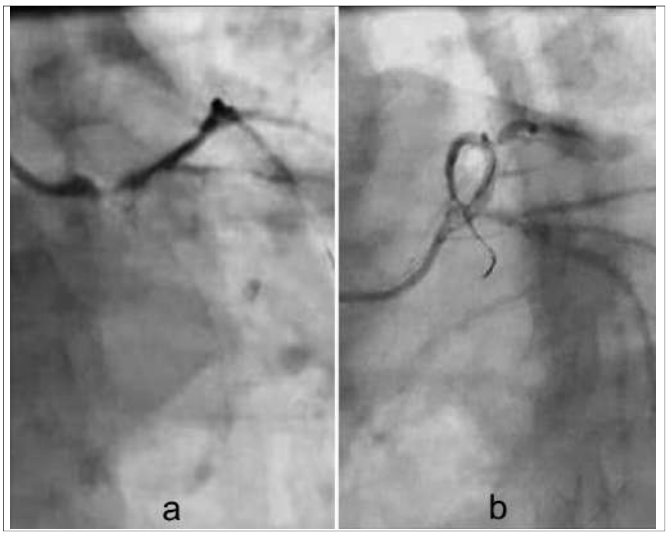

Figure 2: Pre-procedure osteal lesion (a), compared with post-procedure good coronary flow (b).

DISCUSSION

Ever since the clinical practice of coronary angioplasty, stenting of bifurcating lesions account for great therapeutic challenge to deal with.[3] For T-shaped bifurcations with angle >70, T technique is used. In cases where angle is <70, modified TAP technique can be performed. It gives good ostium coverage and is superior to T technique.[4]

In T-stent and small protrusion (TAP) technique, Main Branch (MB) is stented first, followed by stenting of Side Branch (SB). Using the struts of MB stent, SB is dilated and stented, with 1-2 mm proximal part of SB stent positioned within the MB stent. During SB stenting, MB Balloon is left uninflated.

POT refers to expansion of stent from the proximal stent edge to just proximal to carina, using a short oversized balloon. POT might be used in case of large difference in diameters between proximal and distal MB, or difficult SB rewiring.[2] Using two non-compliant balloons, Final Kissing Balloon Inflation (FKI) may be performed,[5] that allows full expansion of stent in proximal MB, providing better scaffolding of SB ostium.[6]

TAP technique allows full coverage of bifurcating lesions, facilitating FKI.[7] It can be used, provided technical facilities and expertise are available, keeping in mind patient selection. It is of great clinical importance in the practice of interventional cardiology. Successful results of TAP technique in dealing with critical osteal disease in this old patient, with compromised LVEF are indicative of the importance of this technique in clinical practice.

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Swirl Sign – A CT Angiogram Alarm Sign for Congenital Trans-Mesenteric Hernias

Ahmad Uzair Qureshi[1], Maliha Fansur[2] and Sidra Shoaib Qureshi[3]

ABSTRACT

Congenital trans-mesenteric hernias (CTMHs) are caused by rent in the mesentry of bowel. The lesions commonly present in pediatric age group. Adult CTMHs usually present with complication of the problem. The diagnosis is often late and is associated with morbidity as well as mortality due to bowel ischemia. There is no specific sign associated with this condition. A case is reported of a patient diagnosed with CTMH on high suspicion by the radiologist based on her finding of abnormal "swirling" of superior mesenteric artery on CT angiogram. Swirl sign on CT angiogram warrants an early surgical consultation to prevent any morbidity.

Key Words: *Internal hernia. Trans-mesenteric defects. Swirl sign. Whirlpool sign. CT angiogram. Gordian knot.*

INTRODUCTION

Congenital trans-mesenteric hernias (CTMHs) are rare forms of internal hernia. The presentation is usually in pediatric age group.[1,2] However, these may report in adults as incidental finding or a complication such as obstruction or strangulation of bowel. The condition is associated with morbidity and mortality.[3,4] These hernias develop due to bowel atresia pre-natally. The size of defect varies from a few centimeters to large defects.[5] Diagnosis is based on clinical suspicion.[5,6]

However, we report a case in which "swirl sign" or a "whirlpool sign" on computed tomography (CT) angiogram, which led to the clue for the diagnosis.

CASE REPORT

We report a case of 32-year female who was referred to us with off and on intractable epigastric colicky pain aggravating with meals and upper abdominal discomfort, relieved by episodes of vomiting 1 - 2 hours after the meal. Symptoms of upper abdominal discomfort would start 10 - 20 minutes postparandially and always followed the same sequence. She used to pass stools once every 3 - 4 days of normal consistency and colour. Lately, patient was afraid of taking meals and lost 6 kilogram in the past year.

On examination, she was thin, lean, patient with flat abdomen and no abnormal finding. Bowel sounds were hyperactive after taking meals. No other abnormal findings were noted and her per rectal examination revealed normal stool.

Her basic biochemical investigations were within normal range including complete blood examination and chemistry screens. She underwent four ultrasound abdomen performed during the past six months with no positive finding except one report suggesting distended small bowel. Her upper gastro-intestinal (GI) endoscopy was done, which was normal and scope examined her all the ways to the duodenum.

Working diagnosis of superior mesenteric artery syndrome was made, and CT angiography was performed. CT angiogram showed a normal angle between the superior mesentenic artery (SMA) and aorta. The orientation of SMA and vein were normal. However, distal SMA was seen to "Swirl" distally into a possible small bowel volvulus (Figure 1).

She was explored with preoperative diagnosis of small bowel volvulus and subacute intestinal obstruction

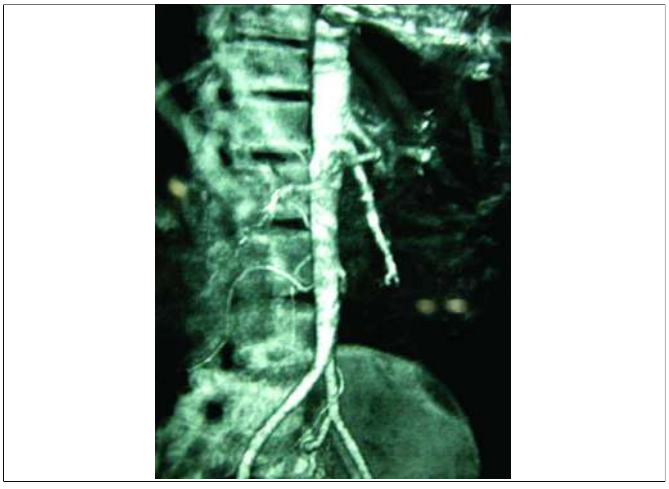

Figure 1: Swirl sign on CT angiogram.

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through a midline laparotomy. On exploration, distended small bowel was seen herniating through a 20 x 15 cm congenital defect in the mesentery of small bowel (Figure 2), reported in literature as the "Gordian knot" of obstructed intestine (Figure 3). Almost whole length of ileum and proximal jejunum was herniating through the defect. The herniation was reduced carefully to untwist the bowel loops and defect closed using interrupted absorbable sutures (Figure 4). Postoperative recovery was uneventful and patient was discharged on 3rd post-operative day.

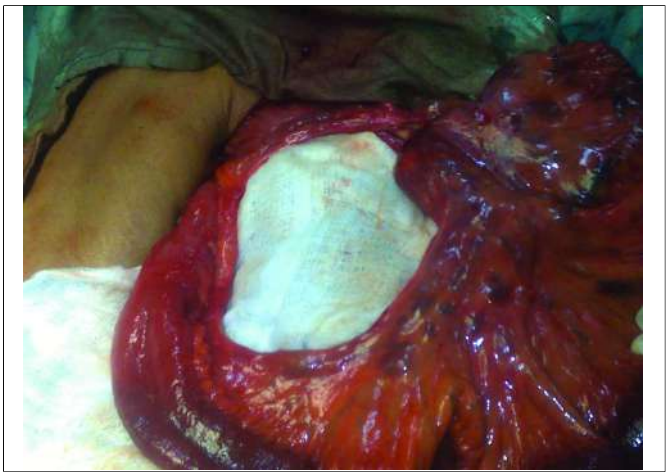

Figure 2: Large mesenteric defect.

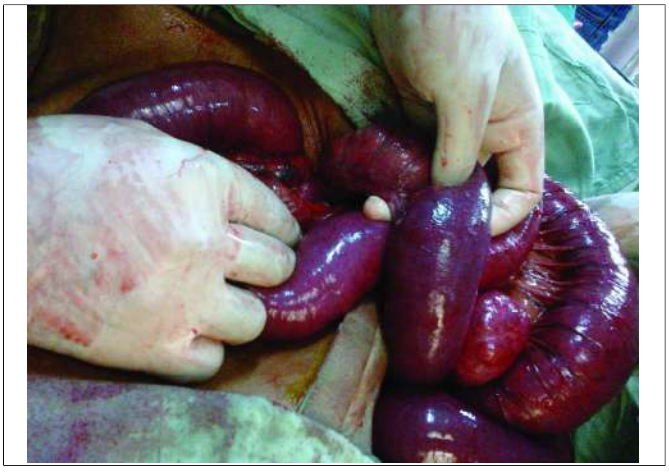

Figure 3: Gordian knot.

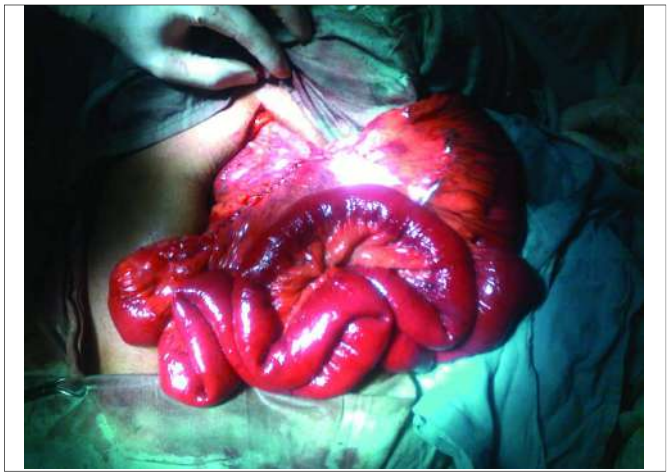

Figure 4: Repair of the mesenteric rent.

DISCUSSION

Patients with CTMH usually present with ischemia due to vague symptoms and absence of specific and sensitive investigation.[8] Common mode of presentation among adults is with full blown peritonitis or obstruction with impending bowel ischemia.[8] Incidental findings of hernias are also reported for abdominal operations during unrelated reasons. Routine CT abdomen has only been able to suspect the hernias once signs of bowel necrosis have set-in. Lack of a diagnostic sign is suggested by the reported number of cases being reported late and associated morbidity of the reported cases in the literature. Even a few cases have been reported to be diagnosed on autopsy. Diagnosing the disease at a pre-gangrenous stage is of paramount importance.

A comprehensive research article, published nearly 40 years ago, suggested the diagnosis of CTMH using CT angiogram.[9] In this case, the patient underwent a number of investigations and consulted surgical colleagues for her symptoms. However, none of the investigations were able to diagnose the cause of pain or the CTMH. "Swirl sign" or "Whirlpool sign" on a CT scan are considered a lead for intestinal volvulus or intestinal obstruction. These signs have been associated with CTMH by various authors.[9,10] We suggest that the diagnosis of such rare cases should be kept in mind and a CT angiogram should be advised in such cases where the cause of abdominal pain is not relieved by simple treatment modalities and the radiologists should be asked to comment on the presence of 'swirl sign' for SMA in cases with episodic crampy abdominal pain.

In cases where the diagnosis of CTMH is delayed or missed, the morbidity is high and can range from subacute intestinal obstructions to bowel ischemia, peritonitis or even death of the patient due to undiagnosed peritonitis.

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Tuberous Sclerosis Complex with Gingival Enlargement in an Adolescent

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ABSTRACT

Tuberous sclerosis complex (TSC) is an autosomal dominant, multisystem genetic disorder. It is characterised by formation of benign hamartomas, neurofibromas, and angiofibromas located in different organs. We describe a case of a 13-year boy who complained of gingival enlargement. Clinical examination showed distinctive dermatological signs like hypopigmented macules, shagreen plaques, miliary fibromas, fibrous plaques and multiple angiofibromas. Oral manifestation included localised gingival enlargement. Gingivectomy was performed and the excised tissue was submitted for histopathological examination. The microscopic examination of gingival tissue revealed multiple bundles of collagen fibres with proliferating fibroblast and multiple proliferating blood vessels in the connective tissue. The clinical and histopathological findings were consistent with gingival angiofibromas of TSC. Gingivectomy allowed the patient to have better function and aesthetics. Periodontal examination in conjunction with dermatological examination is important for early diagnosis of TSC.

Key Words: *Gingival enlargement. Gingivectomy. Neurocutaneous syndromes. Tuberous sclerosis.*

INTRODUCTION

Tuberous sclerosis complex (TSC) is an autosomal dominant disorder causing hamartomas and neoplastic lesions at various sites throughout the body.[1] The classical triad of TSC consists of seizures, mental retardation, and angiofibroma.[2] In 1862, von Recklinghausen was the first to describe the condition.[2] It is associated with mutations of two genes: TSC1 (hamartin) and TSC2 (tuberin). These proteins, hamartin and tuberin, form a complex that regulates cell proliferation and differentiation.[1,3] Recent studies estimate a frequency of 1/6,000 to 1/10,000 live births and a population prevalence of around 1 in 20,000 and two-thirds of all cases do not have any familial history.[3]

TSC manifests with variable signs and symptoms together with cutaneous (approximately 70% of cases), neurological (approximately 50% of cases), cardiac (approximately 30% of cases), renal and oral (approximately 11% of cases) involvement.[4] Oral manifestations consist of dental enamel pitting in both primary and permanent dentitions and oral fibromas.[5] Gingiva is the most common site for oral fibromas appearing as small nodules in the gingiva or as gingival enlargement involving attached and interdental gingiva.[5]

Here, we present a case report of TSC in a pediatric patient that showed definite dermatological and periodontal features. It is essential to highlight the importance of periodontal examination for the early identification of the disease.

CASE REPORT

A 13-year male patient reported to the outpatient department of Dental Institute, Rajendra Institute of Medical Sciences, Ranchi, India in November, 2016. The patient complained of gingival enlargement that was increasing in size over the last 6 months, leading to compromised aesthetic, poor masticatory function, and oral hygiene procedures.

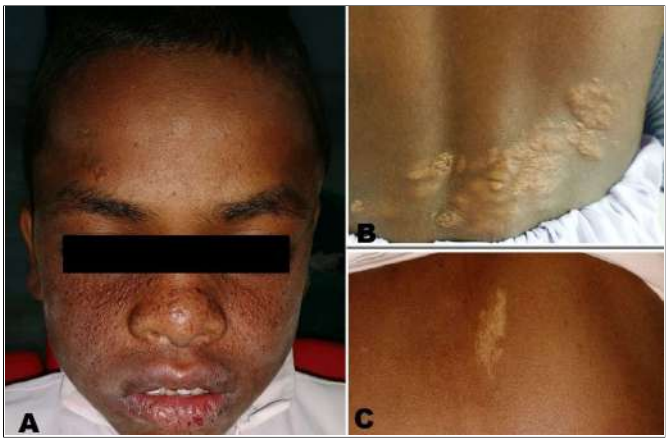

Figure 1: (A) Multiple angiofibromas seen on face, involving malar region, nose and forehead. (B) Shagreens patches. (C) Ash-leaf spot.

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The general clinical examination of the patient revealed multiple facial angiofibromas, shagreen patches, hypomelantic maculae, fibrous plaques, and miliary fibromas. The facial angiofibromas were asymptomatic, multiple, 1-3 mm in size, well defined, round to oval, brown, firm, smooth papules spreading bilaterally over malar region of face, and crossing the bridge of nose, exhibiting a symmetrical butterfly wing-like pattern (Figure 1A). Multiple miliary fibromas and fibrous plaques were present on neck and forehead as well. Multiple leathery raised patches, having bumpy/orange peel-like surface (shagreen patches) were present in the lumbosacral area (Figure 1B). Hypomelanotic maculae (ash-leaf spots) measuring 5 cm in size were present over upper back region (Figure 1C).

Intraoral examination revealed enlargement of the anterior region of maxillary and mandibular gingivae. The swelling was more pronounced between laterals and canines with ballooning enlargement of gingival papillae. The right lateral incisor had gingiva covering till two-thirds of the crown. The overlying gingiva was pink, non-erythematous, firm in consistency, having a smooth surface with loss of stippling (Figure 2A). Other findings included displacement of teeth with shift in midline of maxillary central incisor and spacing present in anterior aspect of maxillary and mandibular arch.

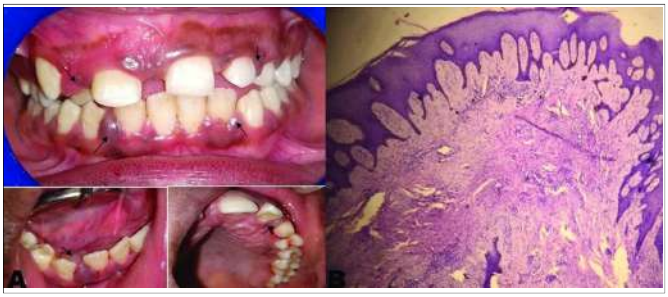

Figure 2: (A) Gingival enlargement involving maxilla and mandible anterior segment. (B) Photomicrograph showing connective with proliferating fibroblast, dilated capillaries and bundles of collagen fibers (hemotoxylin and eosin; original magnification x 10).

Physical examination of the patient did not show any signs or symptoms of cardiovascular, endocrine, respiratory, immune, musculoskeletal or neurological disorder. A routine hematological investigation conducted was within normal physiologic limits. Urine examination showed calcium oxalate crystals and traces of protein. Echocardiography (ECG) and fundoscopy examination showed no abnormalities. Magnetic resonance imaging (MRI) of brain showed solitary well defined nodules in subependymal location in lateral wall of body of bilateral lateral ventricles. No genetic test was performed due to financial constraints of the patient.

The diagnosis of TSC was confirmed by complete physical and oral examination, which included features of major and minor criteria proposed by Nosthrup and Krueger in 2012. After complete oral and periodontal examination, phase I therapy was planned for the patient, which included gentle scaling followed by institution of oral hygiene instructions. Examination on subsequent visit showed the persistence of the gingival overgrowth, which did not subside even after adequate plaque control measures. Surgical procedure was planned to reshape and recontour the gingivae to its original form, using gingivectomy and gingivoplasty techniques. An informed consent was taken from the parents prior to the procedure. Under local anesthesia, external bevel incisions were made, using Orban's and Kirkland knife over the maxillary and mandibular anterior segment of gingiva and the gingiva was reshaped. Post operative instructions and medications were prescribed to the patient. The excised tissue was sent for histo-pathological examination. The section revealed parakeratinised squamous epithelial lining 15-20 cells thick, with multiple slender branching rete ridges. The underlying connective tissue comprised of multiple bundles of collagen fibres with proliferating fibroblasts and multiple dilated blood vessels. Overall, the histopathological features were compatible with oral angiofibroma (Figure 2B). The clinicopathological correlation was conclusive of gingival angiofibromas in association with TSC.

The patient was followed-up 10 days after the surgical procedure and the healing took place uneventfully. Periodontal evaluation was done every month till six months and no recurrence of gingival enlargement was observed.

DISCUSSION

TSC is a complex, multisystem disease that requires a lot of knowledge, care, and follow-up with a multidisciplinary approach in order to provide an effective diagnosis and management of the patient. At the Tuberous Sclerosis Consensus Conference 2012, the clinical diagnostic criteria of TSC were revised and a new classification system, based on major and minor findings, was established. Skin and dental findings comprise 4 of 11 major features and 3 of 6 minor features in the diagnostic criteria.[1] A definite diagnosis is defined as the presence of at least 2 major features or 1 major and 2 minor features and a possible diagnosis is defined as the presence of either one major feature or $\geq$2 minor features.[1] In this patient, a definite diagnosis of TSC was made according to the presence of 3 major criteria (hypomelanotic macules, shagreen patches and multiple facial angiofibromas) and one minor feature (gingival fibromas). Genetic test was advised to the patient but was not done due to financial constraints of the family.

Gingival fibromas are listed as minor feature and occur in about 20-50% of individuals with TSC with greater frequency in adults than children.[1,2] In this patient,

gingival growth involved the interdental gingiva. Similar to this case, Sparling *et al*.[5] and Ammari *et al*.[6] observed gingival fibromas mostly on attached or interdental gingiva. The differential diagnosis of gingival lesions includes inflammatory or drug-induced gingival enlargement. Most patients of TSC suffer from seizures and are treated using antiepileptic drugs, but this patient was not under any medication that could cause gingival enlargement. Most of the studies showed gingival hyperplasia related to usage of antiepileptic drugs, and histological findings were consistent with fibrous hyperplasia.[7-9] The oral hygiene maintenance was adequate and inflammatory enlargement was ruled out. These findings correlated with microscopy which was characteristic of angiofibroma instead of fibrous hyperplasia. Korol *et al*.[10] also found gingival enlargement correlating with angiofibroma in a patient of TSC. The gingival fibromas were removed by gingivectomy under local anesthesia as it interfered with patient function and oral hygiene. Other authors also suggested similar procedure for treatment of gingival overgrowth.[9,10]

The dermatological manifestation observed in this patient included facial angiofibromas, hypomelanotic macules and shagreen patches. Facial angiofibromas occur in about 75% of TSC patients and remain a major feature for diagnosis with onset age between 2 to 5 years.[1] In this patient, the angiofibromas were symmetrically present almost in the form of butterfly wings involving the nasolabial folds, malar region and, nose. Several cosmetic treatments have been suggested including curettage, surgical excision cryosurgery, electrosurgery, dermabrasion, pulsed dye laser and CO_2 laser.[6] However, the recurrence rates are considered to be high. In our report, the patient was too young and was advised to postpone any of the above procedures till 18 years of age. Similar facial and oral papules can be seen in Cowden syndrome, Birt-Hogg-Dube syndrome and multiple endocrine hyperplasia type 1 and should be differentiated from TSC.[5] Hypomelanotic macules or ash-leaf spots appearing at birth or infancy are significant as they are observed in 90% of the individuals with TSC.[1] Shagreen patches are observed in about 50% of individuals and have an onset during the first decade of life.[1] These are large plaques present on the lower back having a bumpy or orange-peel-like surface.

In conclusion, this case report highlights the importance of periodontal examination in achieving the diagnosis of TSC. This case report describes a child patient with TSC having gingival angiofibromas, which were managed with gingivectomy. Skin and oral examinations should be performed annually and are important for early diagnosis and treatment of these patients.

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Kissing Nevus of the Penis

Shan Jiang[1], Yan Chen[1], Taylor E. Hinchliffe[2], Tianfu Wu[2] and Tyring Stephen[3]

ABSTRACT

Kissing nevus is a very rare congenital melanocytic nevus. Here, we describe one case of kissing nevus on the penis. A 15-year boy presented with asymptomatic black to dark brown color patches on his penis. Histopathological findings showed that there were nests and cords of nevus cells in upper dermis. No significant cytologic atypia and mitoses were noted. Immunohistochemical stains revealed a partial positive for HMB-45 only in upper dermis and a stronger positivity for S-100 in almost all nevus cells. We diagnosed the lesion as kissing nevus of penis. The patient and his parents refused further treatment, and the patient is being followed in our clinic.

Key Words: *Kissing nevus. Divided nevus. Melanocytic nevus. Penis.*

INTRODUCTION

Kissing nevus, also called divided nevus, is a rare congenital melanocytic nevus. It is usually located on adjacent parts of the upper and lower eyelids and it appears as one whole lesion when the eyelids are closed.[1] Other types and sites of kissing nevus have been reported, but are rare. Here, we report one case of kissing nevus on the penis.

CASE REPORT

A 15-year boy presented with a 10-year history of asymptomatic black to dark brown color patches on his penis. The color of these lesions had not apparently changed since it had been noticed for the first time, but the size gradually became larger with aging. He was in good health and had no history of trauma. He had no personal and family history of melanoma. On physical examination, two different size of black to dark brown color patches (one was 1.5x1.3 cm, another was 1.3x0.9 cm) were located on the left dorsal portion of the glans penis and inner foreskin, respectively (Figure 1). These two lesions were seen on each side of coronal sulcus. There was non-pigmented normal skin between these two lesions. When the prepuce was retracted, these lesions could overlap each other. A skin biopsy of the lesion on the prepuce was performed. Histopathological findings showed that the epidermis was normal. There were nests and cords of nevus cells in upper dermis and type A nevus cells in the nests contained moderate amounts of melanin granules. No significant cytologic atypia and mitoses were noted (Figures 2A and 2B). Immunohistochemical stains revealed a partial positivity for HMB-45 only in upper dermis (Figure 2C) and a stronger positivity for S-100 in almost all nevus cells (Figure 2D). Based on these findings, we diagnosed the lesion as kissing nevus of penis. The patient and his parents refused further treatment, and the patient is being followed in our clinic.

