CASE REPORT

Spontaneous Idiopathic Unilateral Adrenal Haemorrhage (SIAH)
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ABSTRACT
Spontaneous Idiopathic Adrenal Haemorrhage (SIAH) is an unusual surgical emergency which can present with life threatening massive retroperitoneal bleeding. Most of the cases reported in the literature are associated with use of anticoagulation or underlying adrenal pathology such as tumors or cysts. Since this clinical entity is uncommon and clinical presentation is very indistinct, the diagnosis can be easily missed and can be challenging for the treating physicians. Nevertheless a raised clinical suspicion coupled with advances in radiological imaging have considerably improved the detection of SIAH in recent times. We report an unusual case of a 20 years old healthy female student who presented to our hospital with sudden onset of abdominal pain and shock. She was diagnosed as a case of massive spontaneous idiopathic unilateral adrenal haemorrhage, unaccompanied by any hematologic disorder, trauma or underlying pathology. Although patient was hemodynamically unstable at presentation, she was resuscitated promptly, investigated appropriately, hence recovered uneventfully with conservative management alone.

Key Words: Spontaneous adrenal haemorrhage. Idiopathic adrenal haemorrhage. Shock.

INTRODUCTION
Spontaneous Adrenal Haemorrhage (SAH) is a very uncommon abdominal surgical emergency condition which may be unilateral or bilateral.¹ It is more common in the newborn, but can also occur in older children and even adults.² There is usually an underlying pathology associated with this condition. Adrenal Haemorrhage (AH) is usually seen after anticoagulation therapy, trauma, systemic diseases, surgical stress, sepsis or hypotension.³,⁴ Bilateral adrenal haemorrhage is an important clinical entity and if not recognized it may lead to acute adrenal insufficiency and possible death.⁵ In contrast, unilateral adrenal haemorrhage does not lead to adrenal insufficiency but to life-threatening haemorrhage and can also pose a difficult diagnostic challenge.

The present report describes this rare occurrence in a young female.

CASE REPORT
A 20-year previously healthy female university student presented to Acute Medical Assessment Unit (AMAU) with history of sudden collapse. She woke up in the early hours of the morning with left flank pain which was moderate in intensity and associated with nausea. She thereafter collapsed and was brought to the Hospital by the Emergency Medical Services with in route resuscitation. She was on regular oral contraceptive pills and past history was unremarkable. Clinically she was conscious, pale looking and stable hemodynamically with a pulse of 86 bpm, blood pressure of 110/75 mmHg and SpO₂ of 96% on air. Abdominal examination was unremarkable except for mild left flank tenderness.

During the initial hour of assessment in the medical facility she became dizzy again and hemodynamically unstable; her blood pressure dropping to 70/50 mmHg and pulse rate increased to 120 bpm.

She was resuscitated and an initial provisional diagnosis of ruptured ectopic pregnancy was made. Abdominal ultrasound was carried out in AMAU which showed normal adnexa but detected a large retroperitoneal collection raising a suspicion of retroperitoneal haemorrhage. The initial blood analysis results showed haemoglobin of 82 gm/dl and βHCG in the normal range. This normal βHCG along with normal looking adnexa, lack of free intraperitoneal fluid and retroperitoneal haematoma ruled out ectopic pregnancy. Her repeat blood test showed Hb of 57 g/l with a normal clotting profile. Blood transfusion was started and an urgent CT scan abdomen was organized.

Once stabilized clinically, an emergency CT scan with IV contrast was performed which showed a large retroperitoneal hematoma measuring 12x14 cm in size suggesting an active on going bleeding (Figure 1). Conventional aortic angiogram along selective angiogram of phrenic artery, Coeliac trunk, superior mesenteric artery and left renal artery was performed via right femoral artery with an intent to perform therapeutic embolization. Angiography showed a tiny blush of contrast with a prominent vein which was draining into the left renal vein hence in keeping with venous adrenal bleed. No definite arterial leak was identified and therefore, no embolization was performed (Figure 2). The angiogram catheter sheath was left in the groin, to keep an access.
Spontaneous idiopathic unilateral adrenal haemorrhage

Patient was shifted to surgical high dependency unit for monitoring and serial clinical examination to detect any signs of further re-bleed. She was transfused further 02 units of bloods. Blood samples including serum cortisol, ACTH levels and 24 hours urine metanephrines, to rule out any functioning adrenocortical tumor, were all in the normal reference range.

Patient continued to recover in the HDU where she was kept for next 48 hours with serial haemoglobin monitoring and frequent clinical assessment. She was discharged home on the 5th post-admission and she made an uneventful recovery. She had two post discharge visits in the OPD 02 weeks and 06 weeks apart where she was found making an uneventful recovery and is asymptomatic.

DISCUSSION

SIAH is defined as spontaneous bleeding in adrenal gland which is not associated with prior trauma, anticoagulation therapy or any other identifiable cause. It is a very rare clinical entity as most of the cases of unilateral Adrenal Haemorrhage (AH) are associated with pheochromocytoma and a variety of primary adrenal tumors and cysts.

The adrenal gland is inherently susceptible to bleeding due to its predispension to adrenal medullary venous congestion and thrombosis during stress or sepsis. Due to rarity of the lesion and vague clinical presentation, the diagnosis could be easily missed, however, a raised clinical suspicion coupled with advances in imaging have significantly enhanced the detection of SIAH in recent times. The largest series of AH in adult patients has been presented by Vella et al. from Mayo Clinic, comprising of their 25 years experience of AH in 141 cases. They classified AH into 07 different types according to the likely etiology behind these cases as spontaneous, incidentaloma, anticoagulation therapy, trauma, associated with antiphospholipid- and heparin-associated thrombocytopenia, postoperative and severe stress/sepsis.

The incidence of AH in children is almost seven times greater than that in adults. AH in pediatric population is known to manifest typically as an incidental abdominal mass, vomiting, anemia, jaundice or scrotal swelling. There can be various non-specific signs and symptoms of SIAH at the time of presentation in adult population such as abdominal pain, anemia and shock. Unlike bilateral adrenal haemorrhage, a majority of the patients with unilateral AH do not show any signs of adrenal insufficiency. About half of the patient present with abdominal pain radiating to flank. Acute adrenal crisis may occur if bleeding is bilateral and can be life threatening, therefore, assessment of adrenal function and appropriate glucocorticoid and mineralocorticoid replacement is crucial.

On CT scan, adrenal haemorrhage appears as a mass of more than 4 cm. It is generally heterogeneous, can be with or without calcification and variable density. Adrenal hematomas are usually of soft tissue density but these can be hypo-attenuated centrally with a peripheral rim of higher attenuation, hyper-attenuated centrally with a rim of hypo or hyper density. MRI appears to be more accurate than other imaging modalities for diagnosing adrenal haemorrhage. The most common finding is high signal intensity on T1-weighted images, especially if this is located most peripherally. On T2-weighted images signal intensity can be heterogeneous, however, low signal serpiginous areas are characteristic.

The mainstay treatment of AH consists of volume resuscitation, transfusion therapy and correction of any underlying coagulopathy if existing. In case of severe life threatening bleed trans-arterial embolization can be attempted as this allows for hemostasis, with consolidation of the hematoma and surgery can subsequently be performed if required. Although in majority of these patients there is venous bleed and embolisation is usually not indicated.

REFERENCES


