

Spontaneous intraperitoneal haemorrhage of a renal angiomyolipoma presenting with an acute abdomen

C Choudhury, K Maiti, S Chakravarty, S Bhattacharya

Citation

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Abstract

A 36 year old male patient presented to the emergency department with pain over the right iliac fossa and 2 episodes of fainting over the past 2 days. He was found to have tenderness and rigidity over the right side of the abdomen, tachycardia and severe pallor. A CT scan suggested features of lacerated right kidney. An exploratory laparotomy was performed. 1.5 litres of intra peritoneal blood was found followed by a huge haemorrhagic mass involving the kidney. A right sided nephrectomy was performed and the specimen was sent for histopathological examination. The biopsy report stated that it was an angiomyolipoma. Angiomyolipomas are benign lesions found in the kidney and are usually asymptomatic. Retroperitoneal haemorrhage can occur but intraperitoneal haemorrhage though extremely rare is a dangerous complication.

INTRODUCTION

Angiomyolipomas (AML) are benign slow growing tumours composed of a variable mixture of blood vessels, smooth muscles and fat(1). It may be associated with tuberous sclerosis but are mostly sporadic(8). Most angiomyolipomas are small and asymptomatic (1). They are often an incidental finding on imaging done for other purposes. Occasionally, they may cause hematuria or flank pain from spontaneous hemorrhage which is usually retroperitoneal, and this rarely may result in acute abdomen and shock. However intraperitoneal spontaneous rupture is extremely rare.

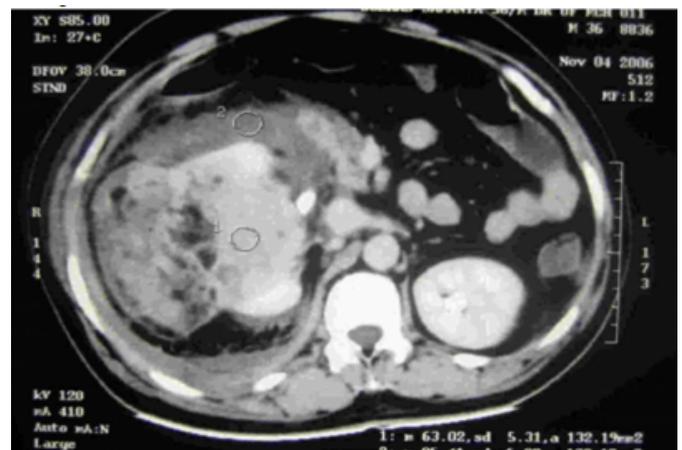
CASE REPORT

A 36 year old male patient presented to the emergency department with pain over the right iliac fossa and 2 episodes of fainting over the past 2 days and no history of trauma. He had no history of haematuria. On examination he was found to have tenderness and rigidity over the right side of the abdomen. He also had tachycardia and severe pallor.

Investigations revealed a blood haemoglobin level of 5.3gm% and a packed cell volume of 16. Contrast enhanced CT scan suggested features of lacerated right kidney in the middle and lower zones with perirenal collection, free fluid in the abdomen and right sided pleural effusion.

Figure 1

Fig 1. CT scan shows renal laceration and perirenal collection

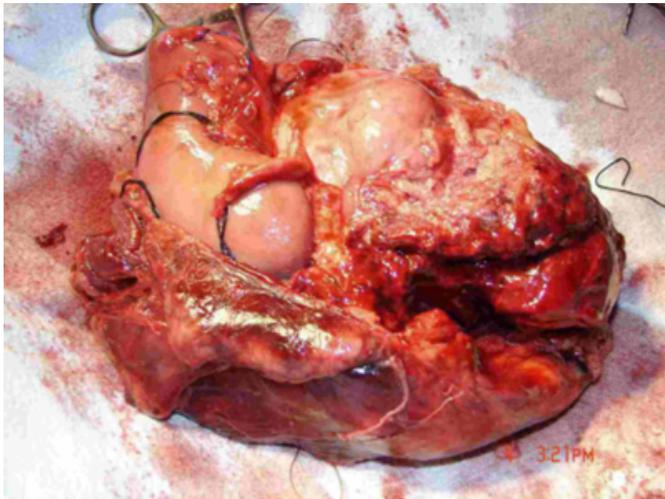


Due to increasing tachycardia and appearance of features of generalized peritonitis an exploratory laparotomy was performed. The operation was performed within 6hrs of presentation. 1.5 litres of intraperitoneal blood was found in the peritoneum. Mobilisation of the hepatic flexure of the colon and Kocherisation of the duodenum revealed a huge haemorrhagic mass involving the kidney. The mass was actively bleeding. The renal vessels were taken under control and a right sided nephrectomy was performed. There was no thrombus in the renal vein and no lesions suggestive of any kind of metastasis involving the peritoneum, liver or abdominal structures were found. The specimen was sent for

histopathological examination.