DISCUSSION

Kissing nevus is an uncommon melanocytic nevus, often located on adjacent sites of the body and separates during embryogenesis. The term "divided" nevus was first used to describe a congenital melanocytic lesion on the eyelids by Von Micheal in 1908. Fuchs presented the name of "kissing" nevus in 1919. The kissing nevus in eyelids is the common clinical type. It may originate during the period of lid fusion, between the 9th and 20th week of gestation. The melanoblasts at this site could be divided when the eyelids separate around the 20th week.[2] However, kissing nevus of the penis is extremely rare. We could find 13 articles with only 19 cases being reported in the English literature. We found that 12 patients (63.2%) were reported from Asian countries. This tendency is consistent with how kissing nevus usually occurs among White or Asian populations.[2] The embryological features of external genitalia could disclose the mechanism for the development of kissing nevus on the penis. During the 11th to 14th gestational week, two invaginations occur in the distal edge of penis. The division of epithelial preputial placode forms the glans and prepuce.[1] There is consensus that kissing nevus forms during this period. However, whether the melanoblasts migrate to the lesion site before or after separation of the epithelial preputial placode still remains controversial. Desruelles *et al*. thought that melanoblasts migration precedes the embryological separation and the kissing nevus may continue to

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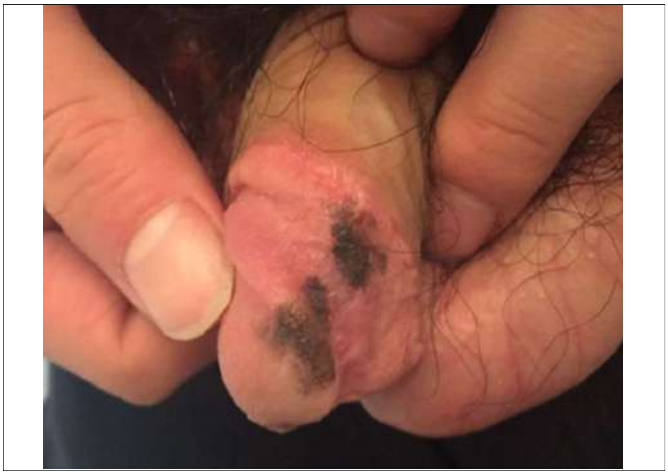

Figure 1: Two black color patches located on the glans penis and inner foreskin. These two lesions were separated by coronal sulcus.

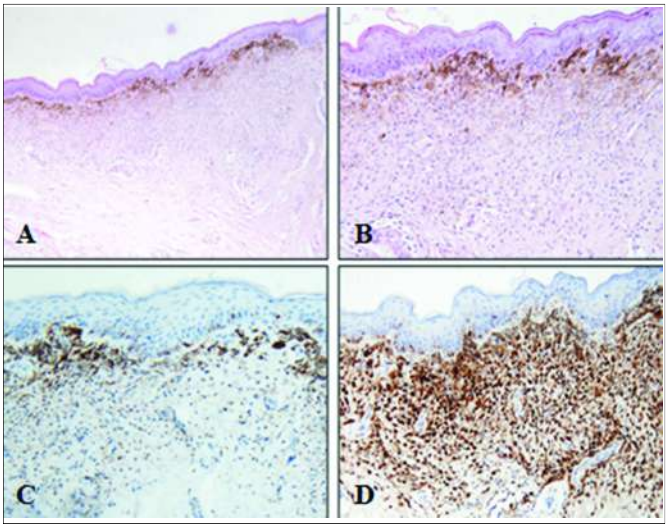

Figure 2: (A and B): Histopathological examination of a black color patch on the inner foreskin. Nests and cords of nevus cells are present at upper dermis and type A nevus cells in the nests contain moderate amounts of melanin granules. No significant cytologic atypia and mitoses are noted. (H&E, A: ×40, B: ×100). Immunohistochemical staining for HMB45 (C: ×100) and S100 (D: ×100). Nevus cells show a partial positive for HMB45 only in upper dermis. However, there is strong reactivity for S100 in almost all nevus cells.

develop independently after this separation.[1] In contrast, Kono *et al*. and Mendes *et al*. suggested that kissing nevus originates from a single pigmented lesion and melanoblasts migrate after completion of embryological separation.[3,4]

The diagnosis of kissing nevus of penis depends mainly on clinical findings and histological evaluation. Histopathological findings show that mitoses and atypia are absent from the nevus cells. Immunohistochemical stainings for S-100, HMB-45, melan-A and Ki-67 are helpful in the differential diagnoses of nevus versus malignant melanoma. Recently, dermoscopy has proved a very useful and non-invasive method in the diagnosis of pigmented lesions on skin or mucous membrane. It can avoid unnecessary surgery or biopsy, and allow long-term follow-up.[4] Dermoscopy may be an ideal alternative method for those patients with kissing nevus of penis.

Therapy for kissing nevus of the penis should consider the functionality and aesthetic outcome. Surgical resection, followed by skin grafting from the lower lip[5] or remnant foreskin,[6] has good results and there was little scaring and no loss of sensation. Laser therapy has also proved to achieve good cosmetic results.[7,8] Of the 19 reported cases of kissing nevus of the penis, only one case of malignant melanoma,[5] and one case of pigmented epithelioid melanocytoma (a borderline melanocytic tumor) have been reported.[9] Therefore, close follow-up without non-invasive therapy, such as dermoscopy, is also an alternative option.

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Pseudohypoaldosteronism Type II: A Young Girl Presented with Hypertension, Hyperkalemia and Metabolic Acidosis

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ABSTRACT

Pseudohypoaldosteronism (PHA) type II is an extremely rare disorder which presents with hypertension, hyperkalemia, and normal anion gap metabolic acidosis. PHA II is also known as familial hyperkalemic hypertension, Gordon syndrome, and chloride shunt syndrome. PHA II is an autosomal dominant disorder and is caused by mutation in WNK1, WNK4, CULLIN3, KLHL3, OSR, SPAK gene. The expression of these proteins is limited to the distal convoluted tube and collecting duct of the kidney. PHA II usually responds to salt restriction and thiazide diuretics. We are reporting here a case of 16-year girl who presented with generalised fatigue and shortness of breath, and blood pressure (BP) of 220/110 mmHg. Laboratory investigation showed hyperkalemia, normal anion gap metabolic acidosis, and hypercalciuria. Workup for secondary causes of hypertension was negative. She responded to thiazide diuretics and her BP is well controlled, and acidosis and hyperkalemia are corrected.

Key Words: *Pseudohypoaldosteronism type II. Gordon syndrome. Hypertension. Hyperkalemia. Hypercalciuria. Metabolic acidosis. Thiazide diuretics.*

INTRODUCTION

Pseudohypoaldosteronism type II (PHA II) is a rare syndrome and is also known as familial hyperkalemic hypertension, Gordon syndrome, and chloride shunt syndrome. It is inherited as an autosomal dominant pattern.[1] It is caused by mutation in WNK1 and WNK 4 genes (with no lysine kinases) on chromosome 12 and 17, respectively. Some researchers have reported the genetic defects in Kelch-like 3 (KLHL3) or Cullin 3 (CUL 3), OSR (oxidative stress-responsive kinase), SPAK (Ste20-related proline alanine-rich kinase) etc. as a cause PHA II.[2] PHA II is manifested by hypertension, hyperkalemia, normal anion gap metabolic acidosis, decreased renal potassium excretion, hypercalciuria, low or low normal plasma renin and variable level of aldosterone, either low or normal. Thiazide diuretics effectively reverse the hypertension and hyperkalemia.[3]

We are reporting a case of PHA II, which responded to thiazide diuretics.

CASE REPORT

A 16-year Kuwaiti girl presented with generalised fatigue and shortness of breath for four hours. She visited a polyclinic and her blood pressure (BP) was 220/110 mmHg. She gave history of seasonal bronchial asthma for which she was taking salbutamol inhaler as per requirement. When she arrived to Emergency Department, her vital signs were: BP of 160/90 mmHg, pulse at 120 beats/minute, and O_2 saturation 96% on room air. Systemic examination, including fundoscopic examination, was unremarkable. She was neither in a state of exacerbation of bronchial asthma nor in hypertensive pulmonary edema.

Investigations showed normal blood counts and renal functions. However, potassium level was 6.1 mmol/L, HCO3 was 18 mmol/L, anion gap was 11.1, and calcium level was 2.3 mmol/L. Arterial blood gases (ABG) analysis showed metabolic acidosis. Coagulation profile urine microscopy and chest X-ray were normal. ECG showed sinus tachycardia with heart rate of 120 beats/minute without left ventricular hypertrophy. 24-hour urinary protein and creatinine clearance was normal and 24-hour urinary catecholamines were normal, whereas 24-hour urinary calcium was high at 8.5 mmol/day (normal 2.5-7.5). TSH, aldosterone (124 pmol/L) and renin (<5.6 ng/L) levels were normal. Echocardiography showed concentric left ventricular hypertrophy with ejection fraction of 65%. Ultrasound of abdomen/pelvis normal, Doppler of renal vessel, MRI of adrenals, MRA of renal arteries and immunological investigations (ANA and ANCA) were all normal.

Initially, she was managed with amlodipine 5 mg OD, and calcium resonium 30 gm TDS with lactulose to treat her hyperkalemia. Later, she was switched to hydrochlorothiazide and amlodipine. Her home BP readings are controlled on thiazide diuretic, and currently she is off the amlodipine. Her electrolytes are shown in Table I.

Her family was screened with serum electrolytes and one of her sisters had high serum potassium levels. She

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Table I: Comparison of serum electrolytes before and after thiazide therapy.

Electrolytes	Pre-thiazide	Post-thiazide
Potassium	5.7 & 5.8	5.1* & 4.9**
Chloride	116 & 115	117 & 115
Bicarbonate	17	16 & 19

**Done after one week; ** after two weeks of thiazide diuretic.*

need genetic analysis, especially WNK gene, which has not done yet.

DISCUSSION

PHA II is the opposite of Gitelman's syndrome, which presents with low blood pressure, hypokalemia, hypochloremic metabolic alkalosis, and hypocalciuria.[4] Initially, it was considered to be caused by a genetic defect in the NaCl cotransporter (NCC), the target transporter of thiazides. However in 2001, PHA II was reported to be caused by abnormalities in two types of genes known as WNK1 and WNK4 genes.[5] Later, Yang *et al*. confirmed the WNK4 mutation in mice, manifesting hypertension secondary to electrolyte abnormalities, acidosis and increased circulating blood volume.[6] There are reported cases in which PHA II was not linked with genetic mutation in WNKs. Some researchers have reported PHA II is caused by the genetic defects in Kelch-like 3 (KLHL3) or Cullin 3 (CUL 3), OSR (oxidative stress-responsive kinase), SPAK (Ste20-related proline alanine-rich kinase) etc.[2] The expression of these proteins is limited to the distal convoluted tube (DCT) and collecting duct (CD) of the kidney. The WNK1 and WNK4 genes are located on chromosome 12 and 17, respectively.[7] WNK4 gene negatively regulates the thiazide sensitive NCC in the DCT, which leads to volume expansion due to increased sodium and chloride reabsorption; which ultimately results in hyperchloremia and hypertension.[8] Moreover, WNK4 defect also antagonises the aldosterone sensitive renal outer medullary potassium (ROMK) channels of DCT, which leads to decreased potassium secretion through ROMK channels and hence cause hyperkalaemia.[9]

All these findings, i.e. hypertension, hyperkalemia, hyperchloremic metabolic acidosis, hypercalciuria were present in this reported case. Moreover, hypercalciuria typically occurs with WNK 4 gene mutation in subjects of PHA II.[10] In this reported case, hypercalciuria meant that she might have mutation in WNK4 gene. In PHA II, hypercalciuria is caused by increased NCC activity due to defect in WNK genes. Uninhibited NCC activity causes increase sodium reabsorption and decrease calcium reabsorption in PCT which results in hypercalciuria and hypertension. Another mechanism for hypercalciuria is due to down regulation of transient receptor potential V5 channel (TRPV5) activity. It reduces calcium reabsorption in DCT, which may result in urinary calcium loss and osteoporosis.[7]

Thiazide diuretics effectively reverse hypertension and metabolic abnormality in patients of PHA II. Thiazide diuretics are the pharmacological inhibitor of the NCC activity, hence inhibiting the NCC activity normalises hyperkalemia, hyperchloremic metabolic acidosis and hypercalciuria. Moreover, thiazides are six times more sensitive in treating hypertension in patients with PHA II than in individuals with essential hypertension.[7] Hyperkalemia, hypertension, hypercalciuria with hyperchloremic metabolic acidosis in the presence of normal renal function may alert clinician to think of pseudohypoaldosteronism type II. It is associated with mutation of multiple genes, especially WNK genes; and thiazide diuretics effectively reverse the metabolic abnormalities, including hypertension.

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Acute Onset of Quadriplegia Secondary to Hypoparathyroidism: Mimicker of AMAN Variant of GBS

Syeda Saba Zaidi[1], Tahira Siddiqui[1] and Sarwar Jamil Siddiqui[2]

ABSTRACT

Acute onset of quadriplegia is a rare phenomenon seen with hypocalcemia due to hypoparathyroidism. We describe a 33-year gentleman who presented with weakness of all four limbs and areflexia. ECG showed QT abnormality. Nerve conduction study revealed normal sensory and significantly low motor CMAP amplitudes in both the upper and lower extremities. This nerve conduction study can be seen in acute motor neuropathy of various etiologies, among which GBS is the most worrisome. Our patient found to have low calcium and parathyroid hormone level. His symptoms improved after calcium replacement. Occurrence of quadriplegia in hypoparathyroidism, and its improvement after correction of calcium, suggests crucial role of calcium in neuromuscular transmission. One should suspect electrolyte imbalance, like hypocalcemia in patients presenting with nerve conduction features of AMAN variant of GBS.

Key Words: *Hypocalcemia. Quadriplegia. Hypoparathyroidism. Nerve conduction study. Acute motor neuropathy.*

INTRODUCTION

Primary hypoparathyroidism is defined as inadequate parathyroid hormone activity, due to which serum calcium concentration falls below reference range. Causes of primary hypoparathyroidism can be acquired or hereditary. On the other hand, secondary hypoparathyroidism is a state in which parathyroid hormone levels are low due to primary pathologies that lead to hypercalcemia. Common clinical characteristics of hypoparathyroidism include muscle spasm, paresthesias, perioral numbness, tetany and seizures. Other neurological manifestations are depression, psychosis, movement disorders like chorea, athetosis, bradykinesia, hemiballism and peripheral neuropathy.[1,2]

We describe a case of hypocalcemia due to idiopathic primary hypoparathyroidism presented with acute onset of quadriplegia and sequential changes in nerve conduction study. To our knowledge, this is the first case report which elaborates the significance of low motor CMAP amplitude changes in hypocalcemic weakness.

CASE REPORT

A 33-year gentleman, who was in a good health a day before presentation, suddenly developed stiffness in the lower limbs, followed by heaviness and a tendency to fall. Over the next 24 hours, his weakness progressed to an extent that he was not able to lift his legs off the bed and had difficulty in holding things in his hands.

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On examination, he was alert and oriented. Visual fields, optic discs and extraocular muscles were normal. There was no facial paralysis; gag reflex was intact and neck flexion was strong. Bulk was normal, tone was decreased in all four limbs, and power grade was 0/5 in all limbs (MRC scale), except in right upper limb where it was 1/5. Reflexes could not be elicited even on reinforcement. Plantars were equivocal. Joint position, vibration, touch, position were intact.

Investigations showed normal hemoglobin, leucocyte count, blood glucose, urea, serum creatinine, electrolytes, albumin, and liver function. Serum calcium level was 5.6 mg/dl (8.5-10.2 mg/dl) and phosphate level was 2.6 mg/dl (2.5-4.5 mg/dl). Serum magnesium was 1.5 mg/dl (1.7-2.2 mg/dl). Serum PTH was 5.98 pg/ml (16-87 pg/ml). Vitamin D level was found to be 20.6 ng/ml (< 30 ng/ml). Electrocardiogram revealed QTc of 0.54 s.

Electrophysiological study was done. Motor study in upper extremities show low CMAP amplitudes of median nerve on both sides, with normal distal latencies and conduction velocities. Right ulnar nerve showed low CMAP amplitude, normal distal latencies and conduction velocities. In lower extremities, Peroneal and tibial motor studies showed low CMAP amplitudes, normal distal latencies and conduction velocities. Absent H reflex was consistent with polyneuropathy. Although peroneal F responses were absent, this finding was of unclear significance because peroneal F responses are difficult to obtain in some normal individuals (Table I). Sensory nerve conduction studies were normal.

A diagnosis of hypocalcemia due to hypoparathyroidism leading to significant motor CMAP amplitude changes in both upper and lower extremities was made. Patient received 10 ml of calcium gluconate IV (90 mg elemental calcium). His symptoms started to improve after 24 hours. He was kept on oral combination of Calciferol:100IU

Table I: Motor nerve conduction study.

		Latency		Dist	Amplitudes		NCV		F-lat		Duration	
Nerve-muscle	Stim-site	R	L		R	L	R	L	R	L	R	L
Median(APB)	WRIST	3.3	3.4	7	1.8	0.5	24.5	NR	24.5	NR	4.1	5.6
Median (APB)	ELBOW	7.2	6.8	23	1.1	0.5	59	68			4.7	5.6
ULNAR (ADQ)	WRIST	3.3	2.3	7	1.3	4			26.9	24.5	6.7	6.8
ULNAR (ADQ)	D. ELBOW	6.5	5.4	19	1.3	3.4	59	61			7.6	7
ULNAR (ADQ)	P. ELBOW	8.2	6.9	10	1.2	3.7	59	67			7.6	7.3
Post TIBIAL (AH)	ANKLE	6.4	6.6	9	0.7	0.9			47.2	49.4	14.7	10.9
Post TIBIAL (AH)	KNEE	15.4	15.3	40	0.6	0.8	44	46			13.5	10.2
PERONEAL (EDB)	ANKLE	3.7	5.7	8	0.5	0.5			NR	NR	10.6	8.9
PERONEAL (EDB)	D. KNEE	10.6	12.1	37	0.5	0.4	54	58			11	11.9
PERONEAL (EDB)	P. KNEE	12.2	13.6	9	0.4	0.5	56	60			11	11.6
PERONEAL (TA)	DFH	2.7	2	10	0.2	0.2					10.1	9.4
PERONEAL (TA)	PFH	4	3.2	9	0.2	0.2	69	75			9.3	8
H REFLEX	POP. FOSSA	NR	NR									

and Calcium: 400mg, thrice daily. He started walking in next few days with support and discharged home with outpatient physiotherapy advice.

DISCUSSION

Hypoparathyroidism is a condition of parathyroid hormone deficiency. Primary hypoparathyroidism is defined as inadequate parathyroid hormone activity, due to which serum calcium concentration falls below reference range.

While on the other hand, secondary hypoparathyroidism is a state in which parathyroid hormone levels are low, due to primary pathologies that lead to hypercalcemia.

Primary hypoparathyroidism can occur due to acquired or inherited causes. Common causes of acquired hypoparathyroidism include surgical removal of parathyroid adenoma and autoimmune parathyroid disease. Other rare causes include radioactive iodine treatment, sarcoidosis, hemosiderosis, hemochromatosis, and parathyroid gland infiltration by metastatic deposits. Inherited hypoparathyroidism occur with other developmental anomalies such as failure of adrenal glands, thyroid gland, ovaries, thymus associated with oral candidiasis, baldness and vitiligo.

Literature review revealed five cases on effect of low calcium on membrane potential and subsequent changes in nerve conduction studies. To our knowledge, this case is the first of its kind in published literature.

Gomez reported a 25-year gentleman, known case of hypoparathyroidism.[3] At the age of 20 years, patient developed weakness of limbs with depressed tendon reflexes. On nerve conduction studies, found to have decreased nerve conduction velocities of median motor and ulnar nerves. Nerve biopsy showed axonal degeneration.

Dionisi described a 15-month child with distal sensorimotor peripheral neuropathy and hypoparathyroidism, which was recovered after treatment with vitamin D over five months.[4]

Gay and Grimes described a 68-year man with large fiber sensory neuropathy and hypoparathyroidism.[2] It also improved after vitamin D and calcium therapy for 6 weeks.

Goswami reported another case of idiopathic hypoparathyroidism whose nerve conduction studies revealed axonal sensorimotor neuropathy.[5] He received calcium and vitamin D for two years. There was slowly progressive betterment in neuropathy both clinically and on electrophysiologically. The occurrence of peripheral neuropathy in conditions associated with hypocalcemia such as hypoparathyroidism and osteomalacia and its reversibility after normalisation of calcium and vitamin D suggests its crucial role in the functioning of the peripheral axons.

Decreasing concentration of calcium reversibly increased inward sodium currents, moderate depolarization and increased nerve excitability, this results in failure of excitation of muscle fibers by supramaximal stimulation of peripheral nerves, thus resulting in decreased CMAP of tested nerves. This mechanism is similar to the changes seen in hypokalemic flaccid weakness.[6]

Hypoparathyroidism was diagnosed in our patient on the basis of low serum calcium, normal serum phosphate, and very low serum PTH. Nerve conduction studies showed severely reduced motor CMAP amplitude, a pattern similar to pure motor axonal neuropathy, which can be seen in AMAN variant of GBS or porphyria. Patient improved markedly after calcium and vitamin D replacement.

Appropriate diagnosis always requires a combination of careful history, examination and accurate interpretation of diagnostic testing. Failure to recognize the easily reversible causes of acute quadriparesis can lead to erroneous diagnosis, inappropriate treatment, and significant morbidity, related to treatment complications. This case report highlights the fact that nerve conduction studies can be misleading in patients presenting with flaccid quadriparesis. One should suspect electrolyte

imbalance, like hypocalcemia, in patients presenting with nerve conduction study of AMAN variant of GBS.

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A Gastric Duplication Cyst Initially Mimicking Staghorn Calculus

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ABSTRACT

Gastric duplication cyst is an uncommon anomaly typically found in the greater curvature of the stomach. It is usually diagnosed in children and quite difficult to diagnose in adult because of variable presentation. We present a 76-year woman who was admitted for abdominal pain of few weeks' duration. Her X-ray showed calcification in the region of left kidney, and she was subsequently managed with a presumptive diagnosis of staghorn calculus. Computerized Tomography (CT) scan of her abdomen disclosed calcification in the gastric wall for which an endoscopic ultrasound and biopsy was done. Findings were consistent with the presence of gastric epithelium and a diagnosis of gastric duplication cyst was made. Surgery was the continuation of care. The presence of symptomatic gastric cyst in an elderly patient is very rare. This is the first case in which gastric duplication cyst mimicked staghorn calculus on abdominal X-ray. Consequently, this diagnosis, though rare, should be considered in the differentials of upper abdominal pain.

Key Words: *Calculi. Staghorn. Cysts. Abdominal pain. Gastric duplication cyst. Stomach. Congenital anomaly. Gastric wall calcification.*

INTRODUCTION

A gastrointestinal duplication cyst is an uncommon abnormality, usually involving the ileum.[1-3] Gastric duplication cysts are very rare with an incidence of about 4 - 8% among all gastrointestinal cysts.[2] In addition, most cases are typically diagnosed in infancy or early teens.[4]

This case report describes a 76-year lady who was initially misdiagnosed and treated for staghorn calculus, which is a unique case.

CASE REPORT

A 76-year Caucasian lady with medical history remarkable for gastric polyps, gastroesophageal reflux disease (GERD), diverticulosis without any episode of diverticulitis, who endorsed abdominal pain of several weeks' duration. Pain was gradual in onset, 10/10 at its worst, located at the left lower quadrant, non-radiating with no specific aggravating or alleviating factors. Of note, the patient had presented with similar complaints a couple of weeks prior to onset of the current symptoms. At that time, X-ray of the abdomen showed calcification in the left kidney region (Figure 1). Nephrologist was consulted and initial impression was staghorn calculus. Patient was advised to drink plenty of water and workup for staghorn calculus was done, which was unremarkable.

On physical examination, patient's vital sign disclosed BP 116/76 mmHg, pulse rate 85 bpm, respiratory rate 18 per minute, and body temperature was 98.4°F. On general examination, patient did not appear to be stable and not distressed. Abdominal examination revealed left lower quadrant tenderness with no guarding or rebound tenderness. Basic laboratory work revealed a normal WBC of 7300 per cubic millimeter. CT scan of abdomen was carried out to rule out diverticulitis in view of history of diverticulosis. It disclosed a calcified mass in the posterior inferior gastric wall. Based on these clinical and radiological assessments, upper GI endoscopy was planned to rule out gastrointestinal stromal tumor or other neoplastic etiologies. It showed gastritis and a gastric polyp. Further workup with endoscopic ultrasound revealed the presence of cystic area with presence of calcification. Biopsy was taken which revealed presence of gastric epithelium with no atypical cell, typical of a gastric duplication cyst.

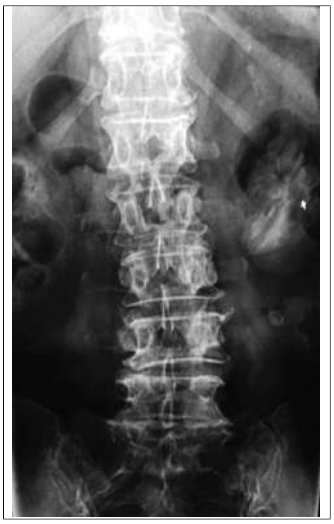

Figure 1: X-ray showing calcification similar to staghorn calculus.

