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

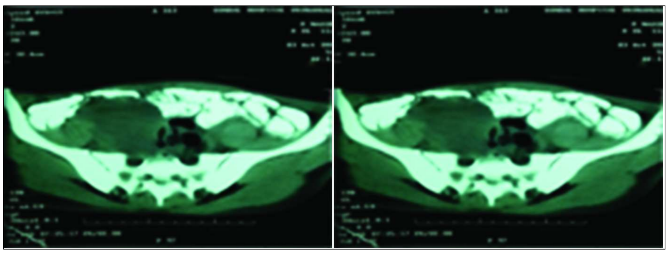

Figure 1: CT-scan showing pelvic mass.

[1] *Department of Urology / Medicine[2], KEMU/Mayo Hospital, Lahore.*
[3] *Department of Medicine, Allama Iqbal Medical College, Lahore.*
[4] *Department of Urology, Postgraduate Medical Institute (PGMI), Lahore.*

Correspondence: Prof. Safdar Shah, Head, Department of Urology, Hemodialysis and Renal Transplantation, KEMU/ Mayo Hospital, Lahore.
E-mail: drsafdar-ali@hotmail.com

Received: April 28, 2016; Accepted: October 30, 2017.

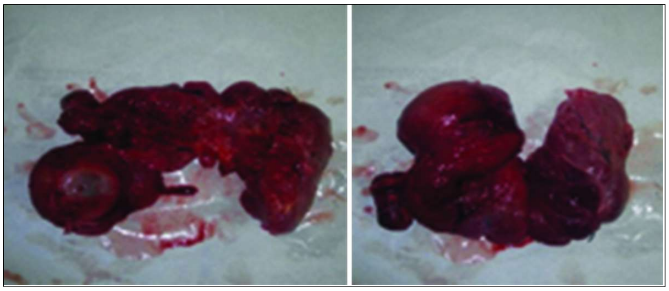

Figure 2: Excised pelvic mass.

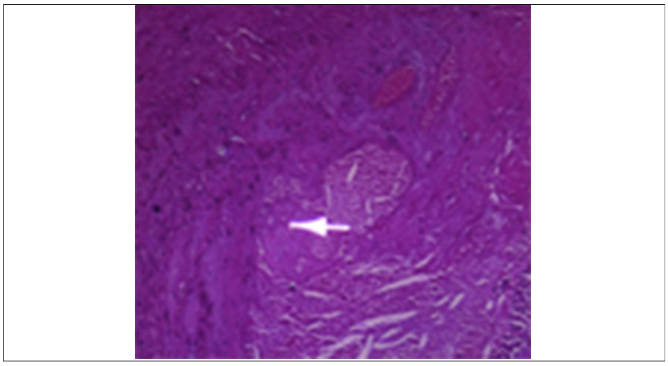

Figure 3: Histological examination shows myxoid stroma and Schiller-Duval bodies.

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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