Figure 2

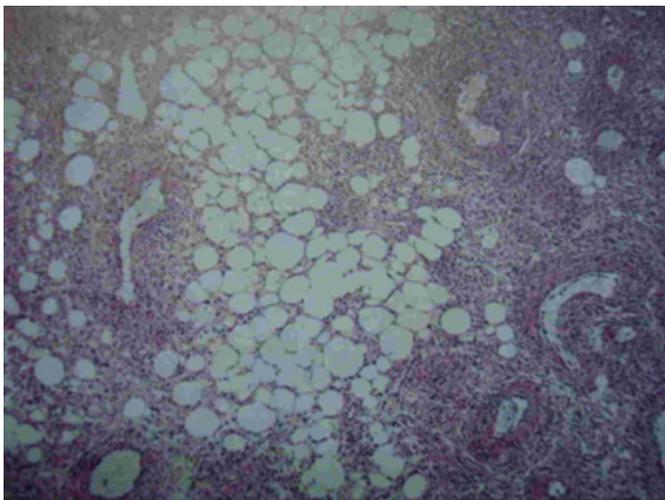
(Fig 2). Kidney with mass



Meanwhile a work up of the patient was done to rule out any signs or symptoms of lung , bone or brain metastasis as is commonly seen in renal cell carcinoma. They revealed no positive finding. A detailed history and examination ruled out the presence of any paraneoplastic manifestations. The biopsy report stated that it was an angiomyolipoma of the kidney.

Figure 3

(Fig 3) Histopathology show angiomyolipoma



Postoperatively the patient made an uneventful recovery. He was screened for tuberous sclerosis but was not found to be suffering from it.

DISCUSSION

Renal AML is a relatively infrequent benign tumour observed in 0.3% of the population and accounts for 3% of

all solid renal masses (1). As many as 25% of patients with renal AML can present with spontaneous rupture and subsequent haemorrhage into the retroperitoneum (2) Massive retroperitoneal hemorrhage It is known that spontaneous perirenal haemorrhage is from AML also known as Wunderlich’s syndrome has been found in up to 10% of patients and represents the most significant and feared complication(3,4) often a surgical emergency necessitating great efforts in terms of diagnosis and treatment (5) as we faced here. However only 6 cases of spontaneous rupture in the peritoneal cavity have been reported in medical literature(6)

There are two types of angiomyolipomas -- one associated with tuberous sclerosis and the other is known as isolated angiomyolipoma. AML is found in approximately 20 % of patients with tuberous sclerosis and are frequently bilateral and asymptomatic(7).But in patients without tuberous sclerosis renal AML can be unilateral and tend to be larger than those associated with tuberous sclerosis.(8) This case is an example of a huge AML not associated with tuberous sclerosis.

AML is diagnosed commonly by CT scan in which the high fat content present in the lesion makes it pathognomonic(9). In this case the large amount of blood and necrosis present surrounding the lesion made a preoperative diagnosis of the presence of a ruptured AML difficult.

The patient had presented to us with hypovolemic shock but preoperatively there was no clear reason as to why. He did not have a history of an obvious trauma that could cause intraabdominal bleeding nor was there any history of a predisposing diseased organ system. When the CT scan revealed the renal laceration it still seemed incongruent to the history although it explained his symptoms.Exploration revealed a haemorrhagic mass and it was apparent that renal tumor had ruptured. Renal cell carcinoma is the commonest renal mass but a presentation as spontaneous rupture is rare(10) AML can rupture but intra peritoneal rupture is extremely rare. While no intervention is required for tumours <4 cm(11) nephrectomy seems to be is the only option in ruptures of such large tumours(6)

We present this case as it can teach us mainly two things: firstly a patient can present with an acute abdomen due to rupture of renal tumour and secondly renal AML not related to tuberous sclerosis can present with a spontaneous rupture in the peritoneum not just retroperitoneum.

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Author Information

Chandan Roy Choudhury, MS

Department of Surgery Medical College, Kolkata

Krishnendu Maiti, MS MRCS

Department of Surgery Medical College, Kolkata

Saurav Chakravartty, MBBS

Department of Surgery Medical College, Kolkata

Saptarshi Bhattacharya, MS

Department of Surgery Medical College, Kolkata