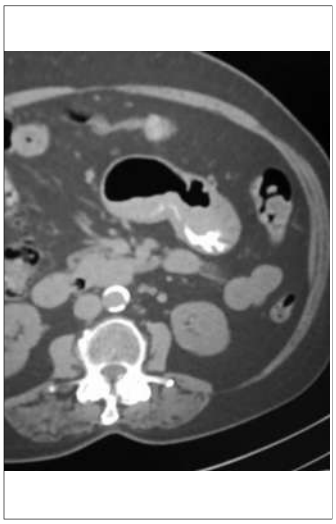

Figure 2: CT scan abdomen showing calcification of the posterior gastric wall.

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Patient was subsequently referred to surgery for further management. She underwent enucleation of the cyst through minimally invasive approach. She had uneventful postoperative course and was discharged on the third postoperative day.

DISCUSSION

Gastrointestinal duplication is a uncommon anomaly that usually involves ileum but may involve any part of the gastrointestinal tract.[1,2,5] Gastric duplication cyst is most commonly located along the greater curvature.[4,6] In adults, the diagnosis is usually challenging because of the vast differences in presentation of the patients.[7,8]

Most of the cases are diagnosed in children. Gastric duplication cyst manifest usually during childhood and majority are diagnosed before the age of one year.[4,5] In adults, presentation is non-specific and include a wide spectrum of symptoms including epigastric fullness, abdominal pain, nausea and vomiting.[1,2,4,5,7,8] Because these cysts usually lie on posterior border of stomach, they have the potential to compress the adjacent organs including pancreas, adrenal gland, kidney, and spleen. However, benign cysts usually do not present with compression symptoms, presence of compression of adjacent organs usually indicate conversion to carcinoma.[1]

Advances in technology have engendered a paradigm shift from intraoperative diagnosis to preoperative diagnosis with the help of imaging such as endoscopic ultrasound (EUS). Diagnostic criteria for gastric duplication cyst comprise stomach wall adjoining cyst wall; cyst and wall of the stomach should be surrounded by smooth muscle; and the cyst wall be composed of gastric epithelium or any kind of gut mucosa.[3,9] CT and EUS are imaging modalities of choice for diagnosis of gastric cysts. Classical finding on contrast-enhanced CT scan is a thick-walled cystic lesion with enhancement of the inner lining with occasional calcification.[10] Biopsy usually shows similarity between the mucosal lining of duplication and the region of the gut, where it is found.[5]

Differential diagnoses include gastrointestinal stromal tumor, benign and malignant neoplasm of GI tract. Natural history of gastric duplication cyst is difficult to outline. Complications include gastrointestinal bleeding, obstruction, torsion, hemorrhage and neoplastic transformation, all of which can be presenting symptoms.[3] Out of 11 reported cases of malignancy, 8 were adenocarcinoma.[4] Because of the risk of neoplastic transformation, it is important to remove the cyst completely. Various methods are used for removal of cyst, one of which is enucleation. Enucleation is considered the gold standard therapy as it causes the least disruption of normal anatomy.[9]

This is a unique case report of gastric duplication cyst as it showcases occurrence in an unusual age, at the same time masquerading as a staghorn calculus. We recommend consideration of gastric duplication cyst during the workup of upper abdominal pain.

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Managing a Large-Size Adrenal Cyst by Hand-Assisted Laparoscopic Surgery in a Young Male

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ABSTRACT

Adrenal cysts are not common and are most often non-functional and asymptomatic. Most of these cysts are found incidentally. These lesions exhibit a broad histomorphologic spectrum and may vary from benign cysts to malignant cystic neoplasms. Our patient was a 21-year male who presented with abdominal discomfort and epigastric pain and was diagnosed with left adrenal cyst on ultrasound, which was later confirmed by CT scan.

Key Words: *Adrenal cyst. Asymptomatic. Abdominal.*

INTRODUCTION

Cysts of the adrenal gland are rarely encountered in clinical practice. Presenting signs and symptoms are variable. Reportedly, adrenal cysts cases are common in women aged forty to sixty years.[1] The currently accepted classification of adrenal cysts includes four subtypes as pseudocysts (39%), endothelial cysts (45%), epithelial cysts (9%) and parasitic cysts (7%).

Furthermore, they usually present with nonspecific clinical and radiologic findings.[2] Parasitic and cancerous cysts require removal along with the entire adrenal gland, especially in view of the fact that the current diagnostic imaging yields a high probability of characterizing the lesion in the preoperative period and permits the investigator to use a suitable surgical approach.[3]

Pseudocysts often result from hemorrhage within the adrenal gland. These may also result from severe stress, birth trauma, and surgery. Adrenal cysts can also occur in association with benign and malignant tumors. In adrenal cysts, the overall incidence of malignancy is deemed to be around 7%.[4]

A complete endocrine evaluation is recommended for adrenal cysts of size 5 cm or greater (as confirmed by abdominal CT) or smaller ones in which malignancy cannot be ruled out. Small asymptomatic or nonfunctional adrenal cysts can be followed clinically without intervention. When adrenal cysts are 6 cm or greater, symptomatic or functioning or malignancy is suspected on imaging, then surgical exploration is recommended. CT scan is better for diagnosing adrenal cysts, since it can reveal adrenal cysts, which are low-density masses with smooth borders and thin walls.[4] Surgical interventions can be open, i.e. cyst enucleation or en bloc adrenalectomy, or laparoscopic intervention.[1] The laparoscopic approach, which involves minimal invasiveness, has proved to be safe and results in lesser morbidity.[1] Total adrenalectomy or laparoscopic partial adrenalectomy can be performed in cases of larger cysts in which most of the adrenal gland is compromised.[1,4]

CASE REPORT

A 21-year male presented to Shifa International Hospital with complaints of left upper abdominal pain, radiating to back and resulting in heaviness in abdomen for 15 days. He also had loose motions but had no history of any fever, vomiting, or other complaints. His laboratory reports were all within normal ranges. His preoperative blood tests were normal. Serum electrolytes including potassium and chloride were 4.4 mEq/L and 103 mEq/L, respectively. His serum creatinine was 0.75 mg/dl, serum lipase and amylase were 8U/L and 41U/L, respectively (within normal limits). CT scan showed left suprarenal cystic mass (Figure 1). A decision was made to remove it surgically by hand-assisted laparoscopy technique. Consent was taken from the patient for this surgical intervention. Incision was made on left paramedian site above the level of umbilicus. Gel port was inserted and then with the help of trocar, gas insufflation was done. Endoscopic camera was inserted via gel port and three other working ports were inserted under camera vision into the abdominal cavity. Then, left colon was mobilized medially and superomedial side of left kidney was identified. Left suprarenal area was reached and adrenal cyst was separated from surrounding structures with difficulty, especially from the spleen and pancreas, which were lying very close to it. Partial left adrenalectomy was done. During the procedure, there was a rent of approximately 0.5 cm in the left diaphragm and parietal pleura was damaged. It

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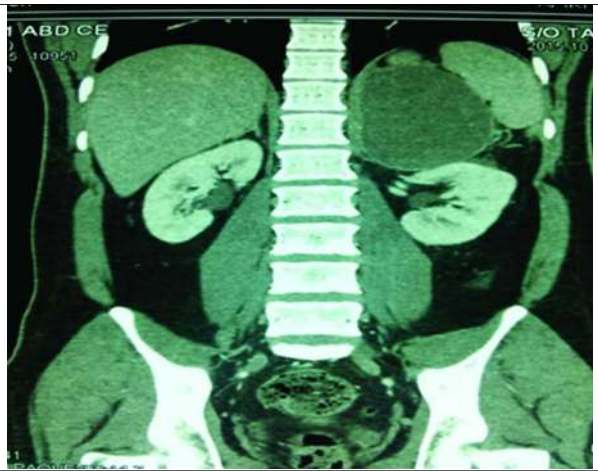

Figure 1: CT scan showing huge adrenal cyst on left side abutting the spleen.

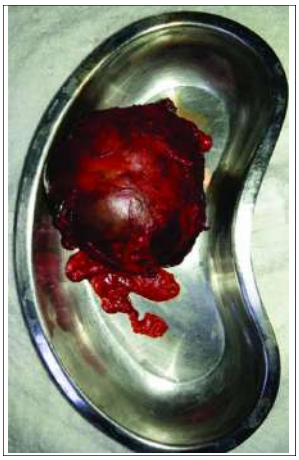

Figure 2: Gross appearance of adrenal cyst specimen after removal from patient.

was repaired intraoperatively immediately. The specimen of adrenal cyst with some adrenal gland was removed and sent for histopathology evaluation. Postoperatively, patient had sluggish bowel sounds, and was kept nil per oral. After 3 days, he had normal diet intake and was improving. He was discharged from hospital on 4th post-operative day. On follow-up after 2 weeks, he had completely recovered and was leading normal life. He had no symptoms of pain.

Grossly, the cyst was well-circumscribed and weighed 443 grams and measured 10.5x10.0x7.0 cm (Figure 2). Upon serial sectioning, yellow-color adrenal gland was identified in the wall measuring 1.0x0.5 cm. On microscopic examination, the sections of cyst showed extensive hemorrhage and necrosis. There was no evidence of malignancy. A final diagnosis of hemorrhagic cyst was made.

DISCUSSION

Adrenal cysts are uncommon lesions, with a reported incidence at autopsy of 0.064% to 0.18%.[1] Until 2010, about 600 cases have been reported in medical literature.[3] Mosty, adrenal cysts are non-functional but some cases are reported with non-specific symptoms, e.g. flank or abdominal pain.[5]

On histopathology, adrenal cysts vary from benign to malignant lesions. However, the cysts most commonly found in adrenal glands are endothelial (45%) followed by haemorrhagic pseudocysts (39%), epithelial cysts (9%), and parasitic (7%). Some reports show that pseudocysts are the most common of all adrenal cysts. Surgically treated cases show that endothelial cysts represent 48% and haemorrhagic pseudocysts are found in 42%. The proposed pathogenesis of adrenal pseudocysts include vascular neoplastic growth, malformation, and haemorrhage into the adrenal parenchyma. An uncommon condition of atraumatic haemorrhage can be categorized as stress, haemorrhagic diathesis or coagulopathy, neonatal stress, underlying adrenal tumours and idiopathic disease.[6] Some theories suggest that adrenal hemorrhage is more common in neonates.[6]

Incidental malignancy can also occur in adrenal gland. Adrenocortical cysts showing benign appearance have been found to have malignant features on extensive sampling.[4] In cases where malignancy is not suspected, percutaneous aspiration is the treatment option.[4] Surgical treatment is the only choice for cysts more than 6 cm.[4] Functional tumors should be treated following appropriate preparation.[4]

The ELISA test is best used to find out echinococcal origin of the cyst, and if malignant element is to be ruled out then fine needle biopsy is used with up to 85% sensitivity. Adrenal hormone evaluation is performed with the application of generally available laboratory tests.[3,7] As with the use of laparoscopic techniques, other methods can also be used to preserve the non-diseased part of adrenal gland.[3] However, hormone-dependent cysts, along with bacterial and parasitic ones, require a more radical procedure. In such patients, most often the treatment of choice is classical adrenalectomy in addition to cystectomy due to the need of removing the entire cyst.[3] Three factors which are important while making management plan include: (a) the functional status of the cystic lesion, (b) the probability of any incidental malignancy, and (c) the possibility of complications that can happen like hemorrhage into the cyst.[4]

For benign adrenal cysts, laparoscopic adrenalectomy is the most safe and effective treatment option. Its advantages over open surgery include shorter hospital stay, lesser amount of bleeding, and improved cosmetic appearance. With surgical removal, the chances of recurrence of cysts is rare as compared to techniques using aspiration of cysts contents.[6]

We had a young patient who underwent successful removal of adrenal cyst with partial adrenalectomy by hand-assisted laparoscopic technique. There was some difficulty in the surgery due to large size of the adrenal cyst, and its close proximity and adhesions to the spleen and the pancreas. There was a diaphragmatic rent during surgery, which was repaired on the spot. Postoperatively, patient had good recovery. There were complications of sluggish bowel sounds and vomiting postoperatively which were settled on 3rd postoperative day. Overall, we had a successful removal of the cyst

and satisfactory outcome of the whole surgical management of the cyst.

In conclusion, adrenal cyst is a rare entity which can be of large size as in our case. Even such a large adrenal cyst can be successfully removed laparoscopically without major complications peroperatively and postoperatively.

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Occult Pneumothorax: What Do We Need to Do?

Huma Sabir Khan, Abdul Wadood and Mahmood Ayyaz

ABSTRACT

Occult pneumothorax is a condition in which a patient's clinical examination and chest radiograph are normal; but there is pneumothorax on computed tomography (CT) scan. We here describe two cases of trauma whose initial survey and chest X-ray showed normal lung markings bilaterally; but CT scan done subsequently, showed pneumothorax which was managed by chest intubation. There is still an ongoing debate about the management of occult pneumothorax. Simple observation is recommended for an asymptomatic pneumothorax. However, chest intubation is recommended for patients undergoing a surgery or mechanical ventilation due to the fear of converting a close pneumothorax into a tension pneumothorax.

Key Words: *Occult pneumothorax. eFAST (extended focused assessment by sonography for trauma). Chest intubation.*

INTRODUCTION

Occult pneumothorax is a pneumothorax that is neither suspected clinically nor is evident on the plain radiograph; but rather identified on computed tomography (CT) scan or ultrasound.[1] The increasing use of eFAST (extended focused assessment by sonography for trauma) and CT scan has enabled emergency care physicians and trauma teams to diagnose occult pneumothorax, which might not be evident on a chest radiograph.[2-4] Chances to miss a pneumothorax increase, if radiographs are interpreted by trauma teams instead of qualified radiologists. Management of occult pneumothorax is still under debate. However, chest intubation appears to be the best policy for patients who need ventilatory support.

CASE REPORT

Case 1: A 24-year male presented to surgical emergency after a motor vehicle accident with polytrauma. He was hemodynamically stable. There was fracture of right femur and right tibia/fibula accompanied by tenderness in right hypochondrium. Initial supine chest X-ray and FAST (Focused assessment with sonography for trauma) were normal. However, there was clinical suspicion of blunt abdominal organ injury; hence, the patient underwent CT scan chest, abdomen and pelvis with intravenous contrast. CT scan revealed small right-sided pneumothorax which had been missed on clinical examination and supine chest X-ray (Figures 1 and 2). There was also perihepatic and perisplenic fluid with grade-II liver injury. Right-sided intubation was done. Skeletal traction was applied for the femur fracture by orthopaedic team. Abdominal injury was managed conservatively. Chest tube was taken out on 3rd day and

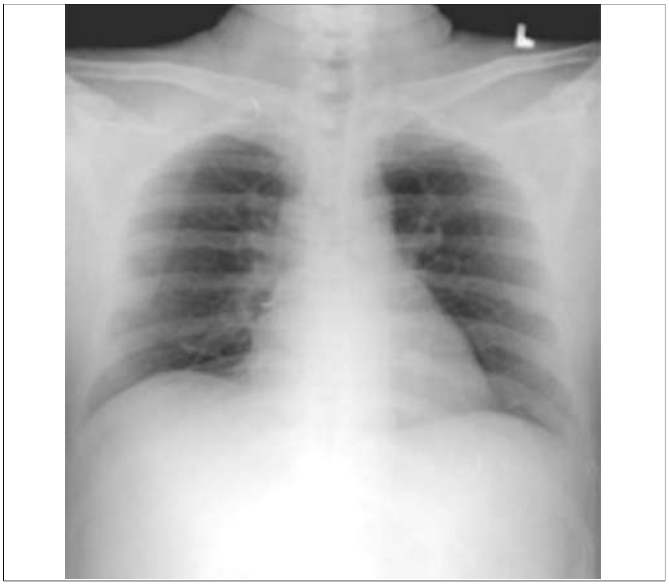

Figure 1: Anteroposterior chest radiograph of patient showing no evidence of pneumothorax.

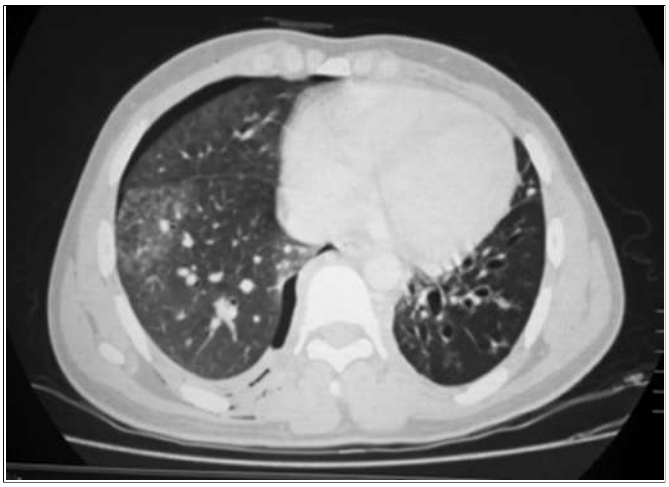

Figure 2: CT scan of patient showing right sided occult pneumothorax.

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patient was referred to orthopaedic department for management of femur fracture.

Case 2: A 20-year male presented to emergency after motor vehicle accident. Primary survey showed intact airway, breathing and stable vitals. There was an imprint mark in left hypochondrium along with slight tenderness. Patient underwent chest X-ray and FAST examination, both of which were normal. Hence, patient underwent chest, abdomen and pelvis CT scan, which showed right sided pneumothorax and grade-III liver injury. Patient was managed conservatively for pneumothorax and splenic injury by simple observation. There was no respiratory distress. Repeat CT scan was done three days later, which showed complete resolution of pneumothorax. Hence, patient was discharged and remained fine on follow-up.

DISCUSSION

The incidence of occult pneumothorax is reported to be 5% in literature.[1,2] As the conservative management of blunt trauma patients is increasing, the incidence of recognition of occult pneumothorax is also increasing.

Occult pneumothorax rate up to 76% are usually due to the fact that anteroposterior (AP) chest radiographs done in trauma patients are reported by trauma team instead of qualified radiologists.[2] This difference is due to the fact that trauma team functions in difficult conditions in emergency situations. The other factor is the low sensitivity of an AP radiograph. An erect chest X-ray is more sensitive as compared to an AP chest X-ray in detecting pneumothorax.[1] AP chest X-ray is 75% sensitive in identifying pneumothorax as compared to ultrasound, which has a 98% sensitivity.[3] CT scan is taken as gold standard for diagnosing pneumothorax, especially occult pneumothorax.[1] However, there are certain clinical signs which can guide the clinicians towards possibility of an occult pneumothorax. These include subcutaneous emphysema, rib fractures, and lung contusions.

Management of occult pneumothorax is still variable. It is recommended that observation of small occult pneumothoraxes, without tube thoracostomy in trauma patients not receiving mechanical ventilation, is likely safe.[2] In patients who have to undergo mechanical ventilation, tube thoracostomy is advised to prevent development of tension pneumothorax.[2,4] However, some studies suggest conservative management for hemodynamically stable patients undergoing mechanical ventilation for operation or short intensive care unit (ICU) stay.[5,6] Observation is advocated in occult pneumothorax in hemodynamically stable patients because tube thoracostomy is associated with upto 22% rate of major complications including infections, vascular and lung parenchyma injuries.[1] Also the length of stay in patients with tube thoracostomy is longer.[7]

Hence, in trauma patients with clinical signs of thoracic trauma, a high index of suspicion should be maintained for possibility of an occult pneumothorax even if chest radiograph appears normal. CT scan is the definitive investigation to identify occult pneumothorax. It can be managed by simple observation or a tube thoracostomy depending on patient's stability.

In trauma patients with subtle signs suggestive of a pneumothorax which is not evident on a chest radiograph, CT scan should be done to identify occult pneumothorax. Patient management can be either conservative or tube thoracostomy, depending on patient's stability.

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Uterine Arteriovenous Malformations after Suction Evacuation of Missed Miscarriage

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ABSTRACT

Uterine arteriovenous malformation (AVM) is an uncommon but life-threating source of bleeding. AVM is an abnormal connection between uterine arteries and veins. Patients typically present with vaginal bleeding following miscarriage (medical/surgical) or cesarean section. The treatment of choice depends on the symptoms, age, desire of fertility, localization and size of the lesion. Uterine artery embolization is the first choice in symptomatic patients of reproductive age group. We report a case of AVM presenting after dilation and evacuation with extensive lesion, which was successfully treated with bilateral uterine artery embolization.

Key Words: *Vaginal bleeding. Arteriovenous malformation. Uterine artery embolization.*

INTRODUCTION

Uterine arteriovenous malformation (AVM) is an uncommon but a life-threating condition, due to profuse or irregular bleeding from abnormal connection between artery and vein.[1] The incidence or prevalence of AVM is unknown. Only 100 cases have been reported since 1926.[2] The few available case reports reflect the rare nature of uterine AVM. AVM is uncommon in nulliparous women and has been reported in patient with age group ranging from 18-72 years.[3] The lesion can be congenital or acquired. Congenital AVMs are rare and arise from anomalous differentiation of primitive capillary plexus which results in abnormal connections between arteries and veins.[2] Acquired AVMs are caused by uterine trauma and instrumentation, such as, in dilatation and curettage, therapeutic miscarriage, trophoblastic disease, cesarean section, endometriosis or endometrial cancers.[2] Angiography has been proved as gold standard test for the diagnosis of AVM.[3] Recently, color Doppler ultrasound has been used for obtaining a reliable diagnosis. It showed reversal of flow and colour mosaic pattern.[4]

We report a case of AVM after dilatation and curettage in a primigravida which was successfully treated with bilateral uterine artery embolization.

CASE REPORT

A 21-year primipara was admitted through clinic with complain of heavy vaginal bleeding for the past two days. She had uterine curettage 7 weeks back due to missed miscarriage at 8 weeks of gestation. Over previous few weeks, she reported on-and-off vaginal bleeding episodes, for which, she visited clinic twice. Medical treatment, antifibrinolytic agents, and oral progesterone were given but she did not respond to treatment. Hence, she was advised admission for workup and further management. On admission, her general examination was normal except pale appearance. The pelvic examination showed no active vaginal bleeding, slightly enlarged uterus and no adnexal abnormality.

Laboratory evaluation revealed a hemoglobin of 7.9 g/dl with hematocrit (Hct) 25.3% and normal platelet counts of 351x109/L. Her serum beta human chorionic gonadotropin (βHCG) level was 2.7 miu/ml, which confirmed that she was non-pregnant. Transvaginal scan at the time of admission revealed the endometrial canal widened with heterogeneous debris and multiple cystic spaces with marked vascularity.

Doppler ultrasonography on the following day showed a highly vascular anterior uterine wall. The endometrio-myometrial interface was lost due to AVM (Figure 1). The patient was referred for consultation with an interventional radiologist and uterine artery embolization was planned and done on the same day. Two packs of blood were transfused before procedure. Bilateral uterine artery embolization was done with (355-500 um) polyvinyl alcohol (PVA) particles. The pre- and post-embolization images of patient are shown in (Figure 2). No post-procedural complication occurred. Patient was hemodynamically stable with slight spotting after the procedure.

The patient was discharged two days after the procedure. She returned for her follow-up on the 10th postoperative day when she had no symptoms and was vitally stable.

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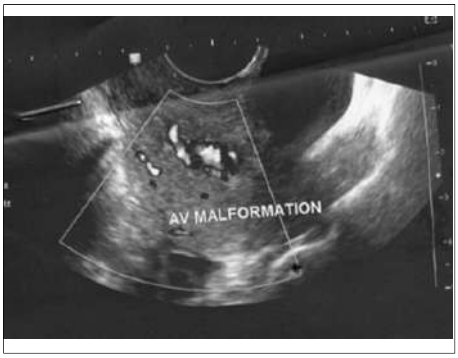

Figure 1: Colored Doppler ultrasound imaging feature of uterine arteriovenous malformation.

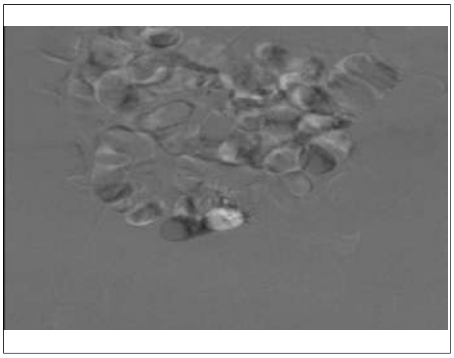

Figure 2 (a):): Angiogram of uterine artery showed an arteriovenous malformation (AVM).

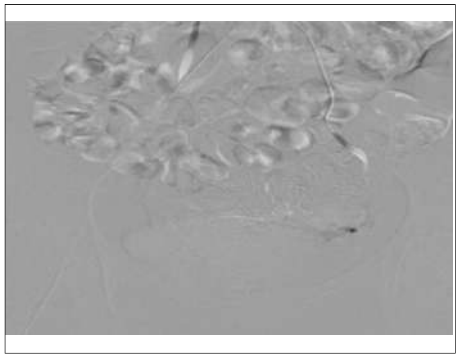

Figure 2 (b): Post-embolization showed complete embolization of the AVM.

DISCUSSION

In women of reproductive age group, the most common causes of abnormal uterine bleeding are complication related to pregnancy. Uterine AVM, although uncommon, should be considered in patients with unexplained uterine bleeding after abortion. Uterine AVM results in sudden and massive vaginal bleeding that may endanger life , suggestive of arterial hemorrhage, when the vessels are ruptured from iatrogenic sloughing of endometrium during dilation and curettage.[3]

Many imaging methods have been used to diagnose AVMs. These include ultrasound, computed tomography (CT), angiography and magnetic resonance imaging (MRI); although in recent practice, color Doppler ultrasonography is the modality of choice to diagnose AVM, as it increases the accuracy of ultrasound.[5]

Important differential diagnosis includes retained products of conception and gestational trophoblastic disease, because of the hypervascular appearance with turbulent flow. In such cases, the serum βHCG levels can help in confirming the diagnosis.[1]

Treatment of uterine AVM varies from expectant and medical management to surgical management. Medical management includes danazol or gonadotropin releasing hormone analogues in patients with mild hemorrhage.[6,7]

The treatment of patients with AVM depends upon the patient age, size and site of lesion and desire to retain fertility. The patient can be offered intervention options from minimally invasive uterine artery embolization to definitive surgical hysterectomy.[8]

Angiographic arterial embolization has now-a-days become the preferred management, because it is minimally invasive and preserves fertility. It is considered in cases where women experience recurrent or severe bleeding and become hemodynamically unstable.[9] The advantages of arterial embolization include ≥95% success rate, lower complication rate and avoidance of surgical risks.[3,10] The procedure-related side effects include low grade fever, infection or pelvic pain. The complications of uterine arterial embolization are negligible when performed by interventional radiologist.[3]

In the current case report, the AVM was diagnosed by color Doppler scan and successfully managed with uterine artery embolization.

In conclusion, uterine AVMs are uncommon, life-threatening clinical condition, which should be considered in patients with unexpected heavy and irregular vaginal bleeding after delivery or any surgical procedure involving the uterus.

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Fatal *Elizabethkingia Meningoseptica* Cholangitis Following Biliary Stent Placement

Khalid Ahmed[1], Syeda Atia Qudsia[2], Abdul Rehman[1] and Syed Hani Abidi[1]

ABSTRACT

Elizabethkingia (E.) meningoseptica is a ubiquitous gram-negative bacillus belonging to the genus *Chryseobacterium* and has been reported to cause nosocomial infections in both the immunocompromised and immunocompetent patients. E. meningoseptica can colonize the biliary tree after endoscopic procedures; and cholangitis, caused by this organism, is associated with a favorable prognosis. Here, we report a fatal case of cholangitis secondary to E. meningoseptica that developed following biliary stent placement. This case suggests that E. meningoseptica can be a cause of potentially fatal biliary tract infections in patients who undergo biliary tract endoscopic procedures. Clinicians must not disregard this organism as a contaminant (or colonizer) as a delay in diagnosis and treatment can lead to a fatal outcome, as seen in this case.

Key Words: *Elizabethkingia meningoseptica. Chryseobacterium meningosepticum. Cholangitis. Multiple antibacterial drug resistance. Biliary stent. Complication.*

INTRODUCTION

Elizabethkingia (E.) meningoseptica, formerly known as Chryseobacterium meningosepticum, is an oxidase-positive, catalase-positive, non-glucose fermenting, and gram-negative bacillus that belongs to the genus *Chryseobacterium*. Eponymously named after Elizabeth King who first described this bacterium in 1959, this ubiquitous organism has been traditionally considered as a contaminant of blood cultures.[1] Reports published over the past two decades have shown that this bacterium can cause infections in both immuno-compromised and immunocompetent patients.[2] *E. meningoseptica* has been recognized as a frequent colonizer of bile, especially following endoscopic biliary tract procedures. Although it rarely causes biliary tract infection, cholangitis due to *E. meningoseptica* is generally associated with a favourable prognosis.[3]

Here, we report the case of an elderly lady who developed cholangitis due to *E. meningoseptica* following biliary stent placement and died subsequently because of septic shock.

CASE REPORT

A 70-year female with past history of hypertension and ischemic heart disease underwent work-up for obstructive jaundice at our hospital. She was diagnosed to have unresectable, peri-ampullary carcinoma and underwent palliative biliary stenting. Shortly after being discharged from the hospital, she presented again to the emergency department with fever, abdominal pain, and shortness of breath. On physical examination, she was febrile, tachycardiac, tachypneic and icteric. She was maintaining 92% oxygen saturation on room air. Chest auscultation was notable for harsh vesicular breathing with bilateral fine crackles.

Laboratory investigations revealed a total leukocyte count of 25.24 × 10^9 cells/L (reference: 4.0-11.0 × 10^9 cells/L) with predominant neutrophilia (94.4%) and platelet count of 575 × 10^9 cells/L (reference: 150-400 × 10^9 cells/L). Results of liver function tests included alanine aminotransferase of 61 IU/L (reference: less than 36 IU/L), aspartate transaminase of 335 IU/L (reference: less than 40 IU/L), alkaline phosphatase of 238 IU/L (reference: 75-120 IU/L) and total bilirubin of 10.4 mg/dl (reference: 0.3-1 mg/dl) with direct and indirect bilirubin of 6.3 mg/dl and 4.1 mg/dl, respectively.

A plain chest radiograph was obtained, which revealed bilateral interstitial infiltrates. Ultrasonography of the abdomen was also performed, which revealed minimal dilatation of extra- and intra-hepatic biliary channels. Blood, urine, and sputum cultures were sent and the patient was started empirically on clarithromycin (500 mg 2×/d), metronidazole (500 mg 3×/d) and ceftriaxone (2000 mg 1×/d). However, the patient's condition progressively worsened and she began to develop worsening respiratory failure. Arterial blood gas obtained on supplemental oxygen revealed partial pressure of oxygen of 51 mm Hg (reference: 70-100 mm Hg), partial pressure of carbon dioxide of 28 mm Hg (reference: 36-40 mm Hg), and bicarbonate concentration of 18 mmol/L

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(reference: 22-28 mmol/L). Due to worsening tachypnea and respiratory failure, endotracheal intubation was performed and she was shifted to the intensive care unit. Subsequently, a central venous catheter was inserted through the right femoral vein.

After about 48 hours, the patient continued to have spiking fever and was started on imipenem/cilastatin (500 mg 4×/d) and vancomycin (1000 mg 2×/d). Preliminary reports of blood cultures revealed growth of *E. meningoseptica*, which was disregarded as a contaminant initially. However, the patient's condition did not improve and final reports of tracheal, blood, and urine cultures showed growth of >105 CFU/mL of *E. meningoseptica*. Urine culture also grew vancomycin-resistant *enterococcus fecalis*. *E. meningoseptica* was sensitive to piperacillin-tazobactam and vancomycin, while *enterococcus fecalis* was sensitive to linezolid.

In consultation with infectious disease specialists, antibiotic therapy with piperacillin-tazobactam (4450 mg 3×/d), vancomycin (1000 mg 2×/d) and linezolid (600 mg 2×/d) was instituted (on the fourth day of admission). However, the patient's condition continued to deteriorate and she developed multi-organ dysfunction. Her arterial serum lactate rose to 4.5 mmol/L (reference: 0.6-2.2 mmol/L) and she began to develop hypotension (mean arterial pressure of less than 65 mm Hg). An arterial catheter was inserted for invasive blood pressure monitoring and norepinephrine infusion (3 mcg/minute) was started. Despite vasopressor support, patient went into cardiac arrest and could not be revived.

DISCUSSION

This case emphasizes the importance of considering *E. meningoseptica* as a potential pathogen of the biliary tract infection. This is in contrast with previously published evidence which suggested that cholangitis with *E. meningoseptica* following endoscopic procedures is associated with a good prognosis. Most strains of *E. meningoseptica* are resistant to carbapenems (conferred by the metallo-β-lactamase, BlaB) and this is the only bacterium known to have two chromosomally-encoded metallo-β-lactamase genes.[4] This organism is peculiar in that it is often sensitive to piperacillin-tazobactam, despite being resistant to carbapenem; and vancomycin has good activity against this organism, even though it is a gram-negative bacillus.[5] Due to these unique antibiotic susceptibilities, clinicians must keep *E. meningoseptica* in mind as a potential pathogen in patients who develop biliary sepsis following endoscopic procedures. A delay in recognizing this pathogen or disregarding it as a contaminant could have disastrous consequences for patients, as happened in the present case.

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CASE REPORT

A Young Boy with Persistent Nodules and Hoarseness: A Rare Presentation of Nodular Secondary Syphilis

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ABSTRACT

Syphilis is a venereal disease caused by treponema pallidum, historically known as "great mimicker" because of its variable presentations. Secondary syphilis usually presents with maculopapular or papulosquamous rash. Rare manifestations include papulonodular, annular and lichenoid lesions. Nodules are usually found in benign tertiary syphilis. We present a case of a young boy who presented with five months' history of nodular skin lesions and hoarseness of voice. Venereal disease research laboratory (VDRL) was initially negative owing to prozone phenomenon. However, histopathology was confirmatory and he responded to benzathine penicillin. We present this case because of rare clinical presentation with persistent nodular lesions, unusual age group, and negative initial VDRL test.

Key Words: *Secondary syphilis. Nodular. Hoarseness.*

INTRODUCTION

Syphilis was historically termed as a "great mimicker" due to its varied and unusual presentations.[1] It is caused by a microaerophilic spirochete, treponema pallidum, a rod shaped motile organism, and is a sexually transmitted disease. Syphilis continues to be a worldwide problem; highest rates are reported in South and Southeast Asia, followed closely by sub-Saharan Africa.[2] It passes through four stages: primary, secondary, latent and tertiary. Primary and secondary stages are self-limiting and may go unrecognized. Primary lesion is a painless chancre which heals by itself. Secondary syphilis has protean cutaneous manifestations which include macular, maculopapular or papulosquamous rash, which is also self-limiting.[3] Unusual presentations include micropapular, nodular, annular and corymbose.[4] Due to diversity of clinical presentations, atypical presentations may cause diagnostic problems and delay in diagnosis with high risk of transmission.

We report a case of a young boy who presented to us with persistent nodular lesions and was diagnosed as secondary syphilis in histopathology.

CASE REPORT

A 15-year boy presented to us in outpatient department (OPD) with intensely pruritic nodular lesions over whole body for the last five months. He was treated with topical steroids and antibiotics without any response. He had hoarseness of voice for two weeks.

On examination, he had symmetrically distributed, infiltrated, erythematous to lichenoid nodules over volar aspect of wrists, limbs, trunk and face with some overlying scale. There was an annular plaque over face and split papules at angle of mouth (Figure 1). Oral cavity examination showed a snail track ulcer over hard palate. His palms showed keratoic papules. A serosangious discharge was observed from nasal mucosa, eyes showed conjunctival hyperemia. On genital examination, there was a healed scar over glans and condylomalata in perianal area. His cervical, axillary and inguinal lymph nodes were papable which were discrete, rubbery, shotty and non-tender. Epitrochlear lymph nodes were palpable. Initially, he denied any sexual contact history. On detailed history, he told that he had a genital ulcer six months back which healed after taking some topical application prescribed by a general practitioner.

Blood complete picture (CP), liver and renal function tests were normal. Human immunodeficiency virus (HIV)

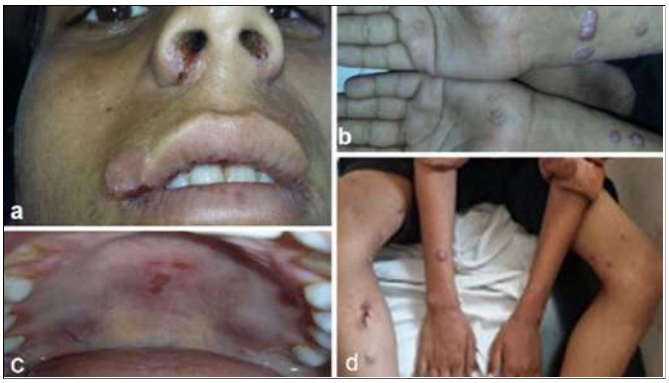

Figure 1: (a) Split papules, inflamed nasal mucosa, annular plaque near angle of mouth; **(b)** Multiple bilateral violaceous nodules over limbs with hyperkeratotic lesions over palms; **(c)** Mucous patch; **(d)** Multiple symmetrical erythematous nodules over limbs.

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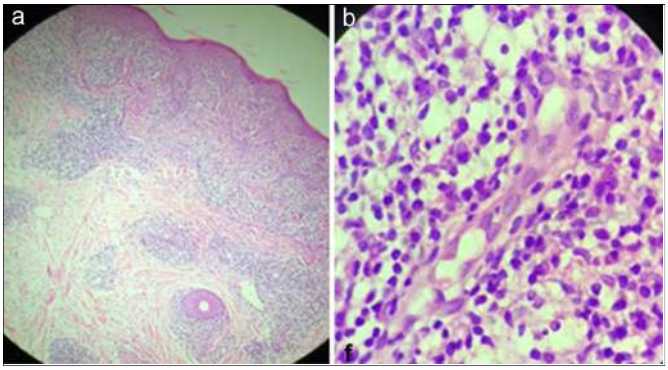

Figure 2: (a) Histopathology reveals perivascular infiltrate; **(b)** Infiltrate comprising of predominant plasma cells and endarteritis obliterans.

serology was negative. Veneral disease research laboratory (VDRL) test was positive in titre of 1:32 and treponema pallidum hemagylutination assay (TPHA) was also positive. Skin biopsy revealed hyperkeratosis and acanthosis, dermis showed dense mixed inflammatory infiltrate predominantly comprising of plasma cells. There was vascular proliferation and endothelial swelling (Figure 2). Report was consistent with nodular syphilis.

He was treated with intravenous benzyl penicillin 1.2 mU. Mucosal lesions as well as skin lesions showed improvement after first dose.

DISCUSSION

Syphilis has a myriad of presentations and was popularly known as "a great imposter" caused by a spiral spirochete, trepenema pallidum, which spreads by direct contact as organism is fragile and cannot survive in environment.[5] Blood-borne and transplacental transmission is also seen.[4] It usually passes through primary, secondary, latent, and tertiary stages. Primary stage is characterized by a chancre which heals with or without treatment and often goes unnoticed.[6] Secondary syphilis has variable and complex manifestations and may sometimes cause diagnostic difficulties. It is important to recognize the disease at this stage because it is highly infectious and is usually self-resolving. If left untreated, it may progress into tertiary syphilis after 5 to 10 years.[1]

After 3 -12 weeks of primary syphilis, secondary syphilis may appear as a rash. About 95% rashes are macular, maculopapular or papulosquamous. There are some rare manifestations like annular, lichenoid, nodular, corymbose and lues maligna.[6] Rare manifestations are usually found in HIV positive patients.[7] Nodular lesions are classically seen in tertiary syphilis and termed as benign nodular syphilis.[8] The presence of nodules and infiltrated plaques is extremely uncommon in secondary syphilis and was first described about 20 years ago. Only few case reports of papulonodular secondary syphilis are reported in literature since then. Such cases may be initially misdiagnosed as sarcoidosis, lymphoma, diffuse cutaneous leishmaniasis or lichen planus hypertrophicus, as in our case.[9] Secondary syphilitic lesions resolve by themselves in most of cases; but in our case, nodules and plaques were persistent for six months, which is rarely seen and causes diagnostic confusion. They may remain localised to face or other body area or may be generalised. Diagnosis is further complicated by variable histological findings of granulomas or lymphoid hyperplasia, which explains the clinical appearance of nodules and plaques. Serological tests may also be negative, owing to prozone phenomenon.[10]

Our patient was a young boy with persistent, symmetrically distributed, nodular skin eruption which was persistent for six months. He had pruritis, which is unusual in secondary syphilis. He was initially misdiagnosed as lichen planus and sarcoidosis. Initial VDRL test was also negative. Histopathology was diagnostic of secondary syphilis. Patient responded to injection benzathine penicillin 1.2 mU with the healing of lesions and improvement of hoarseness.

To conclude, nodular secondary syphilis should be kept in differential diagnosis even if patient does not belong to high risk group; as in our case, it can be managed at this stage to avoid complications of tertiary syphilis.

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CASE REPORT

Endobronchial Tuberculosis Simulating Carcinoid Tumor

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ABSTRACT

Diagnosing endobronchial tuberculosis (EBTB) can be difficult due to the lack of specific signs and symptoms that differentiate it from other respiratory diseases, such as lung tumors. We hereby report a case of a very rare presentation of tuberculosis (TB) in a patient who presented with a dry cough and significant weight loss for 3 months. Chest X-ray and CT scan of the chest showed partial atelectasis and a segmental collapse of the right upper lobe and tumor-like arising from its bronchus along with a large right para-tracheal mediastinal lymphadenopathy, mimicking a metastatic (N2) disease. Fiber-optic bronchoscopy revealed a fleshy, highly vascularized mass occluding the right upper lobe bronchus, and thus the initial diagnosis of carcinoid tumor was made. Mediastinoscopy and biopsy of these mediastinal lymph nodes showed caseating chronic granulomatous inflammation consistent with TB, which changed the diagnosis to EBTB. The patient was treated with first-line anti-tuberculous drugs that led to a full resolution in terms of symptoms, radiological findings and complete disappearance of the endobronchial mass by bronchoscopy. To the best of authors' knowledge, there are no other similar cases in presentation and management in the literature.

Key Words: *Endobronchial tuberculosis (EBTB). Fiber-optic bronchoscopy. Carcinoid tumor.*

INTRODUCTION

The incidence of endobronchial tuberculosis (EBTB) is about 10-15% of patients with active tuberculosis (TB).[1] EBTB most commonly involves the right upper lobe and right main bronchus. The X-ray picture usually shows consolidation or loss of volume.[2] However, presentation as a tumor-like mass is, even more, rarer,[1] and diagnosing this pathology can be difficult due to the unusual presentation, rarity, and the lack of specific signs and symptoms that differentiate it from other respiratory diseases, such as lung tumors or carcinoid. Combination of investigations is required to reach the definitive diagnosis.

CASE REPORT

A 48-year diabetic male who was a non-smoker presented with a dry cough and involuntary significant weight loss for 3 months. He had no other respiratory or constitutional symptoms. He was healthy with no history of major illness in the past. Physical examination was unremarkable. Chest X-ray showed infiltration in the right middle zone with evidence of partial (segmental) lung collapse (Figure 1A). CT scan chest showed partial atelectasis of the right upper lobe and a tumor-like mass arising from its bronchus causing partial occlusion of this bronchus (Figure 1B), with multiple enlarged mediastinal lymph nodes, mainly in stations R3 & R4, mimicking a metastatic (N2) disease, the largest lymph node measuring 4 x 3 cm (Figure 1C).

Fiber-optic bronchoscopy revealed a fleshy, very highly vascularized mass, that tended to bleed to touch at the entrance of the right upper lobe bronchus and was partially occluding the bronchus, highly suspicious of carcinoid tumor (Figure 2). Bronchoalveolar lavage (BAL) and brushing of this mass was performed, which was negative for malignant cells and the initial report for acid-fast bacilli (Ziehl-Neelsen staining) and BAL for polymerase chain reaction (PCR) were also negative. However, BAL samples were sent for TB culture and the results were pending.

Although the bronchoscopic appearance was strongly suspicious of carcinoid tumor, non-small-cell lung carcinoma (NSCLC) could not be excluded completely with this enlarged mediastinal lymphadenopathy. So the decision was made to sample these accessible nodes to rule out atypical form of carcinoid tumor, or to exclude metastatic N2 disease from NSCLC, rather than to biopsy the endobronchial tumor by rigid bronchoscopy and the potential complications of massive bleeding. In addition, an endobronchial ultrasound (EBUS) with needle aspiration was not feasible as the mediastinal lymph nodes were situated in stations R3 & R4, which were very difficult to access by EBUS.

The patient underwent mediastinoscopy and incisional biopsy of the right para-tracheal lymph nodes of station

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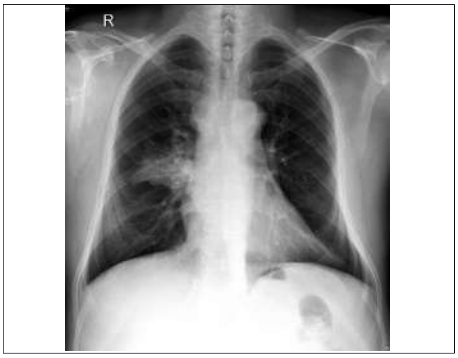

Figure 1A: Chest x-ray showing infiltration in the right middle zone with evidence of partial (segmental) lung collapse.

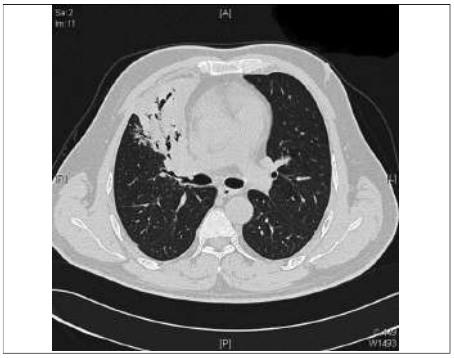

Figure 1B: CT scan chest showing partial atelectasis of the right upper lobe and a tumor-like projection arising from its bronchus casing partial occlusion of this bronchus.

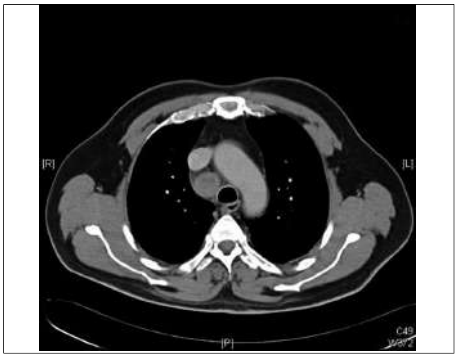

Figure 1C: CT scan chest showing multiple enlarged mediastinal lymph nodes, the largest lymph node, retro cava, measuring 4 x 3 cm.

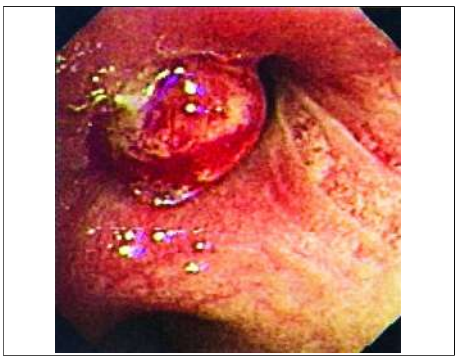

Figure 2: Fiber-optic bronchoscopy showing a fleshy highly vascularized mass at the entrance of the right upper lobe, highly suspicious of carcinoid tumor.

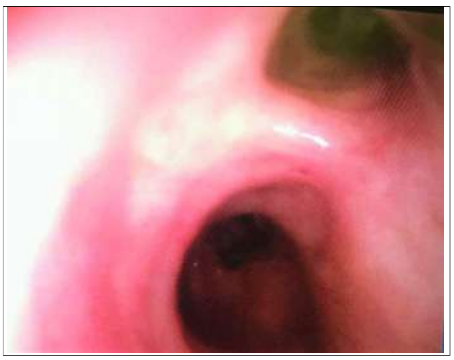

Figure 3: Fiber-optic bronchoscopy after 6 months showing complete resolution of the intra-bronchial lesion.

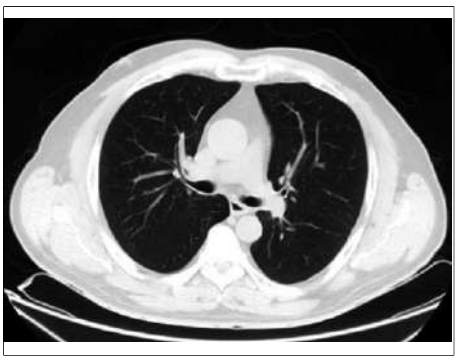

Figure 4: CT scan chest after 12 months showing complete resolution of the lung atelectasis and the tumor-like lesion of the right upper lobe.

R3, which showed chronic granulomatous inflammation with foci of caseous necrosis, with no evidence of malignancy, consistent with TB. Thus, the diagnosis was converted from the initial diagnosis of carcinoid tumor to the diagnosis of EBTB. This result was also confirmed later on by the positive TB culture of both BAL and the lymph node biopsy tissue.

The patient was treated with first line anti-tuberculous drugs for nearly 12 months, and there was marked improvement clinically and radiologically. The patient's symptoms were completely resolved. Fiber-optic bronchoscopy performed six months after initiating the treatment, which showed complete resolution of the lesion (Figure 3).

At six and 12 months follow-up, the patient remained well and asymptomatic and both radiological and bronchoscopy examinations did not show any evidence of recurrence. (Figure 4).

DISCUSSION

Diagnosis of EBTB in adults can be a major dilemma due to a wide variety of symptoms with lack of diagnostic clues in the investigations. It has been reported that EBTB can be confused with many different conditions like asthma and foreign body aspiration.[3,4] In addition, it can be confused with malignancies of an endobronchial tumor, such as in this case.[5] That, not only the findings are non-specific on initial investigations such as chest radiography, but also it is possible not to find any positive findings, and this may be misleading.[6] Until now, there is no single gold standard test in diagnosing EBTB and instead, a combination of investigations is required. This indicates the importance of having high index of suspicion when diagnosing EBTB.[6]

Bronchoscopic examination is considered one of the most important investigations in diagnosing EBTB.[2] Endoscopically, EBTB has been classified into seven sub-types: (i) actively caseating, (ii) edematous-hyperemic, (iii) fibrostenotic, (iv) tumorous, (v) granular, (vi) ulcerative, and (vii) nonspecific bronchitis.[7] However, in our case, the endobronchial lesion had the appearance of tumor-like carcinoid tumor due to high vascularity and easy bleeding rather than EBTB. Thus, the mass was not biopsied. For this reason, it is important to maintain a high index of suspicion and not to be misguided by the result of a single investigation modality.

The most helpful investigation in our case that guided us to the diagnosis of EBTB was the mediastinoscopy and biopsy of lymph nodes, which showed granulomatous inflammation with foci of caseous necrosis. An established modality and promising technique used for examining mediastinal lymph nodes is the endo-bronchial ultrasound (EBUS) with trans-bronchial needle

aspiration (TBNA). Geake *et al*. concluded that this method can be highly reliable for finding the cause of lymphadenopathy, especially when combing the microbiology and histopathology results.[8] It is worth mentioning that even histopathology may not be positive in nearly 16% of patients.[9]

Delay in diagnosis and treatment of EBTB may lead to devastating complications like bronchial stenosis, as was reported by Kizilbash.[10] Another complication that may occur is fistulization between two bronchi.[10] Both complications require further treatment that may include surgical intervention.

Thus, the true challenge lies in establishing an early diagnosis of EBTB in order to start appropriate treatment as soon as possible, and to prevent complications that may persist even after treatment. These complications were successfully prevented in our case due to prompt treatment after reaching the correct diagnosis of EBTB.

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Epigastric Heteropagus Conjoined Twins

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ABSTRACT

Heteropagus twins are an extremely rare form of asymmetrical conjoined monochorial, monoamniotic twins with an estimated incidence of less than one per one million live births. An often used synonym is parasitic twins. We report a very rare case of epigastric heteropagus twins with a large omphalocoele. The parasite had fully formed pelvis, lower limbs and upper limbs with male genitalia and it was attached to the autosite in the epigastrium.

Key Words: *Epigastric heteropagus. Asymmetrical twins. Omphalocoele.*

INTRODUCTION

Conjoined twins have expected frequency of 1 in 50,000 to 100,000 live births.[1] The term heteropagus conjoined twin was first used by Potter and Craig for asymmetrical conjoined twins.[2] Parasitic twins account for 1-2% of all conjoined twins. The dependent twin, known as parasite, is mostly under-developed, is attached to the more developed twin, called as autosite. Parasite, which is attached to the host epigastrium, is rare and called as epigastric heteropagus. The parasites are completely dependent on the hosts for their growth, mostly they are acardiac, anencephalic, rarely contain thoracic organs, usually demonstrate lower limbs, external genitalia, trunk and sometimes upper extremities with little or no movement in them. The host is usually a normal looking baby but may have associated anomalies, especially cardiac anomalies.[3]

In asymmetrical twins, the parasite is most commonly attached to the host at either hypogastric or suprapubic region, whereas attachment at epigastric region is rare with only 45 reports to date.[4] Surgical separation of epigastric twin is not difficult but the outcome is dependent on associated anomalies in the autosite.

We present a very rare case of heteropagus epigastric twins with a large omphalocoele. Surgical separation was planned but could not be performed due to death of twins, secondary to severe birth asphyxia sustained at birth.

CASE REPORT

A male baby was born at 39 weeks of gestation to a 22-year G2P1 by spontaneous vaginal delivery. The baby was delivered with great difficulty due to obstructed labor as the mother had irregular antenatal visits and was non-booked case, so fetal malformations were not detected antenatally. The family had one alive healthy girl with no history of twins in family.

After delivery, the baby was referred to our hospital for pediatric surgical consultation and further management. He was brought at 24 hours of life with severe respiratory distress. He was passing urine and stools normally. Weight was 4,100 g, length 46 cm and head circumference 35 cm. Physical examination showed an 8x10 cm omphaloceole and a parasitic twin attached to the epigastrium with two well-formed upper and lower limbs and male external genitalia with anal opening (Figures 1 and 2). There was total lack of movement in extremities of parasite. Hematological and biochemistry workup was within normal limits. X-ray showed developed bones in the lower limbs of the parasite. The host did not show any anomaly. Echocardiography showed complex congenital heart disease. Contrast enhanced CT scan of abdomen and chest showed dextrocardia in the host and a walled-off anterior abdominal wall defect was noted with segments III and IVb of liver, small bowel loops and omentum projecting into it, representing omphalocele. A parasite twin was seen attached to the right side of thorax of host twin. All limb bones of parasite twin were visualized. Thorax and abdomen of parasite twin were not visualized. Both

Figure 1: Epigastric heteropagus conjoined twins with a large omphalocoele.

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Figure 2: A parasitic twin attached to epigastrium.

kidneys were seen lying in the pelvis and were fused; and the ureters could not be visualized. Arterial supply of the parasite twin was coming from anterior intercostal arteries of the host twin. Venous drainage was through median umbilical vein draining into inferior vena cava.

Initial management included resuscitation, fluids, intravenous antibiotics and ventilatory support. Elective surgery was planned but baby went into sudden cardiac arrest, owing to the perinatal asphyxia sustained at birth, and could not be revived after resuscitation.

DISCUSSION

Heteropagus twins are extremely rare and represent only 1 to 2 percent of all conjoined twins.[3] In most of the reported cases, the host is a male. No clear risk factors have been mentioned in the literature so far.

The cause of conjoined twins and therefore of heteropagus twins is not fully clarified. A widely accepted theory suggests an incomplete fission of one zygote, which occurs about 14 days after fertilization. Logroño *et al*. suggest the alternate hypothesis that heteropagus twins originate from the fusion of two separate zygotes at a later stage. In a similar way, Ratan *et al*. mention a fusion theory, also called Ratan's theory. It is postulated that an unbalanced distribution of the placental blood leads to an ischemic insult in the parasite twin with selective atrophy of this twin as a consequence.[4,5]

The limbs of parasite normally do not show any spontaneous movements. This is due to absence of neural innervations of the parasite myoblasts, which leads to incomplete differentiation and consecutively to skeletal muscular atrophy. Therefore, on histology the parasite limb has only fat and bones but no muscle tissue.[6,7]

The host twin often has various malformations, the most common being congenital heart defects, especially ventricular septal defects.[5] There is also a high incidence of associated omphalocele, especially in epigastric heteropagus twins. Spinal cord defects are less commonly seen; whereas, bony and visceral connections between the heteropagus twins can occur but are rarely observed.[4,6]

Due to the rarity of this malformation, there is only limited literature available, mainly consisting of case reports. Omphalocele was present in approximately half of epigastric heteropagus, but major omphalocele was seen only in a few cases.[8] This case was unique for the presence of major ompahalocele, which has been very rarely reported so far.

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CASE REPORT

Ellis-Van Creveld Syndrome in a Neonate

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ABSTRACT

Ellis-van Creveld syndrome is a rare form of skeletal and chondroectodermal dysplasia which affects all the three ectodermal, mesodermal, and endodermal derivatives. It has an autosomal recessive inheritance. This is caused by mutations in 1 of 2 genes, EVC 1 or EVC 2. This syndrome has a constellation of characteristic features that include bilateral post-axial polydactyly, mainly involving the upper limbs, hypoplastic nails and teeth, congenital heart defects, and chondroectodermal dysplasia. It is mainly a disorder of Amish population where incidence of this disease is 1/5000 and its incidence in non-Amish population is 7/1000000. Our patient had all the major characteristic features consistent with Ellis-van Creveld syndrome including post-axial polydactyly, teeth and nail abnormalities, congenital heart defect and skeletal dysplasia. Until now, only five cases have been reported from this region of the world, none of them diagnosed in neonatal life and having characteristic common atrium.

Key Words: *Ellis-van creveld syndrome. Chondroectodermal dyspasia. Neonate. Common atrium.*

INTRODUCTION

Ellis-van Creveld syndrome is a rare form of skeletal and chondroectodermal dysplasia with an autosomal recessive inheritance, first described in 1940.[1] It is caused by mutation in the EVC gene as well as by a mutation in a nonhomologous gene, EVC2, located close to EVC gene in a head-to-head configuration.[2] This syndrome has a constellation of characteristic features that include bilateral post-axial polydactyly, hypoplastic nails and teeth, congenital heart defects, and chondroectodermal dysplasia.[3] The birth prevelance in Amish population is 1/5000 and non-Amish population is 7/1000000.[4] Previously literature research shows that only five cases have been reported from this region, and none diagnosed in the neonatal period.

CASE REPORT

A baby girl born through elective LSCS at full term with Apgar scores of 7 at 1-minute and 8 at 5-minute and a birth weight of 3.3 Kgs presented with the complaints of breathing difficulty soon after the birth. She was the product of consanguineous marriage and was fourth in sibship. Mother was gravida 5 and history was negative for any chronic illnesses. Family history was significant for death of one sibling in neonatal life due to breathing difficulty and skeletal malformations. Record of sequence of events and cause of death for that sibling was not available. Baby was in obvious respiratory distress at presentation and was managed with bubble CPAP.

General physical examination revealed a total length of 41 cm at birth. Head circumference was 32 cm. Examination of oral cavity showed mandibular natal tooth (Figure 1) and extra-oral examination showed V-notch of middle part of the upper lip and fusion of upper lip and gingival mucosa (Figure 2).

Thoracic cavity was narrow and air entry was decreased in bilateral lung fields. Examination of limbs showed shortening of both upper and lower limbs with upper to lower segment ratio of 2 at birth. Hands were sausage shaped and nails were hypoplastic. There was post-axial polydactyly on both the hands on the ulnar side (Figure 3).

Skeletal survey of the patient showed shortened long bones and polydactyly that involved the bones. Polydactyly was post-axial involving the ulnar side of both hands. Parental heights were at the 50th percentile for father and at 25th percentile for mother. Parents examination of limbs and hands was normal. Examination of cardiovascular system of the baby showed a pansystolic murmur at left lower sternal border. Echocardiography was done that showed common atrium, ventricular septal defect (VSD), and pulmonary hypertension. Cardiologist opinion was taken regarding the cardiac malformations, and therapy was started for the pulmonary hypertension. Cardiologist advised a follow-up at two weeks of life. Septic screening was negative and blood counts were normal. Ultrasound abdomen and kidney, ureters, and bladder was done that was normal. Ultrasound brain was done that was also normal.

Patient gradually recovered and was discharged on the seventh day of life with the advice of an early follow-up. Parents were counselled regarding the nature of disease and requirement of a regular follow-up.

DISCUSSION

Ellis-van Creveld syndrome, also known as chondro-ectodermal dysplasia, is a rare autosomal recessive disorder that has a prevelance of 1/5000 live births in Amish population of America and the estimated

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Figure 1: Natal tooth.

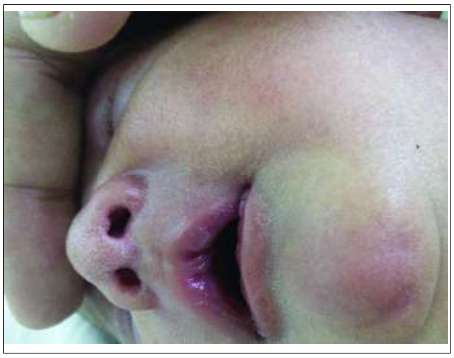

Figure 2: Gingival mucosa.

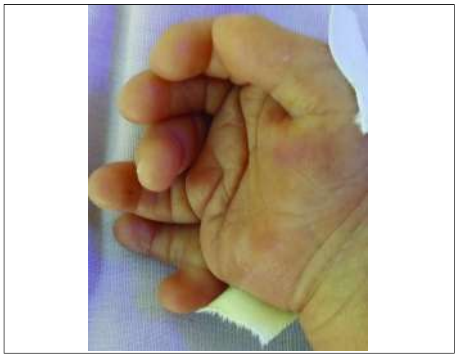

Figure 3: Post-axial polydactyly.

prevelance in non-Amish population is 7/1000000. It involves males and females equally. Autosomal recessive disorder affect 25% of next generation, 50% are carriers and 25% are normal. This patient was the product of consanguineous marriage and there was a history of death of one sibling with similar features.

This syndrome has a constellation of characteristic features that include bilateral post-axial polydactyly, hypoplastic nails and teeth, congenital heart defects and chondroectodermal dysplasia.[3] The polydactyly mostly involves the ulnar side of the upper limbs, and lower limbs are involved in 10% of cases. The polydactyly can be due to bifid metacarpal with duplication of phalanges and intercarpal fusion between hamate and capitate or it may just involve the soft tissue.[5] This patient has polydactyly involving the ulnar sides of both the upper limbs. Nails are hypoplastic and dystrophic. There may be complete absence of nails..Tooth involvement may include neonatal tooth, as present in our case, or partial adontia, small teeth and delayed eruption. Hair may occasionally be sparse.

Cardiac anamolies are a major cause of shortened life expectancy. It is reported in 50-60% of cases.[6] The most commonly reported cardiac defect is a common atrium.[7] Other reported defects include mitral and tricuspid valve defects, patent ductus arteriosus, ventricular septal defect, atrial septal defect and hypoplastic left heart syndrome. This patient also had common atrium, ventricular septal defect and pulmonary hypertension.

Chondrodystrophy in this syndrome is characterized by acromesomelic dwarfism involving the middle and distal parts of the upper limbs resulting in short stature. There is progressive distal limb shortening, symmetrically affecting the forearms and lower legs. Adult height ranges from 119-161 cm. Other features that may be present include genu valgum, curvature of humerus, talipes equinovarus, talipes calcaneovalgus and pectus carinatum with a long narrow chest. Genitourinary system may also be involved and manifestations include renal agenesis, nephrocalcinosis, megaureters, hypospadias, epispadias, vulvar atresia and cryptorchidism.

The cognitive and motor development may be normal with occasional CNS anomalies. Prenatal diagnosis can be made with intrauterine growth retardation, skeletal malformations and cardiac defects on ultrasound images. Definitive diagnosis is possible by sequencing EVC and EVC2, that is identified in two-thirds of patients with Ellis-van Creveld syndrome.[8] Due to lack of availability of genetic studies, the diagnosis was made on the basis of clinical features and additional tests, such as skeletal survey and echocardiography.

A multidisciplinary approach that involves a paediatrician, cardiologist, orthopedist, dentist, psychologist and clinical geneticist is required for the management of patients. Approximately half of patients die in early infancy as a consequence of cardiorespiratory problems. End organ involvement may include renal involvement (nephrotic syndrome, nephrolithiasis and renal failure), hepatic involvement (progressive fibrosis and hepatic failure) and hematological abnormalities that range from myelodysplastic changes with dyserythropoiesis to acute leukemia.[9]

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MELAS: A Complex and Challenging Diagnosis

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ABSTRACT

Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) is a rare multisystem disorder and is the most common maternally inherited mitochondrial disease. This condition has a special predilection for the nervous system and muscles. Typical findings on brain imaging include stroke-like areas, calcification of basal ganglia and brain atrophy. This accounts for the disease being, both clinically and radiologically, mistaken for ischemic stroke. The differentiation features from stroke include comparatively young age of the patients, site of the lesions, and relative overlap between the cerebral vasculature territories. In this case report, we discuss a 16-year male with clinical and radiological features highly suggestive of MELAS syndrome. Since this disease is rare and its clinical presentation is complex, it is among the most challenging to diagnose. It is particularly difficult to differentiate between ischemic stroke and MELAS. Magnetic resonance imaging (MRI) with diffusion weighted imaging (DWI), susceptibility weighted imaging (SWI) sequences and MR spectroscopy may aid in establishing the diagnosis. A particularly characteristic feature of MELAS syndrome is that recurrence may occur in locations different than previously noted, which was also seen in our patient.

Key Words: *MELAS. Stroke-like lesions. MRI. Mitochondrial disease.*

INTRODUCTION

Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) is the most common maternally inherited mitochondrial disease with multisystem involvement and a special predilection for the nervous system and muscles. Up to 80% of cases show a common defect located at nucleotide 3243 of the mitochondrial DNA (mtDNA) gene owing to a point mutation involving transposition from A to G, which encodes for transfer RNA (tRNA) for leucine.[1]

Abnormalities involving tRNA affect multiple parts of the respiratory chain. These are hypothesized to cause abnormal protein production throughout the mitochondria. Therefore, the body shifts its functions to derive energy primarily from anaerobic metabolism. This manifests as accumulation of lactic acid, depletion of Nicotinamide Adenine Dinucleotide (NAD) stores causing predisposition to hypoxic injury, resulting in death of neurons in the cortex. This is believed to be the basic pathogenesis of the disease.[2,3]

CASE REPORT

A 16-year male patient was initially admitted to our hospital because of non-specific abdominal pain, headache, nausea, vomiting and focal seizures. His initial neurological examination was normal. A study of cerebrospinal fluid (CSF) samples excluded neuro-infection. Electroencephalogram (EEG) was normal. Computed tomography (CT) scan showed areas of hypoattenuation in bilateral cerebellar hemispheres, raising the suspicion of ischemic changes (Figure 1A). MRI was performed, which revealed areas of high signal on DWI and no significant dropout on Apparent Diffusion Coefficient (ADC) mapping in the cerebellar hemispheres (Figures 1B and C). The patient was managed on the basis of subacute cerebellar infarcts.

Subsequently, a year later, the patient presented to the Emergency Department with seizures, headache and vomiting. This time the neurological exammation revealed a right-sided visual field defect and decreased visual acuity. EEG showed left temporo-occipital sharp and slow waves with diffuse theta and delta slowing. Extended diagnostic tests were performed. Thyroid antibodies and autoimmune workups were negative. CSF studies revealed a leukocyte count of 2 and an increased lactate level of 4.8 mmol/L. Herpes simplex polymerase chain reaction (PCR) and gene xpert for mycobacterium tuberculosis were negative. Homocysteine levels were normal and workup for porphyria was negative. Left deltoid muscle biopsy showed no evidence of degenerative, regenerative changes, atrophy or inflammation or ragged-red fibers.

MRI was done which showed gyral thickening of the left parieto-occipital cortex extending to the temporal lobe and areas of abnormal, high signal on FLAIR and T2 images with relative loss of cortico-subcortical differentiation, and reduction of signal intensity in these areas on T1 images representing gyral edema (Figures 2A and B) and (Figures 3B and C). This time, there was diffusion restriction in these lesions on DWI and

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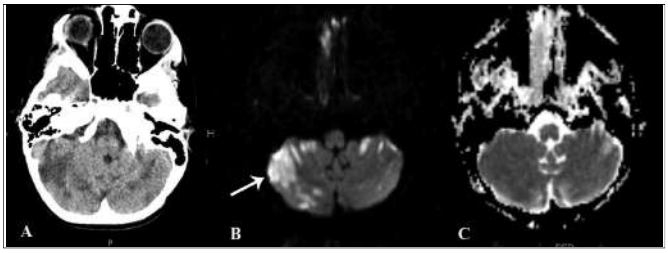

Figure 1: (A) Initial CT scan revealed hypoattenuating areas in bilateral cerebellar hemispheres which were suggestive of ischemic infarcts (B) MRI showed high signal in the cerebellar hemispheres on DWI (C) (white arrow) without any significant dropout on ADC maps.

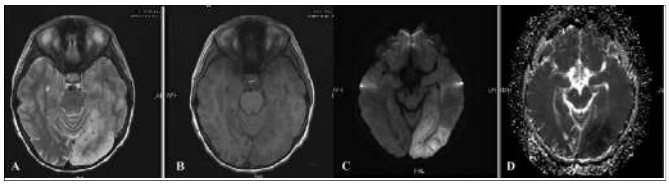

Figure 2: T2, T1 images of MR brain done a year later revealed signal abnormality in the left parieto-occipital lobe along with diffusion restriction in this area on DWI and dropout on ADC mapping.

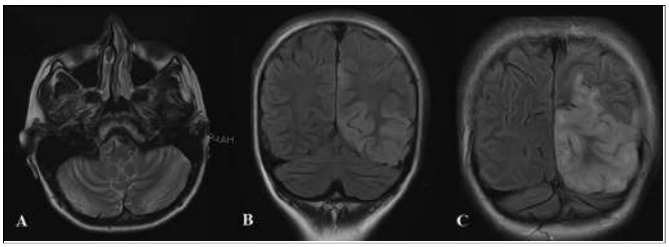

Figure 3: (A and B) Previously noted stroke like lesions in the cerebellar hemispheres had almost completely resolved on this examination (B and C) FLAIR images revealing gyriform edema and subcortical involvement in the left parietal and occipital lobes.

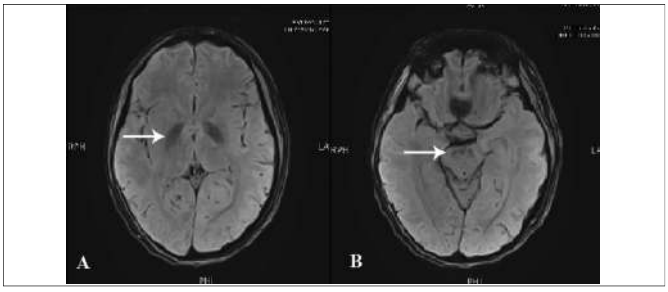

Figure 3: SWI images showing signal dropout in bilateral globus pallidus, substantia nigra and red nuclei (white arrows).

corresponding decrease in cortical signal intensity on ADC maps (Figures 2C and D).

The previously noted stroke-like areas in bilateral cerebellar hemispheres had, however, resolved (Figures 3A and B). Increased signals were noted on susceptibility weighted imaging (SWI) in bilateral globus pallidus, red nuclei and substantia nigra, which were likely representing iron deposition/calcifications (Figure 4). The whole clinical scenario, investigations carried out during hospitalization, information from history-taking, and CT and MR images of the brain with recurrent, migratory stroke-like episodes were all highly suggestive of MELAS syndrome.

DISCUSSION

MELAS is a rare multisystem disorder and is the most common maternally inherited mitochondrial disease having a special predilection for the nervous system and muscles. Presentation of patients with this mitochondrial disorder is usually delayed until childhood after a period of normal development, owing to time taken for metabolite build-up and exhaustion of cell's coping mechanism. Most patients, before the age of 40, have history of multiple stroke-like episodes. These recurrent stroke-like episodes progressively cause widespread damage leading to loss of cognitive mechanism. Characteristic signs and symptoms may include muscle weakness, recurrent headache, weakness and even, vision loss. Patients develop increased anion gap metabolic acidosis from lactic acid buildup that presents as vomiting, abdominal pain, loss of bowel control, fatigue, muscle weakness and breathing difficulty. MELAS syndrome may also present with ataxia and myoclonus as well as endocrine system abnormalities like diabetes mellitus, hypoparathyroidism and hypogonadism.[4]

The stroke-like areas are non-vascular and linked to mitochondrial mutation leading to oxidative phosphorylation dysfunction, which subsequently causes encephalopathy. The combination of lactic acidosis, multiple nonvascular strokes, encephalopathic psychosis, diabetes, and sensory neuronal hearing loss cause severe dysfunction leading to increased mental disabilities, physical disabilities, and eventually death. Death may also occur due to cardiac failure, pulmonary embolism, or renal failure.

Typical findings on brain imaging include stroke-like areas, calcification of basal ganglia, and diffuse brain atrophy.[5] In 90% of patients with MELAS, the focal neurological symptoms are well correlated with stroke-like lesions in the corresponding brain areas.[6] Hypodense areas resembling infarcts are seen on CT imaging. This accounts for the disease being, both clinically and radiologically, mistaken for ischemic stroke initially. The differentiation features from stroke include comparatively young age of the patients, site of the lesions (parietal, temporal and occipital cortices are especially prone), and relative overlap between the cerebral vasculature territories.[7]

The stoke-like areas in MR studies manifest as enlarged gyri with enhanced T2 signals. Subcortical white matter involvement is not unusual. Previous reports have suggested that DWI and ADC sequences prove extremely helpful in distinguishing between stroke-like lesions of MELAS and ischemic stroke. The stroke-like lesions in MELAS syndrome, show increased signal on

DWI (T2 shine through) with minimal or no change on ADC because these lesions are probably due to vasogenic edema rather than cytotoxic edema, as seen in ischemic stroke. Cytotoxic edema causes diffusion restriction with corresponding dropout on ADC mapping.[7,8] This finding was observed in our patient on the initial MRI, revealing the cerebellar stroke-like lesions. They did not show any significant dropout on ADC mapping. However, dropout on ADC was observed in our case in the subsequent MRI with lesions in the parieto-occipital lobe, suggesting that this feature is not always characteristic. Kim *et al*. reported that increased, normal and decreased ADC values were found in an equal number of patients with MELAS, suggesting that cytotoxic edema gradually evolves following an acute stroke-like episode in such patients, and this may overlap with hyper-perfusion and vasogenic edema.[9]

Lesions in the subacute phase may sometimes show contrast enhancement due to increased permeability resulting from congestion or reperfusion or as a result of blood-brain barrier damage.[5,9] Imminent improvement is seen in the clinical picture as these stroke-like lesions subside with passing time. A particularly characteristic feature of MELAS syndrome is that recurrence may occur in locations different from previously noted, which was also seen in our patient.[5,10,11] MR spectroscopy is an additional tool that may reveal elevated lactate peaks, suggesting disturbed anaerobic processes.[7,8]

The differential diagnoses of MELAS syndrome, besides ischemic stroke, includes vasculitides bearing resemblance to Moyamoya and Kawasaki diseases, viral encephalitis, status epilepticus, Creutzfeldt-Jakob disease, Wilson's disease, hypoxia, and other mitochondrial disorders such as Leigh's disease, Kearns-Sayre Syndrome, and myoclonic epilepsy with ragged-red fibers.[5] This condition is usually associated with a poor prognosis and outcome, as there is no effective treatment for MELAS syndrome established, hence the resultant high morbidity and mortality rates.

In summary, we report a patient with clinical and radiological picture of MELAS syndrome. Since this disease is rare and its clinical presentation is complex, it is among the most challenging to diagnose. It is particularly difficult to differentiate between ischemic stroke and MELAS syndrome. MRI and MR spectroscopy may aid in establishing the diagnosis; however, the radiological features are often non-specific and may overlap. It is also important to consider the other differential diagnoses.

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CASE REPORT

A Rare Case: Rupture of Internal Pudendal and Uterine Artery in a Vaginal Delivery

Novera G. Chughtai and Raheela Mohsin Rizvi

ABSTRACT

The management of puerperal hematomas after normal delivery has always been challenging for obstetricians. Vulvar, vulvovaginal, or paravaginal hematomas are common. On the other hand, retroperitoneal hematomas are uncommon and can be life-threatening. The diagnosis of vascular injury is rarely made preoperatively as atonic or traumatic postpartum hemorrhage (PPH), uterine rupture and amniotic fluid embolism are more common differential diagnoses. Injury to internal pudendal and uterine vessels is extremely rare in cases of vaginal delivery and, therefore, the literature on this topic is very scarce. We present a rare case of both internal pudendal and uterine artery rupture in a normal vaginal delivery, which led to massive postpartum hemorrhage. The diagnosis was made on Magnetic Resonance imaging (MRI) and arterial embolization was performed. This case stresses on the need for careful post-delivery monitoring for revealed postpartum hemorrhage. Vascular injury is a rare life-threatening cause of obstetric shock, and active; and timely operative intervention can prevent morbidity and mortality.

Key Words: *Postpartum hemorrhage. Maternal mortality. Hematoma. Internal pudendal artery rupture. Uterine artery rupture. Vaginal delivery. Complication.*

INTRODUCTION

Massive postpartum hemorrhage is one of the most feared complications of vaginal delivery. Uterine atony, retained placenta, and vaginal and cervical tears are the common causes. Hematomas due to arterial rupture are rare and their insidious nature may delay the diagnosis and cause severe hemorrhagic shock and even death.[1]

During pregnancy and postpartum period, rupture of uterine vessels is a rare life-threatening condition. The risk of maternal and perinatal mortality is reported as high as 40 and 30%, respectively.[2] Spontaneous rupture of uterine vessels has been reported in literature, more frequently in the antenatal and less commonly in the postpartum period. The broad ligament is the most common site being involved in 75% of cases.[3]

In cases of massive postpartum hemorrhage, when mainstay methods of suture and packing fail, arterial embolization becomes an excellent alternative to definitive treatment, laparotomy.[4]

This case report describes the above rare complication in a vaginal delivery of a primipara.

CASE REPORT

A 22-year primipara was referred to our hospital emergency services with complaints of heavy bleeding immediately post normal vaginal delivery of a 5 lbs baby two hours back. On admission, she was tachycardiac and hypotensive. Per abdominal examination revealed no abnormality. On pelvic examination, she was found to have a right sided vulval hematoma, sized approximately 8x10 cm with massive vaginal bleeding. Hematological investigations showed severe anemia with a hemoglobin of 3 gm/dl and deranged coagulation profile. She was immediately taken for examination under anesthesia (EUA) and to proceed accordingly. EUA revealed a right sided vulvo-vaginal hematoma, which was drained; active bleeding was also seen from right lateral cervical tear. Hemostasis was secured by repair of cervical tears, few sutures were applied at hematoma base and vaginal packing. Patient was given packed cells and blood products to prevent disseminated intravascular coagulation (DIC).

During her postoperative recovery, she was found to be hemodynamically stable; but despite multiple transfusions, her hemoglobin remained below 8 gm%. An initial diagnosis of DIC was made and conservative treatment given; but in view of her deteriorating condition with active vaginal bleeding and recurrence of lateral vaginal wall hematoma of 10x10 cm, internal hemorrhage from uterine/pudendal artery was thought of. MRI angiogram was planned, which showed active breach in the right uterine artery and the right internal pudendal artery and a large vulvo vaginal hematoma (Figure 1).

The case was discussed with radiology interventionist and an angio-embolization was performed. The vaginal pack was retained for 24 hours and the patient was kept in the special care ward. She remained well and was

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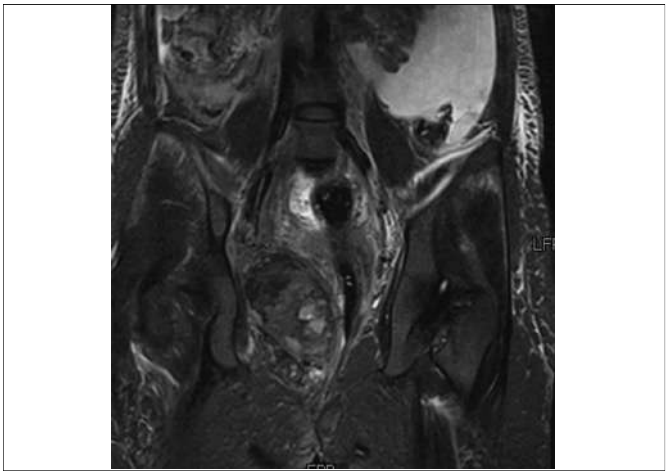

Figure 1: Coronal MRI image of the vulvo-vaginal hematoma.

discharged on the sixth postoperative day. Her subsequent visits in gynecology clinics showed smooth and uneventful recovery with resumption of normal menstrual cycle.

DISCUSSION

Many cases of puerperal hematomas, secondary to uterine artery rupture, have been described in literature, both in pregnancy and postpartum period; but there has been no report of both the uterine and internal pudendal arterial rupture in normal delivery to date.

Puerperal hematoma is an uncommon complication of childbirth with a potential for serious morbidity and possible mortality. Their incidence is 1 or 2/1,000 deliveries. Puerperal hematomas, however, are not unavoidable.[5]

The initial conservative treatment options in the management of vulvo-vaginal hematomas include suturing and vaginal packing. This is similar to what was done in this case which was initially managed conservatively by examination under anesthesia and vaginal packing. Serial complete blood count and coagulation profile were performed, which showed a continuous drop in hemoglobin and warranted further investigation including MRI. Guerririo *et al*. describe the sonographic follow-up of vulvo-vaginal hematoma with MRI correlation. The authors concluded that MRI provides a detailed mapping of the lesion and excludes retroperitoneal involvement.[6]

Case series on the management of retroperitoneal hematomas after vaginal delivery have reported various methods, including conservative approach and surgical interventions, such as laparotomy and evacuation of the hematoma, pelvic arterial embolization,[7] and even hysterectomy.[1]

Embolization of the bleeding artery can be considered when surgical interventions cannot guarantee success. The bleeding artery can be localised through digital subtraction angiography.[8] An interventional radiologist can then embolize the bleeding vessel supplying the site.[8] Pelvic arterial embolization is a good option in the management of puerperal hematomas, if the patient is hemodynamically stable and the necessary equipment and staff are available. If both of these conditions are not met, as in this case, then laparotomy is indicated.[9]

Ligation of the internal iliac artery has been successfully used in postpartum hemorrhages for more than five decades. It provides a reduction of 85% in pulse pressure and a 50% reduction in blood flow in the distal vessels, including uterine artery, internal pudendal artery and middle rectal artery.[10] Internal iliac artery ligation is an effective technique that requires surgical skill.

Rupture of internal pudendal and uterine artery in a vaginal delivery is a rare diagnosis and this is the first case report in this context. In cases of massive postpartum hemorrhage refractory to conservative measures, and facilities available, then MRI and pelvic angiogram are useful diagnostic tools for diagnosis of arterial rupture; though expensive, but timely angio-embolization can prevent morbidity and loss of a precious life.

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CASE REPORT

Hydatid Cyst of Skeletal Muscle Presenting as Soft Tissue Tumour

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ABSTRACT

Hydatid disease is helminthic infection caused by tape worm *echinococcus granulosis*. It commonly involves liver (65-75%) and the lungs (25 to 30%). Involvement of bones and skeletal muscle is very rare, i.e. 3 to 5%. This is the report of a 21-year girl, city resident, presented with mass involving left proximal thigh. The mass was painless and had gradually increased in size over three years. MRI revealed solid cum cystic lesion in vastus medialis muscle. Tru-Cut needle biopsy of the lesion revealed necrotic material. Intraoperative findings were of different sized cystic lesions, typical of hydatid cyst. The cysts were removed intact. No recurrence was seen in 11-month follow-up. Hydatid cyst involving skeletal muscle is very rare entity, but should be considered while making differential diagnosis of soft tissue mass.

Key Words: *Hydatid cyst. Skeletal muscle. Soft tissue tumour. Echinococcosis.*

INTRODUCTION

Hydatid disease or echinococcosis is helminthic infection which is most commonly caused by the tape worm *echinococcus granulosus*. Liver (65-75%) and lungs (25-30%) are involved in most cases. Hydatid cyst of the musculoskeletal system represents approximately 3% of all hydatid disease cases.[1] Even in endemic area, hydatid cyst involving skeletal muscle is rare (0.7-0.9%).[2] Hydatid cyst grows slowly in a spherical manner. A pericyst (fibrous capsule), derived from host tissue, develops around it. The cyst wall consists of two layers, ectocyst (outer) and the endocyst (germinal layer). Brood capsules are small, intracystic cellular masses in which future worm heads develop into *scoleces*. Free brood capsules and *scoleces* found in the hydatid fluid constitute the hydatid sand. Hydatid cyst involving skeletal muscles presents as slowly growing painless mass. However, in some cases the cyst is complicated by nerve compression or infection, so that it mimics an abscess or a malignant tumor.[3] Since its clinical features are closely related to those of soft-tissue tumor, therefore making preoperative radiologic diagnosis of hydatid cyst is very important in order to avoid biopsy. Serology and MRI may not be helpful in diagnosing primary skeletal muscle hydatid cyst in all cases. A negative serological test does not rule out the diagnosis of *echinococcosis*. However, it is considered as the gold standard investigation for the disease. Skeletal muscle hydatid cyst should be included in the differential diagnosis of soft tissue masses involving limbs, especially in areas where it is endemic. The mainstay of treatment is to excise the cyst followed by treatment with antihelminthics.

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CASE REPORT

A 21-year girl, urban resident, with no known comorbid condition, presented with history of painless mass over left proximal thigh just below inguinal ligament for the last three years. It had gradually increased in size. There was no history of weight loss or contact with domestic livestock. On examination, a 7 x 10 cm ill-defined mass was palpable over anteromedial aspect of left upper thigh extending up to inguinal region. It was smooth, non-tender, firm in consistency, not attached to the overlying skin which was normal in colour. It was adherent to the muscles. Left inguinal lymph nodes were palpable. Abdomen and chest examination were normal. Baseline investigations were normal. X-ray left thigh was unremarkable. MRI left thigh revealed cystic cum solid, multi-septated leasion in vastus medialis muscle, 17.4 x 5.6 x 5.2 cm in size, medially abutting the femoral vessels and Sartorius (Figures 1a and 1b). Ultrasound guided core biopsy of the lesion revealed necrotic material.

Patient underwent exploration of the mass. Longitudinal incision made over anteromedial aspect of upper thigh. Underlying great sephanous vein was ligated along with venous engorgement around. Muscles were dissected and a cystic lesion was identified just behind the vastus medialis muscle, separated from the adjacent structure with blunt dissection. Multiple daughter cysts and a thick capsule was identified (Figures 2a and 2b). Diagnosis of hydatid cyst involving skeletal muscle was made on

gross inspection. Parent and the daughter cysts were removed intact carefully to prevent rupture. Cavity washed with hydrogen peroxide and copious amount of normal saline and then closed. The postoperative course was favourable characterized by clinical improvement and patient was started on albendazole. At 11-month follow-up, there was no clinical and radiological (MRI) evidence of recurrence.

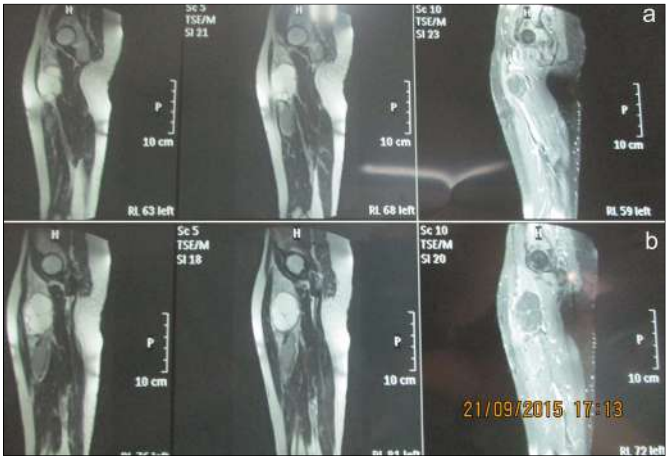

Figure 1: MRI appearance of the lesion: (a) axial view, (b) sagittal view.

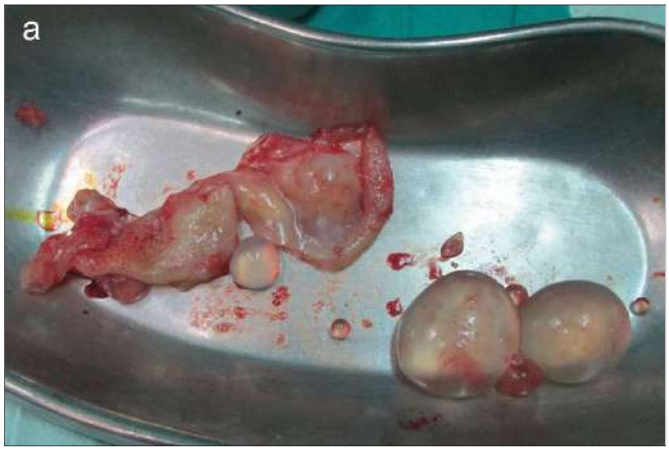

Figure 2a: Gross appearance of the dissected lesion.

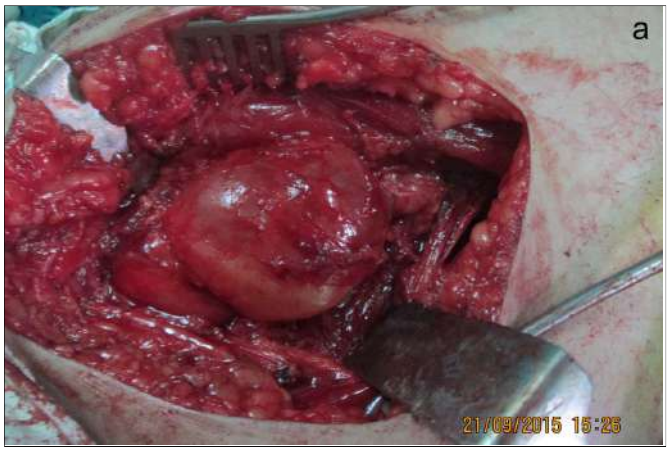

Figure 2b: Multiple daughter cysts and a thick capsule.

DISCUSSION

Primary involvement of skeletal muscle in hydatid disease is not common, and often isolated. A case series of nine patients with primary skeletal muscle hydatid cyst show the lesion confined to the muscles, with no involvement of bone. The location was the thigh in six patients (Biceps in two, and quadriceps in 4 patients), and the popliteal fossa (gastrocnemius) in one, the arm (triceps) in one, and the shoulder (infraspinatus) in one patient each.[4] The clinical presentation depends on the size and site of the lesion. In most of the cases, these lesions present as slowly growing painless mass.

It has been suggested that the low prevalence of this form of disease could be due to the physical barriers to the haematogenous dissemination of cysts in the form of hepatic sinusoids and pulmonary capillaries. In addition to this, higher lactic acid level in skeletal muscle and mechanical factors, such as contractility, may hinder the development of cyst.

The rarity of primary skeletal muscle hydatid cyst causes difficulty in preoperative diagnosis and often it is mistakenly considered as soft tissue tumour. Serology is not always helpful in making diagnosis of primary skeletal muscle hydatid cyst. A negative serology does not rule out the diagnosis of *echinococcosis*.[5] Arazi *et al*. found that indirect hemagglutination teast was positive in only 27% of patients (4 out of 15) in their case series of musculoskeletal hydatid disease.[6] Ultrasonography (US) is a non-invasive and inexpensive. It is most commonly used imaging modality in the diagnosis of this disease, and cysts could be further classified according to the US criteria of Gharbi.[7] CT is superior in detecting calcification in cyst wall as well as association with bone. MRI is an important imaging modality not only for the detection, but for characterization of soft tissue masses also.

Incisional biopsy and marginal excision in case of hydatid cyst involving skeletal muscle is contraindicated, because this can lead to dissemination and the allergic reaction as the hydatid cyst fluid contains significant amount of protein which is extremely toxic to host. The excisional biopsy was planned of this patient keeping in mind the diagnosis of soft tissue tumour; but it remained inconclusive. Different types of serological tests like indirect hemagglutination (IHA) and ELISA may help in diagnosis. In this case, these tests were not done as there was no suspicion of primary muscular hydatid cyst.

Complete surgical excision of the cyst is the treatment of choice.[8] For inoperable cases, medical treatment with or without percutaneous aspiration-injection re-aspiration (PAIR) is regarded as alternative option for the treatment.[9,10] If it is technically not easier to remove the cyst completely (large, complicated cysts or cysts which are firmly adherent to the surrounding muscles). Intraoperative drainage and irrigation with scolicidal agents is an alternative. In any case, the spillage of the

parasite should be avoided intraoperatively in order to prevent recurrence of the disease. However, if it occurs, then adjuvant therapy with albendazole is found to decrease the recurrence significantly. Complete surgical resection without antihelminthic therapy in seven patients (in a case series of 9 patients), was successful with no recurrence observed in a follow-up period ranging from 1 to 8 years.[4]

In this case, the suspicion of a soft tissue tumor other than a hydatid cyst limited the administration of albendazole preoperatively; but intraoperative wash out with hydrogen peroxide was performed repeatedly in order to prevent the recurrence. On follow-up, there was no recurrence.

Hydatid cyst, present in skeletal muscles, may confuse with a soft tissue tumor when the investigations performed cannot clarify the diagnosis. This is further confusing, when there is a history of primary hydatid lesion in lung or liver or previous surgical history of cyst excision. A higher index of suspicion of hydatid cyst in appendicular soft tissue mass should be considered, especially in endemic regions.

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CASE REPORT

Caroticocavernous Fistula: Successful Reversal by Endovascular Treatment

Muhammad Mateen Amir[1] and Naveen Abubakar Bugvi[2]

ABSTRACT

A 22-year male presented in outpatient department with right sided proptosis and chemosis, after a roadside accident two months back. He was on treatment for orbital cellulitis and taking steroids with antibiotics, both orally and topically. He was also on topical antiglaucoma agents. Detailed examination revealed bruit on auscultation. On magnetic resonance imaging (MRI), dilated superior ophthalmic vein led to diagnosis of caroticocavernous fistula (CCF) and the patient was referred to Neurosurgery Department of Lahore General Hospital (LGH), Lahore, where digital subtraction angiography (DSA) and balloon catheterization by a radiologist in the same sitting, resulted in successful closure of fistula and amelioration of all the signs and symptoms within six weeks.

Key Words: *Caroticocavernous fistula. Proptosis. Digitial subtraction angiography.*

INTRODUCTION

Carotid cavernous fistulas (CCFs) are communications between the cavernous sinus (CS) and internal carotid artery (ICA). Barrow classifies fistulas into four types.[1] Type A are of direct type due to direct communication of ICA with the CS, usually as a result of closed head trauma. These fistulas are due to tear in the arterial wall and are of high flow type. They rarely resolve spontaneously, and often require intervention.[2] Other types of fistulas are of indirect type due to abnormal communication between meningeal branches of internal or external carotid arteries. Type B are communications between meningeal branches of internal carotid and CS, while Type C are communications between meningeal branches of external carotid artery and CS. Type D fistulas are communications between meningeal branches of both external and internal carotid arteries with CS. These fistulas have low blood flow; and consequently, the clinical features are less severe. Diagnosis of CCFs is usually delayed for a few weeks because the clinical features are due to increased venous pressure in the superior ophthalmic vein. Timing of diagnosis is important, as permanent loss of vision may develop. The most common signs and symptoms are conjunctival hyperaemia, pulsatile exophthalmos, orbital murmur, ophthalmoplegia, decreased visual acuity and pain in the orbit.[3,4]

CASE REPORT

A 22-year male presented in eye outpatient department (OPD) of Al-Khidmat Teaching Hospital, Mansoora with the complaints of right sided proptosis, lid swelling, and chemosis for one and a half month (Figure 1A). Patient gave history of road side accident two months back, for which he remained admitted in emergency department of some local hospital for two days and then discharged in a good condition. He gave history of development of pain, redness, proptosis, and slightly decreased vision after a week following injury, all increasing gradually. He remained under treatment somewhere else where he was diagnosed as a case of orbital cellulitis and was put on steroids with antibiotic, both orally and topically with topical anti-glaucoma as well.

On examination, he had 6/9 vision in right eye and relative afferent papillary defect. There was slight disc pallor but rest of the fundus was normal. His intraocular pressure (IOP) was 26 mmHg in right eye and 14 mmHg in left eye. CT scan, Ocular Coherence Tomography (OCT) and automated visual fields had already been done. His CT Scan was normal; while OCT of right eye showed decreased thickness of retinal nerve fiber layer (RNFL), and automated visual fields showed inferior hemifield loss in the same eye. Detailed clinical examination was done, which showed bruit on auscultation of right orbit. He was advised MRI brain and orbit in suspicion of CCF. On MRI, there was dilated tortuous right ophthalmic vein which further confirmed the diagnosis (Figure 2).

The patient was referred to Neurosurgical Department for further intervention, where digital subtraction angiography (DSA) with ballon embolisation of CCF was done in the same sitting. Right internal carotid artery on DSA showed CCF. There was diversion of flow to CS and its tributaries with almost absence of flow in right ICA (Figure 3A). After closing the fistula with a detachable balloon, the left carotid angiogram showed the right sided intracranial perfusion maintained through the collateral circulation via circle of Willis (Figure 3B).

Symptoms were improved after surgery. There was disappearance of vascular murmur in the orbital region. Proptosis, chemosis, tortuosity and thickening of the

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Figure 1A: Right proptosis 2 months after RSA.

Figure 1B: After treatment.

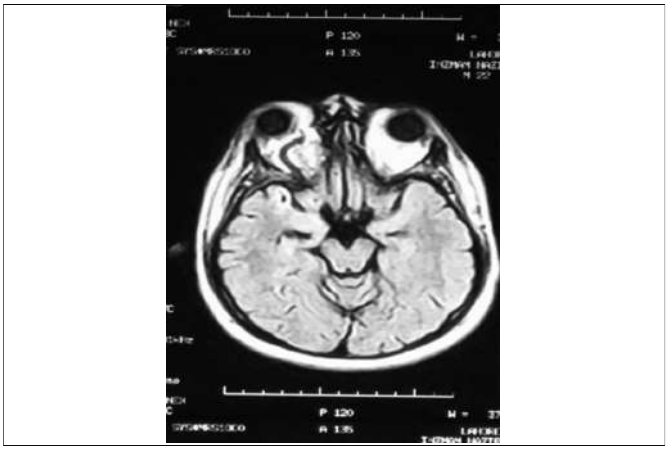

Figure 2: Dilated right superior ophthalmic vein.

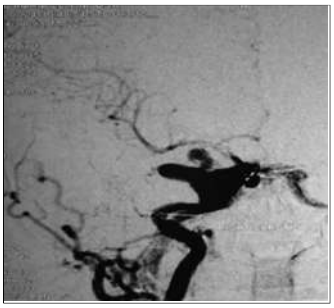

Figure 3A: Right caroticocavernous fistula.

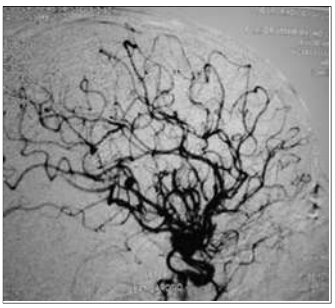

Figure 3B: Left carotid after closure of caroticocavernous fistula.

blood vessels in the sclera disappeared. The raised IOP became normal within 10-15 days without medicines. The patient's vision improved to 6/6 after two weeks. The patient remained symptom-free during two years of follow-up (Figure 1B).

DISCUSSION

CCFs usually present with red eye due to episcleral congestion and chemosis. It is not rare to miss the diagnosis as reports are showing that CCF was mistaken initially as conjunctivitis. The wrong diagnosis misleads the clinician; and delay in diagnosis may damage vision.[5] These patients may present in dental department after maxillofacial trauma.[6] The definite diagnosis can be confirmed by angiography. Glaucoma is a complication that occurs in CCFs. Increased episcleral pressure and vortex venous pressure may result in increased IOP, causing secondary glaucoma.[7,8] Auscultation for potential murmur is an appropriate clinical approach. MRI or CT scan helps in diagnosis. Indirect signs of these fistulas such as engorgement of the cavernous sinus region or abnormally dilated superior ophthalmic vein are present. This patient, having red eye and proptosis, was misdiagnosed initially as orbital cellulitis by some observers as there was no visible pulsations present and because signs appear late. Presence of murmer on auscultation led us to investigate possibility of CCF. MRI further confirms the presence of CCF; and DSA with balloon catheterization helps in cure. Ocular manifestations such as proptosis, chemosis, and glaucoma disappear after closure of fistula with balloon catheterization.

Direct CCFs are treated successfully in 90% of all cases and the mortality and morbidity rates are very low. Endovascular treatment for CCF has widely replaced surgical treatment for the last two decades. Direct delivered coils or balloons to the fistula either using trans-arterial or transvenous approach have become the standard conventional endovascular treatment.

Complete occlusion of the CCF with preservation of carotid artery is the ideal end result of endovascular treatment.[9,10] Surgical treatments remain next option when endovascular treatment fails or is not possible. Spontaneous resolution of CCF has been reported rarely.[11] In summary, most CCFs patients visit ophthalmology departments, and early diagnosis and referral for treatment rest on ophthalmologists.

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A Rare Presentation of Cyclitis Induced Myopia

Umar Ijaz, Asad Habib and Hassan Sajjad Rathore

ABSTRACT

Unilateral cyclitis leading to myopia is a rare and clinical relevant entity. In clinical settings, pseudomyopia is generally encountered in the form of accommodative spasm, which is always bilateral. Cyclitis due to inflammation, on the other hand, can cause pseudomyopia unilaterally and it is a very rare presentation. A young male with acute anterior uveitis, presented with acute episode of unilateral myopia. When patient was examined on first visit, there were no cells in anterior chamber; so he was started on cycloplegic eye drops, but his condition didn't improve. Examination on subsequent visit revealed cellular reaction in anterior chamber and narrowing of anterior chamber angles on anterior segment optical coherence tomography (OCT). Treatment for uveitis was started and patient's visual acuity and refractive error improved. Pseudomyopia is a known complication of several drugs and certain medical conditions. The possible mechanism is supraciliary exudation causing relaxation of zonular fibers and increased convexity of the crystalline lens. Myopia in the setting of a mild cellular reaction can easily be missed and has not been reported yet to the best of authors' literature search.

Key Words: *Myopia. Uveitis. Iridocyclitis.*

INTRODUCTION

Myopia or near-sightedness means that light rays coming from a distance are unable to focus on retina, but they focus short of retina causing a blurred vision. Either it can be due to enlarged eye ball (axial myopia) or due to increase dioptric power of lens itself (lenticular). Some medical conditions and drugs are also known to cause a myopic shift. Commonly reported medical conditions are hyperglycemia, scleritis, choroidal inflammation, Vogt-Koyonagi-Harada disease, and juvenile idiopathic arthritis.[1] Certain drugs which are reported to cause a myopic shift include corticosteroids, sulfa drugs, acetazolamides and topiramate.[2-6]

The relationship between uveitis and pseudomyopia is of clinical significance; and can easily be missed. It is rarely reported in the literature,[7] and is, therefore, worth publishing.

CASE REPORT

A 35-year man reported with complaints of decreased vision from his right eye for the last few days associated with mild redness and pain. On examination, he had visual acuity of 6/36 improving to 6/6 with refraction of -3.00 diopter sphere (DS), while that in left eye was 6/6. Autorefractometer (AR) also reported a refractive error of -3.25 DS (Figure 1). Color vision and near visual acuity was normal in both eyes. Both anterior and posterior segment of both eyes were normal. Keeping in mind the possible diagnosis of ciliary spasm, patient was started on cyclopentolate eye drops and then atropine eye drops, but no improvement in visual acuity was observed. Next visit revealed a visual acuity of 6/75 right eye improving to 6/9 with refraction of -4.00 DS along with cells in anterior chamber (AC) in right eye and pharmacologically dilated pupils bilaterally. Posterior segment examination was normal. Intraocular pressure (IOP) measured by Goldman applanation tonometer was 12 and 18 mmHg in right and left eye, respectively. Both these new findings, i.e. difference in IOP and cells in AC, pointed towards cyclitis. To confirm, anterior chamber optical coherence tomography (OCT) was done to measure the anterior chamber angle, which came out to be 17.5 degrees in right eye as compared to 58.9 degrees in the left eye (Figure 2). Patient was started on steroid eye drops 3 hourly and systemic steroids 1 mg/ kg body weight in divided doses.

Patient was asked for follow-up after three days. Cells in AC disappeared, redness and conjunctival congestion disappeared, and visual acuity improved to 6/9 with a refractive error of +0.50 DS on AR. Patient was advised

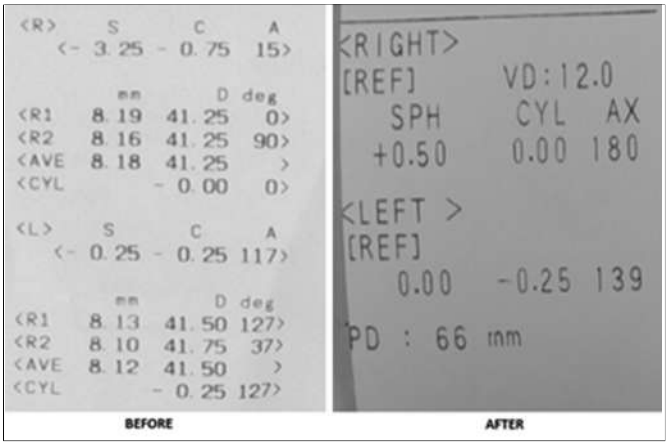

Figure 1: Autorefractometer showing refractive error before and after treatment.

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to continue topical medication and review after two weeks for repeat anterior chamber OCT, which revealed an open angle of 49.3 degrees in the right eye (Figure 2). Vision improved to 6/6 and anterior chamber was quiet. Patient was discharged after follow-up and advised to come back for consultation, if symptoms reappear.

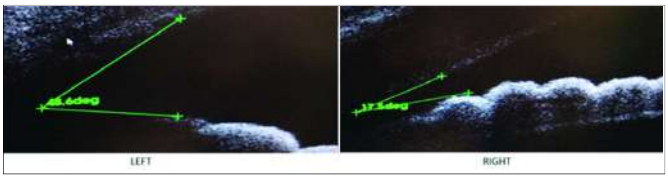

Figure 2: Anterior segment OCT showing angle measurements before treatment.

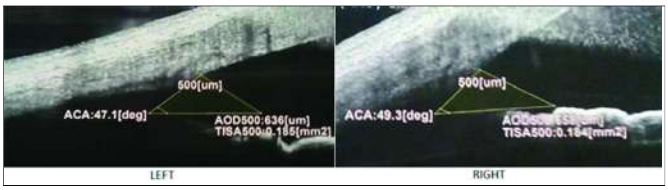

Figure 3: Anterior segment OCT showing angle measurements after treatment.

DISCUSSION

Pseudomyopia may be defined as the sporadic and temporary shift of the refractive power of the eye towards myopia. It may occur alone as a sporadic entity or it may manifest as the spasm of synkinetic reflex, i.e. with pupil constriction and convergent strabismus. The aetiology of this condition varies from organic, traumatic, iatrogenic causes to functional and drug-related causes.[8,9] Pseudomyopia has also been documented in inflammatory conditions like sclero-choroidal inflammation and multifocal choroiditis, though its manifestation in iridocyclitis has never been documented.[10]

When this patient presented to the outpatient department for the first time with blurred vision in right eye, he was suspected as a case of simple accommodative spasm due to increase in refractive power of the eye and absence of any organic pathology, after going through an extensive history and examination protocol. Therefore, he was prescribed cycloplegic eye drops initially and was called for follow-up. On subsequent visits, the myopic shift kept on increasing and initial signs of acute anterior uveitis started to appear. Further investigations and imaging results were consistent with narrow angle, possibly secondary to cyclitis. The patient was then started on topical and oral steroids, to which he responded effectively and his vision returned to normal. In our opinion, the cycloplegics (cyclopentolate, atropine) did not relieve his myopia, initially because there was a persistent edema of the ciliary body which was further relaxing the zonular fibers, making the lens more globular. As soon as the patient was started on steroids, the ciliary edema subsided, the ciliary muscles assumed their normal position, and the pseudomyopia was relieved.

In conclusion, acute unilateral iridocyclitis can present as pseudomyopia. To the best of our knowledge, this was the first case in our setting which presented as pseudomyopia in a backdrop of acute iridocyclitis, and such a rare presentation of this disease has never been reported before. In contrast to treatment by cycloplegics only, this disease entity responds well to the routine acute anterior uveitis treatment.

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Cold Agglutinins in Peripheral Blood with Atypical Cells with an Owl-Eye Appearance in Bone Trephine

Ayesha Majeed Memon and Farheen Karim

ABSTRACT

Autoimmune hemolytic anemia (AIHA) is a form of hemolytic anemia in which red cells lysis occurs due to presence of an autoantibody. Association of AIHA is well known with lymphoproliferative disorders, especially with non-Hodgkin's lymphoma. However, AIHA in association with Hodgkin's lymphoma is seen occasionally. Of the AIHA associated with Hodgkin's lymphoma, most are of warm type or mixed type. Cold AIHA, as seen in our case, is very rare in Hodgkin's lymphoma.

Key Words: *Hodgkin's lymphoma. Autoimmune. Hemolytic anemia.*

INTRODUCTION

Autoimmune hemolytic anemia (AIHA) is an acquired clinical condition which is characterised by the presence of autoantibodies that bind to the surface of circulating red blood cells, leading to hemolysis and shortened red blood cells survival. AIHA is very rarely reported in patient with Hodgkin's lymphoma, with an approximate incidence of around 0.2-4.2%.[1,2] Case reports and reviews have shown that AIHA occurs mostly at stages III and IV of Hodgkin's Lymphoma.[3] Most of the cases reported show an association of Hodgkin's lymphoma with either warm or mixed type of AIHA.[4]

Here, we report an unusual case of cold type AIHA in association with classic Hodgkin's lymphoma.

CASE REPORT

A 20-year female presented with a history of fever, weakness and weight loss for the last 2 months. General physical examination revealed pallor and cervical lymphadenopathy. Baseline complete blood counts showed hemoglobin: 102 g/L, HCT: 5.1%, mean corpuscular volume (MCV): 91.1 fL, mean corpuscular hemoglobin (MCH): 182.1 pg, white blood cells (WBC): 4.5 x 10^9/L, and platelets: 452 x 10^9/L. Peripheral blood film showed numerous red cell agglutinates (Figures 1A and 1B). White blood cells and platelets were normal on film. Considering the red cell indices and presence of agglutinates on peripheral blood film, the blood sample was incubated at 37°C water bath for about half hour. Post-incubation sample was re-run and peripheral blood film reviewed. Red cell agglutinates disappeared after incubation (Figure 1C) and red cell indices were corrected as hemoglobin: 100 g/L [Reference range: 113-145 g/L], HCT: 29.3% [Reference range: 35.4-42.0%], MCV: 89.9 fL [Reference range: 76-96 fL], MCH: 31.6 pg [Reference range: 26-32 pg], WBC: 4.6 x 10^9/L [Reference range: 4-10 x 10^9/L] and platelets: 396 x 109/L [Reference range: 150-400 x 10^9/L]. Direct antiglobulin test [DAT] was positive (3+). Monospecific DAT was positive for C3d

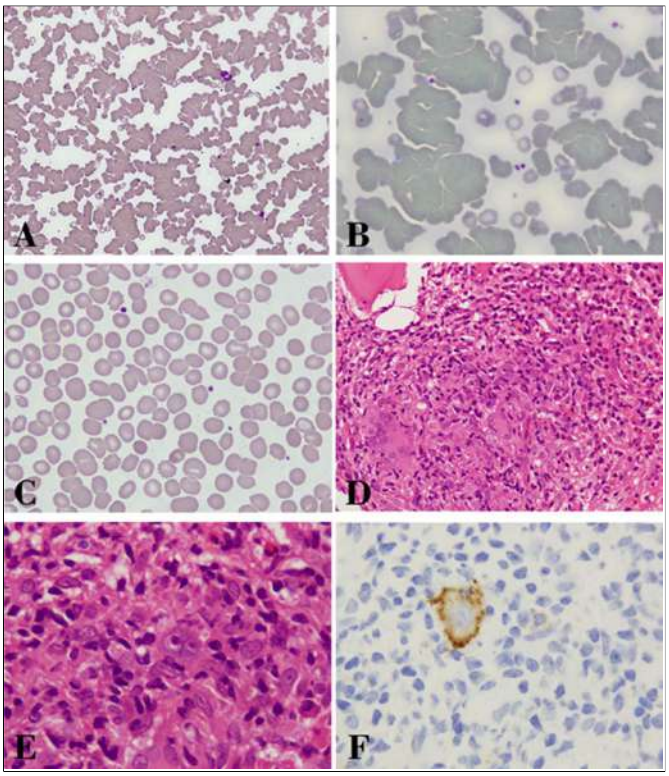

Figure 1: Peripheral blood film showing numerous red cell agglutinates [A and B]. Peripheral blood film after incubation at 37°C showing disappearance of agglutinates [C]. Bone trephine showing few granulomas with multinucleated giant cells [D]. Bone trephine with large mononuclear cells exhibiting owl's eye appearance [E]. CD 30 positive in large atypical mononuclear cells [F].

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(3+) and negative for IgG. Cold agglutinin titre was not done in this patient. Other laboratory findings favouring hemolytic anemia included reticulocyte count of 0.04 proportion of red blood cells [Reference range: 0.003-0.01], lactade dehydroguse (LDH) of 7.51 µKat/L [Reference range: 3.47-6.31 µKat/L], and indirect bilirubin of 25.6 µmol/L [Reference range: 1.71-13.68 µmol/L].

Bone marrow examination was done for workup of cold agglutinin disease. Bone marrow was dry tap. Bone trephine biopsy was a good length specimen showing effaced architecture. Cellular areas showed pleomorphic background comprising of mainly inflammatory cells including lymphocytes, plasma cells and eosinophils. Few granulomas were seen comprising of multi-nucleated giant cells (Figure 1D). Several interspersed large mononuclear cells were also noted exhibiting owl's eye appearance (Figure 1E). Few areas showed intense fibrosis. These large atypical mononuclear cells stained positive for CD 30 (Figure 1F) and negative for CD 20 and CD 3 on immunohistochemical stains. Findings were suggestive of bone marrow involvement with Hodgkin's lymphoma. For further confirmation, lymph node biopsy was advised. Cervical lymph node biopsy was consistent with the Classical Hodgkin's lymphoma. The patient was given 6 cycles of ABVD (doxorubicin, bleomycin, vinblastine, and dacarbazine) and responded to the treatment. DAT was performed after 6 months of treatment and was found to be negative.

DISCUSSION

The exact mechanism of AIHA in Hodgkin's lymphoma is not clear. It is hypothesised that the autoantibodies are produced by neoplastic cells and an immune regulatory phenomenon is involved. It is postulated that there might be an autoimmune mechanism at the initial stages of Hodgkin's lymphoma in which autoantibodies are produced against the neoplastic lesion and erythrocytes as a paraneoplastic phenomenon. In addition, Hodgkin's lymphoma patients are known to have an impaired cell-mediated immunity. This is due to a qualitative defect in T lymphocytes function and decreased number of cytotoxic T cells. Reduction in cytotoxic T cells may cause hyperactivation of B cells leading to increase autoantibody production.[5,6]

This case highlights the importance of diagnosing cold agglutinin disease. Cold agglutinin disease is not indolent, therefore, early diagnosis and identification of underlying cause is very important. While evaluating a case of hemolytic anemia, either warm or cold type, Hodgkin's lymphoma must be kept in mind.

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Botulinum Toxin A and Task-Specific Training for Hand Dystonia due to 5-Year Old Stroke

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ABSTRACT

Focal hand dystonia after stroke, a result of maladaptive plasticity, impairs hand function, affects daily activities, and undermines independence. A 59-year male patient, who had developed focal hand dystonia after suffering from ischemic stroke 5 years ago, received training following an injection of Botulinum Toxin A (BoNTA). Task-specific training for a duration of 60 minutes per day for 3 days per week was provided for 12 weeks. Assessments were done by using arm dystonia disability scale, action research arm test, Fugl-Meyer assessment of upper extremity, and stroke-specific quality of life. Substantial improvement was observed in all the parameters, at short-term follow-up.

Key Words: *Focal dystonia. Task-specific training. Botulinum toxin. Fugl-Meyer assessment. Stroke.*

INTRODUCTION

Dystonia is labelled secondary when it occurs as a result of a known pathological condition, such as stroke. Dystonia had been related to the dysfunction of basal ganglia (especially putamen) in the past. Some studies have also associated dystonia with structural changes or lesions in the brainstem, spinal cord, and peripheral nerves.[1] Dystonia affecting function of a specific body part is termed as focal dystonia.[2]

Post-stroke dystonia manifests in two major forms. Static spastic dystonia occurs at rest resulting in flexion of elbow, wrist, and fingers along with shoulder adduction. It hinders both active and passive movements. Action-induced spastic dystonia, on the other hand, occurs only during particular activities such as writing (writer's cramp). Muscle co-contraction and failure of reciprocal inhibition are core features of dystonia, as a result of extra-pyramidal or basal ganglia disorders.[3]

Task-specific training is the repetition of a specific task until expertise is reached. More challenging tasks are added as a means of progression. Task-specific training may enhance the effects of BoNTA.

CASE REPORT

A 59-year right-handed male presented with complaint of inability to use his right hand. Patient suffered from ischemic stroke (left middle cerebral artery) 5 years ago. He was well-oriented in time, person and place and was fully alert. Sensory examination did not show any notable deviations in both upper and lower limbs. Patient had typical focal dystonia of static spastic type with wrist and fingers fixed in flexion. His lower limb showed muscle strength of 4/5 on manual muscle testing, while different strength grades were observed in various muscle groups in the upper limb (shoulder flexors: 4/5, elbow flexors and extensors: 3+/5, wrist flexors and finger flexors: unable to test as they were fixed in flexed posture due to spastic dystonia). Coordination (both equilibrium and non-equilibrium) and balance were assessed using standardized tools (Rhomberg test, single leg stance etc.) and were found to be normal. The patient walked without any significant gait deviations and did not require assistance. For upper limb assessment, four scales were used; arm dystonia disability scale (ADDS) for measuring dystonia, action research arm test (ARAT) and Fugl-Meyer Assessment of Upper Extremity (FMA-UE) scale for the assessment of arm motor function and stroke-specific quality of life (SS-QOL) for measuring the quality of life.

Patient was injected with BoNTA in flexor digitorum superficialis, flexor digitorum profundus, flexor carpi ulnaris, flexor carpi radialis, biceps brachii and flexor pollicis longus muscles. After one week of injection, patient was provided with task-specific training for 60 minutes/day, three days a week for a total of 12 weeks (Table I). The daily session was broken into 3 sets of 20 repetitions each for every task. Progression in difficulty of exercise was made as patient improved.

Data was collected at baseline then after 4 weeks, 8 weeks and 12 weeks of intervention. ARAT, FMA-UE, and SS-QOL scores and percent improvement, compared to the baseline score, are presented in Table II. Arm motor function during various phases of the rehabilitation is graphically presented in Figure 1.

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Similarly, marked improvement was also observed in ADDS. The patient progressed from category 3 (severe disability) at baseline to grade 2 (moderate disability) after 4 weeks, and grade 1 (mild disability) after 8 and 12 weeks.

Table I: Task-specific training activities across different weeks.

Week I and II
- Reaching for glass on the table
- Pushing bottle aside
- Rolling a tin under hand
- TheraPutty hand exercises
Week III and IV
- Elbow locked and hitting the bottle with wrist
- Bringing glass to mouth
- Bimanual tasks pouring and transfering water
Week V and VI
- Finger tapping
- Folding towel
- Picking cards
- Drawing dots and lines on paper
- Lifting blocks of different colors
- Switching on and off
Week VII and VIII
- Turning pages of newspaper
- Coin swipe
- Picking Ludo bullets
- Catching & throwing ball
- Threading macaronis
Week VIII to XII
- Picking beans of different colors and putting them in different bowls according to colors
- Holding pencil and writing words
- Stacking different coins on one another

Table II: Training-induced changes in arm motor function and quality of life.

	Baseline	4 weeks	8 weeks	12 weeks
ARAT	25	35 (40%)	46 (84%)	49 (96%)
FMA-UE	28	34 (21%)	38 (36%)	47 (68%)
SS-QOL	96	117 (22%)	142 (48%)	151 (57%)

% signifies improvement compared to the baseline score.

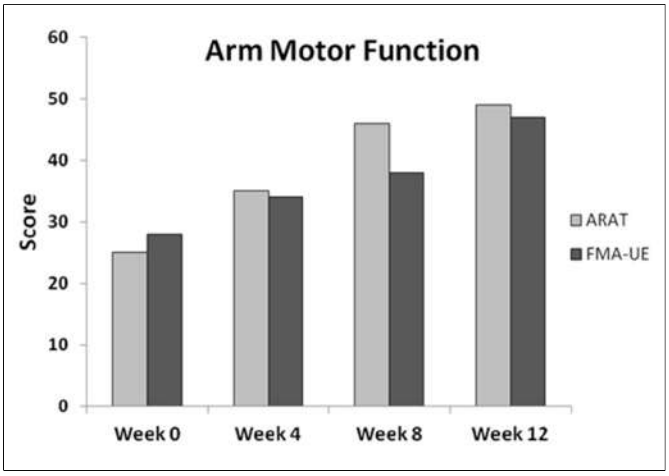

Figure 1: Arm motor function during various phases of the rehabilitation.

DISCUSSION

A combination of BoNTA and task-specific training caused marked improvements in arm motor function, disability, and quality of life. These findings are supported by results of previous studies. For instance, Batla *et al*. recommended BoNTA as the treatment of choice for focal dystonia.[4] Botulinum is safe and effective in treating focal hand dystonia even after more than 10 years of therapy.[5] Rosales explained the rationale for use of BoNTA in dystonia. According to him, dystonia is a multi-segmental disorder that affects not only central but also peripheral nervous system leading to increased muscular tone or muscle spasms, which may cause abnormal posturing of the affected body parts. He explained that these dystonic symptoms can be reversed by chemodenervation including intramuscular administration of BoNTA or phenol.[6] Similarly, Zeuner and coworkers reported that development of dystonia is a consequence of abnormal plasticity. They developed a motor training programme for patients suffering from focal hand dystonia and reported improvements on Fahn dystonia scale, when provided for a period of 4 weeks; however, they recommended studies with long follow-ups in order to get more objective improvements.[7] Furthermore, a review by Thanganatt *et al*. stated that different techniques, beside surgery and other pharmacological treatments, can be used for treatment of focal hand dystonia such as sensory retraining, limb immobilisation, biofeedback, electrical nerve stimulation, and deep brain stimulation. They also reported the beneficial effects of BoNTA and recommended its use for treatment of focal hand dystonia.[8]

Berque *et al*. conducted a study on focal hand dystonia and reported marked improvement in hand function when constraint-induced movement therapy was provided with motor control training.[2] To explain the mechanism, Harvey concluded that plasticity was directly related to task-oriented activities and their incorporation in treatment regimens is imperative in various neurological disorders.[9] Large-scale, multicenter clinical trials to objectively evaluate the effectiveness of combination of BONTA and task-specific training are highly suggested.

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Bipolar Affective Disorder in a Patient of Profound Deafness

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ABSTRACT

Profound deafness is a lifelong impairment, leading to the physical disability as well as poor psychological adjustment. We herein present a mental health disorder rarely seen among the patients of profound deafness. A 16-year deaf and dumb girl, previously treated for depression, presented with unusual laughter, irritability, flight of ideas, decreased sleep, ideas of self importance, and decreased social functioning and educational performance. These problems were understood by the parents via sign language, who interpreted them to the interviewer. Her Young Mania Rating Scale (YMRS) score was 19 and Brief Psychiatric Rating Scale (BPRS) score was 52. She was diagnosed as a case of bipolar affective disorder (BPAD). Marked improvement in the symptoms and social and educational performance was noted after two weeks of the treatment with sodium valproate, resperidone and clonazepam. Treatment options were explained to the patient with risks and benefits, and she was involved in the decision-making. This case report highlights the importance of accurately diagnosing and managing a rare mental health disorder among the physically handicapped people, especially those who cannot communicate effectively and explain their unusual subjective experiences.

Key Words: *BPAD. Profound deafness. Liaison psychiatry.*

INTRODUCTION

Profound deafness is defined as the hearing loss greater than 81 db in which the patient is unable to hear and understand even a voice shouted out loud. Mental health disorders have been studied among the deaf individuals for long and they remain an area of interest for the researchers and mental health professionals due to the diagnostic and therapeutic challenges.[1-3] Deaf individuals are unable to understand the speech of the people around them through their auditory circuit. Due to their communication barriers, they are more prone to emotional, physical and sexual abuse,[4,5] which raises their chances of developing a mental health disorder including the bipolar affective disorder (BPAD).[6]

Depression, psychotic illnesses and impulse control disorders have been commonly encountered among the deaf population, whereas BPAD has been a less frequent diagnosis among this group.[7]

We present a case of BPAD in a young female who has been previously treated for depression and this time presented with a manic episode, which was treated effectively with the routine medication.

CASE REPORT

An unmarried 18-year deaf and dumb female was brought by the parents with the complains of abnormal laughter, irritability, flight of ideas, ideas of self importance, decreased sleep and a decline in social functioning and educational performance. The episode of laughter lasted for 10 to 15 minutes and used to occur once or twice in a day for the last one month. She could not speak; but from sign language and gestures, her increased pressure of thoughts and flight of ideas could be assessed. Parents reported that she used to get exhausted with the sign conversation, and at one moment she was arguing for a Dubai visit and on the next moment she used to ask about the plans of Karachi trip. She had decreased need for sleep, and occasionally the episodes of laughter and prolonged purposeless sign conversation continued for most part of the night. Her academic performance has also declined markedly for the last one month, and her relationship with the siblings and friends became strained due to her tendency to become irritable over minor issues. The patient's family did not report any unusual behaviours or difficulties in daily functioning prior to the onset of current episode of illness about a month ago. There was no history of any head injury or illicit drug use.

She was diagnosed as a case of profound deafness at one year of age when her parents took her to the E.N.T specialist with complains of no response at all to the routine sounds and voices. Brainstem Evoked Response Audiometry (BERA) was done which confirmed the diagnosis by showing no response at all.

She started education at the age of 6 years, in a special school and achieved all other milestones normally without any significant medical, surgical or psychiatric history. She was diagnosed by a consultant psychiatrist as a case of moderate depressive episode one year ago and was put on Ecitalopram 10 mg for 9 months which was then tapered as her symptoms resolved. She was

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off antidepressant medication for 3 months, prior to onset of current episode and there was no significant history of any other psychosocial stressor or childhood abuse. Her parents were trained but siblings were not formally trained in the sign language. There was no positive family history for any psychiatric disorder.

She was a young girl properly dressed in bright colors sitting anxiously on the sofa. Her vital signs and physical examination were unremarkable. She had episode of laughter twice during the interview session and her mood was extraordinarily cheerful. She was constantly trying to interact with the mother through the sign language, which was manifestation of the pressure of her thoughts. Presence of delusions or overvalued ideas could not be elicited. Abnormal laughter could be attributed to the patient responding to an internal hallucinatory experience but no direct evidence could be elicited as she actively denied any visual or auditory hallucination. Her long-term and short-term memory was intact and there was no insight as she told that she is completely fine and her parents are having baseless worries about her health and behavior.

Differential diagnosis included manic episode with or without psychotic features, BPAD, schizophrenia, dissociative reaction, attention seeking behaviour, and acute transient psychotic disorder (ATPD). ATPD was ruled out as duration of symptoms was now more than a month and she previously had a depressive episode as well.

Investigations were performed according to the bio-psycho-social model. All the baseline biological investigations were normal. Serum prolactin was within the reference range. CT-scan brain was also unremarkable. In the light of these findings, no organic cause could be related to her current mental state. Her fresh pure tone audiometry (PTA) results showed profound deafness and there was no response on BERA.

Psychological investigations included the administration of the psychometrics. She completed the self-administered questionnaires with the help of her parents. Her YMRS score was 19 and BPRS score was 52. Generalised anxiety disorder-7 (GAD-7) and back depressive inventory (BDI) scores were within normal range. GAD-7 was applied with the rational to rule out anxiety as she had restlessness and irritability and she could not speak, so quantification with the psychometric scales was done. One year ago, BDI score was 21 when she was treated for the depressive episode. Social investigations included interview from the parents and feedback from the siblings.

After the detailed history, mental state examination and the results of psychometrics, she was put on sodium valproate 500 mg, resperidone 2 mg and clonazepam 0.5 mg daily. Her parents were briefed in detail about the risks and benefits of all the treatment options available, and they agreed upon putting her on combination treatment. After two weeks, she showed marked improvement in the symptoms. Her laughter episodes decreased, sleep improved, and flight of ideas and pressure of thoughts settled to the extent that her social and occupational performance started improving. After one month of treatment, her YMRS score was 9 and BPRS score was 30.

The final diagnosis was BPAD with current episode of mania with psychotic features. After the appropriate treatment, there was dramatic improvement in her condition. On follow-up after 2 to 4 weeks, clonezepam was gradually tapered and sodium valproate and resperidone were continued in the similar dose and detailed briefing was given on their harmful side effects, especially about the teratogenic potential of sodium valproate and associated complications in a young female who is already handicapped. Plan was formulated to taper sodium valproate, once complete remission has been achieved and continue the treatment with mood stabilizing antipsychotics.

DISCUSSION

Adolescent patients suffering from the manic episode of BPAD usually present with grandiosity, over talkativeness, disinhibition and irritability, which are the chief symptoms of this condition, so usually the diagnosis among them is not very difficult.[8] Our patient was unique in a sense that she could not hear or speak, so the presentation was atypical. Routine methods of history taking and mental state examination could not be applied for her evaluation. These limitations made this case a diagnostic challenge for the psychiatric team.

Psychiatric disorders are prevalent among the patients who are hard of hearing or profoundly deaf. Impulse control disorders, ADHD, and intellectual disabilities are more prevalent in this group as compared to the hearing population.[7] This patient had none of these problems in childhood or adolescence. She just had an episode of depression one year ago, which was treated by the psychiatrist with the selective serotonin reuptake inhibitors (SSRIs). Currently, she presented with symptoms mentioned in the previous section which were successfully managed by the mood stabilizer and the antipsychotic drugs. Good clinical response and considerable reduction in the scores of YMRS and BPRS after the above mentioned treatment further supported the diagnosis of BPAD.

This case presented a diagnostic challenge given the limitations of the hearing disabled patient. Valuable information was obtained from the patient's family and the collected data had to be carefully interpreted to reach the diagnosis. Role of psychometrics was significant in overcoming this challenge. Psychometrics

had been used effectively in the past too among this group of population,[9,10] but there are few limitations to their use. In our case, the patient was literate with no gross intellectual disability; therefore, it was feasible to use the self-administered psychometric questionnaires to confirm the clinical diagnosis and also monitor response to the treatment.

The diagnosis of BPAD rests on the clinical criteria set by the international classification of diseases-version-10 (ICD-10), and severity of the manic or depressive episode can be assessed with the help of the psychometric tools. Both these aspects were catered for in this case. Inability of the patient to hear and speak was the biggest hurdle in completing the psychiatric evaluation. Detailed investigation by the bio-psycho-social model and the use of psychometrics served as a key to make this task achievable and can be applied in such group of patients in future, too, to make an accurate diagnosis.

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CASE REPORT

Primary Renal Epithelioid Angiosarcoma

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ABSTRACT

Primary renal angiosarcoma is a rare tumor. Epithelioid variant of primary renal angiosarcoma is extremely rare and aggressive entity described in literature as a few case reports. It presents as solid looking renal mass as other renal cell carcinomas. Management is not well described due to scarcity of cases and aggressiveness of disease. We hereby report a case of epithelioid renal angiosarcoma in a 62-year female who presented with features of infected perinephric collection. Histopathological and immunohistochemical examinations of the renal specimen revealed lower polar renal tumor with atypical epithelioid cells having eccentric nuclei with coarse chromatin and eosinophilic cytoplasm, which were positive for vascular endothelial (FLI-1, CD 34, CD31 and factor VII) as well as epithelial (CKAE1/AE3) immune markers. Based on the histopathological and immunohistochemical findings, the patient was diagnosed with primary renal epithelioid angiosarcoma.

Key Words: *Angiosarcoma. Epithelioid. Renal.*

INTRODUCTION

Angiosarcomas are high grade malignant tumors arising from the endothelium of blood and lymphatic vessels. They make up 2% of all soft tissue sarcomas.[1-3] Most common origin is from the skin while liver, spleen, bone, and breast are the other less common sites.[1,3] They have the worst prognosis among soft tissue sarcomas. Local recurrence occurs in 1/5th of patients after successful treatment, while half of patients die with metastatic disease.[3] Angiosarcoma involving the kidney is usually metastatic from other viscera.[2] Histologically, angiosarcomas vary from well differentiated tumors with variable endothelial atypia to high grade spindle cell neoplasms. There is one variety, which has neoplastic endothelial cells with predominant epithelioid character.[4] Epithelioid variant of primary renal angiosarcoma is extremely rare and described in literature as only a few case reports. It is highly malignant with poor prognosis.[4-6]

We present a case of epithelioid variant of primary renal angiosarcoma in a 62-year female who presented with features of infective perinephric collection. The histopathological examination of the renal specimen provided the diagnosis. To the best of our knowledge, this is the first case report of primary epithelioid renal angiosarcoma from Pakistan. The authors have obtained written informed consent from the patient for written and electronic distribution of the report.

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CASE REPORT

A 62-year female presented in the urology outpatient department at Shifa International Hospital, Islamabad with the complaints of left flank pain and low grade fever for the last 20 days. She was hypertensive with a past history of total abdominal hysterectomy and ovarian cystectomy with benign histopathology. She was treated for pulmonary tuberculosis 25 years back. On physical examination, she was pale with mild tenderness in the left lumbar area but no mass was palpable. Biochemical profile showed hemoglobin, 9.8 g/dl, total leukocyte count (TLC), 10780/µL, and serum creatinine level 1.39 mg/dl. Renal dynamic computer tomography (CT) scan showed a large multilocular, hypodense predominantly fluid-density lesion involving the lower pole of left kidney and extending into perinephric region encasing the proximal ureter along with mild left hydronephrosis and perinephric stranding. There were a few small mildly enlarged para aortic lymph nodes. Keeping in view the ureteric stone and perinephric collection with hydronephrosis, we performed cystoscopy, left retrograde pyeloureterogram with double J stenting. Intraoperatively there was a small stone in lower ureter and a tight stricture in upper third of ureter. Postoperatively, patient recovered well and flank pain subsided. Urine culture showed no growth. Patient was discharged on third postoperative day on oral antibiotics and analgesics in view that the acute episode had settled down and for further workup afterwards. Patient was advised follow-up after one week but was lost to follow-up.

She presented again in emergency department after one and half months with severe left flank pain, vomiting, high grade fever and pyuria. On physical examination, there was marked tenderness in the left half of abdomen. Biochemical profile showed TLC, 29100/ul, serum creatinine, 1.2 mg/dl, C reactive protein (CRP), 465 mg/dl, and cancer antigen 125 (CA 125), 207/mg/dl.

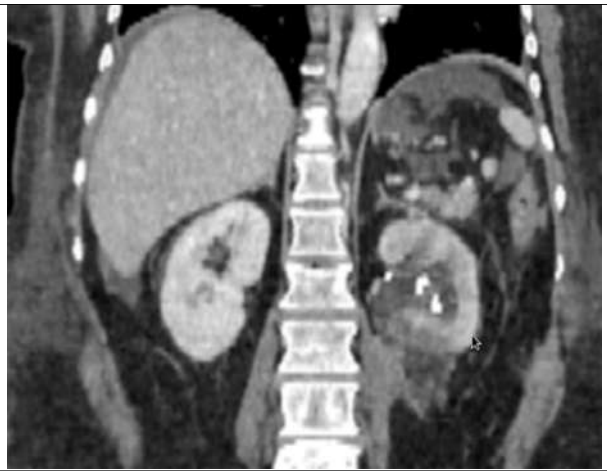

Figure 1: CT scan abdomen and pelvis showing fluid collection along lower pole of left kidney with perinephric fat stranding.

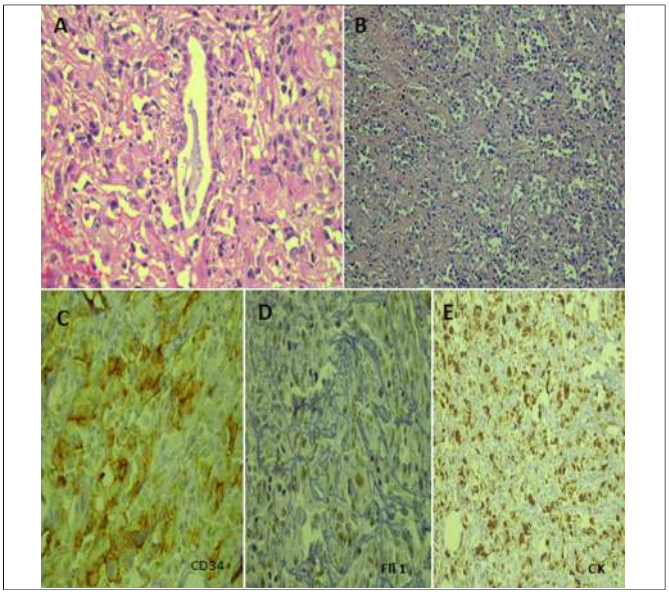

Figure 2: (A) Tumor showing sheets of atypical epithelioid cells. Individual cells have eccentric nuclei with coarse chromatin and eosinophilic cytoplasm (original magnification 200x). (B) Tumor showing vasoformative growth pattern with highly infiltrative vascular channels lined by atypical epithelioid cells. (original magnification 100x). (C, D and E) Immunohistochemistry showing tumor cells positive for CD 34, Fli 1 and CK stains.

Despite giving broad spectrum antibiotics, TLC and CRP remained high. Repeat CT scan showed moderate hydronephrosis with no increase in size of perinephric collection (Figure 1). There were two small hypodense lesions in liver with interval increase in size of left para aortic, preaortic and left common iliac nodes and mild abdominopelvic ascites. Ultrasound guided (USG) aspiration of perinephric collection showed necrotic tissue. Urine cytology, bacterial culture, acid fast bacilli (AFB) staining, gram and potassium hydroxide (KOH) staining as well as aspirate fluid bacterial culture, AFB, gram and KOH staining were negative. USG-guided biopsy of liver lesions was unsuccessful. Hand assisted laparoscopic radical nephrectomy was planned. Per-operatively, there was extensive peritoneal metastasis with no gross pus in renal or perirenal area. Histo-pathology showed necrotic tumor measuring 5.5 × 5.0 × 5.0 cm at lower pole of kidney infiltrating the proximal ureter. Microscopically, there were sheets of atypical epithelioid cells. Individual cells had eccentric nuclei with coarse chromatin and eosinophilic cytoplasm. Also there was vasoformative growth pattern with highly infiltrative vascular channels lined by atypical epithelioid cells (Figures 2A and 2B).

Immunohistochemistry showed tumor tissue positivity for vascular endothelial (FLI-1, CD 34, CD31 and factor VII) as well as epithelial (CKAE1/AE3) immune markers (Figures 2C-2D). Tumor was also present in left adrenal and omental tissue. Diagnosis of primary epithelioid angiosarcoma of kidney was made. Patient was referred to oncologist for chemotherapy.

DISCUSSION

Primary renal angiosarcoma is a very rare tumor. Less than 40 cases have been reported in literature. It usually presents similar to renal cell carcinoma with flank pain, macroscopic hematuria and palpable abdominal mass.[1,3] It occurs most commonly in sixth to seventh decade of life and is rarely found in females.[3,7,8] Environmental exposure to arsenic, thorium dioxide and vinyl chloride are considered as risk factors.[8,9] Primary renal angiosarcoma is a very aggressive tumor and is mostly metastatic when diagnosis is made. It metastasises haematogeneously, mainly to lung, liver and bone.[3,9]

It is usually seen on CT scan as a solid mass with no characteristic signs.[9] Histological findings and immuno-histochemical staining are usually diagnostic.[8] Micro-scopy may show nested and clustered round cells with high nuclear grade. Cancer cells are positive for CD 31, CD 34 and von Wille brand factor.[3,8] Angiosarcomas in which malignant endothelial cell have predominantly epithelial appearance are called epithelial angio-sarcomas. Epithelial cell markers such as broad-spectrum keratins (AE1/AE3) and low molecular weight keratins such as CK8/18, EMA and B72.3 are usually positive in most of the patients.[4,6] In our case, CKAE1/AE3 immunomarkers were positive.

The interesting finding in our case was its atypical radiological appearance, i.e. it was not of a mass but a perinephric collection. Based on this finding and raised TLC and CRP, patient was managed initially for infective collection. This was most likely due to tumor tissue necrosis. Aggressive nature of disease was seen in our case as there were only two months between the occurrence of symptoms and diagnosis but there was extensive metastasis to liver, whole peritoneum, and intra-abdominal lymph nodes.

Initial tumor size and the presence of metastasis at diagnosis are considered significant prognostic factor.

The five-year survival is 32% in patients with tumor size <5 cm, while it is 13% in those with tumor size >5 cm.[2,9]

There is no standard treatment of primary renal angiosarcoma, but surgery is considered as the mainstay of treatment.[1,3] Median survival after surgery is very short (6.28 ±4.96 months).[9] Zenico and colleagues noted that patients with best response, who underwent chemotherapy and radiotherapy, had median survival of 13 months (p>0.005) compared to 7 months with nephrectomy alone.[10] Taxanes and ifosfamide-based chemotherapy are proposed as possible adjuvant therapy.[7] Immunotherapy, including recombinant interleukin 2 and targeted therapies as bevacizumab and sorafenib also give good results.[10]

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Giant Urethral Calculus without Acute Urinary Retention

Zainal Adwin Zainal Abidin[1], Firdaus Hayati[2], Guan Hee Tan[3], Eng Hong Goh[3], Jasman Hafidzul[3] and Md Zainuddin Zulkifli[3]

ABSTRACT

A 46-year gentleman presented with a left-sided lumbar region pain without fever or dysuria. He denied episodes of acute urinary retention. There was a hard mass at the distal urethra with normal laboratory blood tests. Computed tomography urogram revealed a concurrent left renal staghorn calculus and large distal urethral stone. The urethral stone was fragmented via endourologic technique successfully. We report a case of a non-obstructing large urethral calculus in a gentleman with concurrent left renal staghorn calculus and discuss the literature review.

Key Words: *Urethral calculus. Staghorn calculi. Acute urinary retention. Lithotripsy.*

INTRODUCTION

Giant urethral calculi are extremely rare with incidence of less than 1% of all urinary stone diseases.[1] They are endemic in the Middle East and Asia, but rarely exist in the developed countries.[2] These entities occur as a result of stone migration from a proximal source or primarily exist due to strictures, diverticula or other anatomical malformations.[3] Patients typically present with obstructed urination, dribbling, dysuria, and hematuria. Common causative factors are urethral stricture, stasis, or stagnation secondary to a urinary infection, foreign bodies, debris, bladder neck obstruction, lithogenic diathesis, and schistosomiasis.[4] Predisposing factors for *in situ* development of urethral stones include presence of urethral diverticulum or stricture, hypospadias, and meatal stenosis.[5] The stones are usually extracted by using an open technique with occasional closure by reconstructive surgery.[6] Alternatively, endoscopic methods are favoured but there are yet to become a standard management.[7]

We report a unique case of a non-obstructing, large urethral calculus in a 46-year gentleman with concurrent staghorn renal calculus.

CASE REPORT

A 46-year gentleman presented to the emergency department for a complaint of left-sided lumbar region pain for 2 months. The pain was localised and intermittent in nature. He denied any fever or dysuria. His urinary flow was normal without lower urinary tract symptoms or hematuria. Physical examination revealed a tenderness at left lumbar region and a hard mass at the distal urethra, just 2 cm proximal to the meatus. The bladder was not distended. The laboratory blood tests were unremarkable. There was microscopic hematuria with leucocytes seen on urine microscopy. Plain pelvic radiograph showed presence of a calculus in the urethra (Figure 1). Computed tomography urogram showed a left renal staghorn calculus at the mid and lower pole (3 x 1.3 cm) and a large urethral stone (2.5 x 1.2 cm) with multiple smaller urethral stones. He was subjected to laser lithotripsy for the urethral calculus.

A meatal stenosis was noted intraoperatively. He underwent a meatotomy at the dorsal aspect of the glans to allow passage of a rigid cystoscope. A large urethral calculus was seen at the distal aspect of the penile urethra. The calculus was fragmented using laser and lithoclast. All fragmented pieces were removed completely under direct vision (Figure 2). There was no bladder stone and the bladder mucosa was grossly

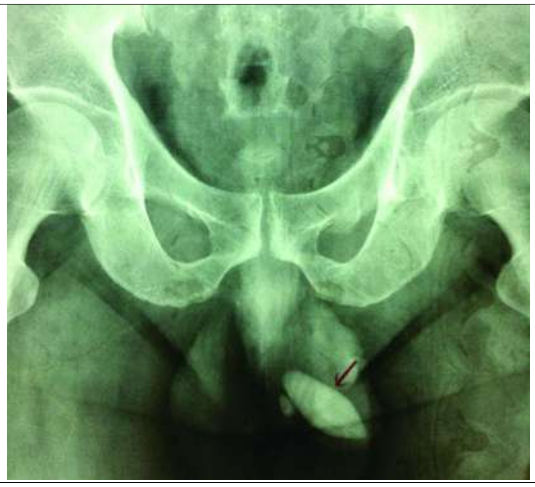

Figure 1: Plain pelvic radiograph showing the calculus (red arrow).

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Figure 2: Fragments of urethral stone removed via endoscopic technique.

normal. Postoperatively, he had urinary catheter inserted for 1 day and after trial of void, he was able to urinate without difficulty. He was discharged the next day and planned for a left percutaneous nephrolithotripsy.

DISCUSSION

Giant urethral calculi are rare urologic entities with incidence of 0.3%.[3] Preponderantly found in the prostatic urethra, they are primarily formed in the prostatic urethra or occur as a result of distal migration from the upper urinary tract. Numerous giant stones have been reported in the literature.[8] Rarely, they can even present as a urethral-cutaneous fistula.[6] Symptoms on presentation are usually urinary retention, frequency, dysuria or stinging in the anus. In this patient, surprisingly there was no acute urinary retention, probably due to the adaptation in urethral lumen size as a result of a long-term, progressive enlargement of the stone.

Most reports contain no data on the constituents of the calculi but Kamal *et al.* reported that 86% of urethral calculi consists of calcium oxalate with a minimum percentage of the stones formed by struvite (magnesium ammonium phosphate) and uric acid.[9] Primary native calculi are usually small and multiple, composed of struvite.[5] They are uniform in structure without a nucleus.[5] They are usually formed either behind a stricture or within a communicating cavity, with the obstruction, stagnation and inflammation being predisposing factors.[5] Migratory stones are much more common and are most often encountered in association with urethral disease and other form of obstruction. They are usually of calcium oxalate or citrate.[5] We postulate that this patient might have had a smaller stone (migrated from the left kidney or upper tract) that travelled to the urethra and lodged proximal to the meatal stenosis, hence becoming a nidus for stone enlargement.

Management of urethral calculi varies according to the size, location and associated disease. Treatment options include retrograde manipulation into the bladder followed by lithotripsy or litholapaxy for smaller stones and ventral meatotomy or urethroscopic methods for larger size.[3] Due to the advancement in equipment and skills, the use of endourology has been shown in recent literature.[10] Successful management of small urethral calculi using laser ablation has been reported by Walker *et al*. and it is a significant advancement in urology.[10] Endoscopic approach provides surgeon with better visualization of the calculus with minimal trauma. This leads to better lithotripsy accuracy, thus minimizing tissue trauma. With giant urethral stones, however, duration of the surgery might be an issue. Further studies are needed to be done to properly assess the advantages and possible complications from laser lithotripsy of the urethral stones.

The standard management of large urethral stones is open surgery, namely urethrotomy with or without urethroplasty.[4] In our patient, we attempted endoscopic treatment using laser and pneumatic lithoclast with successful removal of the calculus completely. We believe the endoscopic modality is feasible and safe, thus should become standard management for urethral calculi.

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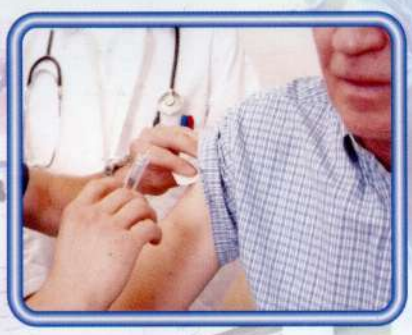

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