

permission to use the flow cytometer for immunophenotyping and the staff of the Clinical Diagnostic Laboratory, University Hospital, KL for the

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Spontaneous Rupture of Renal Angiomyolipoma Presenting as Acute Abdomen

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Summary

Five cases of renal angiomyolipoma which underwent spontaneous rupture are described. These patients presented as an "acute abdomen" for which the diagnosis was not initially apparent. A high index of suspicion is required to make the diagnosis even with modern imaging techniques. The treatment of these tumours is discussed.

Key Words: Renal angiomyolipoma, Spontaneous rupture

Introduction

Renal angiomyolipomas are uncommon hamartomas of the kidney which have a propensity for bleeding. These patients often present acutely as a result of the haemorrhage which may be life-threatening. We report five patients who presented with an acute abdomen and discuss the role of radiological imaging in reaching the diagnosis.

Case 1

The patient was a 48-year lady known to have tuberous sclerosis. She presented with the sudden onset of right sided abdominal pain and swelling. On examination she was pale and tachycardic with a blood pressure of 90/40mmHg. Abdominal examination revealed a large tender mass in her right flank. Her haemoglobin was 9g/dl on admission and she was treated conservatively with blood

transfusion. CT scan of her abdomen showed bilateral angiomyolipomas with evidence of a recent haemorrhage around the right kidney (Fig. 1). Two days later she became hypotensive again and underwent an emergency laparotomy and right nephrectomy. Bleeding from her right kidney was torrential and she succumbed from hypovolemia. Histology of the right kidney confirmed angiomyolipoma.

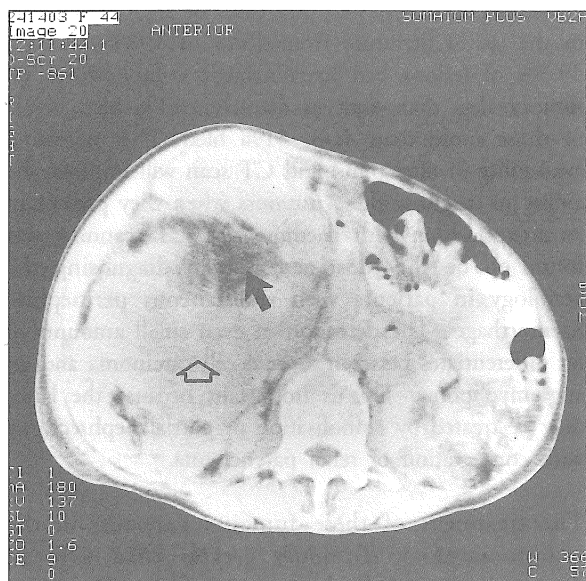


Fig. 1: CT scan of the abdomen showing fat within the right renal angiomyolipoma (black arrow) and the large surrounding haematoma (white arrow)

Case 2

A 50-year male presented to Casualty with the acute onset of right loin pain. There was no history of haematuria of calculous disease. Physical examination revealed an obese gentleman with normal cardiovascular parameters. His abdomen was tender in the right flank but no masses were palpable. Urinalysis showed 20 red blood cells per high power field and a plain abdominal X-ray was normal. He was treated with analgesics with a provisional diagnosis of renal colic and subsequently had an IVU. This showed a mass lesion in the right kidney and a subsequent ultrasound and CT scan showed a 9cm diameter tumour with solid areas and areas of fat density, suggestive of an angiomyolipoma with recent haemorrhage (Fig. 2). He refused all further treatment.

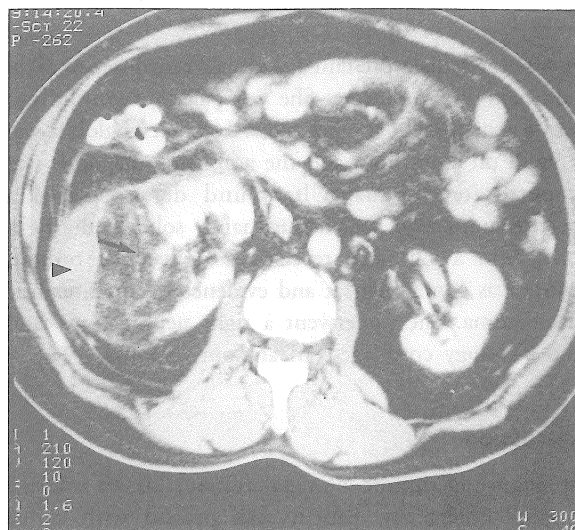


Fig. 2: CT scan of the abdomen showing areas with fat density (black arrow) and perinephric haematoma (arrowhead)

Case 3

A 72-year lady presented to another hospital with right loin pain and postural dizziness. She was found to have a right flank mass and was referred as a colonic tumour. On examination she was pale with a haemoglobin of 3g/dl, and on abdominal examination was found to have a large bimanually palpable mass in her right flank. An urgent ultrasound scan and CT scan showed a large tumour in the right kidney with surrounding haemorrhage. She underwent an emergency laparotomy and right nephrectomy. Histology confirmed an angiomyolipoma and the patient recovered uneventfully.

Case 4

A 50-year lady presented to another hospital with left loin pain of sudden onset. She was treated on clinical grounds as having renal colic but as the pain persisted, she decided to seek a second opinion. An ultrasound scan showed bilateral small tumours in the kidneys with high echogenicity, and a CT scan of the kidneys confirmed the presence of fat in these tumours and haemorrhage from a lesion in the lower pole of the left kidney. In view of the presence of bilateral tumours, an attempt was made to selectively embolise the tumour which had bled but this was unsuccessful. She subsequently underwent a left lower pole nephrectomy and recovered uneventfully.

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Case 5

A 40-year female presented with sudden onset of right loin pain and vomiting. She was pale, tachycardic and had a tender right lumbar mass. Her haemoglobin was 7.9g/dl on admission, and the admitting diagnosis was a perforated viscus. Ultrasound demonstrated a heterogenous mass which was mainly solid in the right flank and a CT scan revealed a right renal tumour with areas containing fat and evidence of a perinephric haematoma. She underwent a right nephrectomy and the histology confirmed an angiomyolipoma.

Discussion

Spontaneous rupture of renal tumours is an unusual surgical emergency. Patients can present with varied symptoms from acute pain mimicking renal colic to life threatening retroperitoneal haemorrhage. In the absence of urological symptoms, the diagnosis may not be made preoperatively, but this is now much less common as ultrasound and CT scanning are becoming more widely used in the assessment of the acute abdomen. Most patients will demonstrate all or some of the symptoms of acute flank pain, lumbar tenderness and signs of internal haemorrhage. The presence of haematuria may help to localise the cause of the acute abdomen to the urinary tract but this is not always present. A high index of suspicion is therefore required to diagnose this condition. The tumours most often associated with spontaneous rupture are renal carcinomas and angiomyolipomas. The latter seems to be much more commonly the cause in Orientals¹. All our patients with spontaneous rupture were Orientals with angiomyolipomas. This is a hamartomatous lesion comprising of fat, blood vessels and smooth muscle elements in varying proportions.

Two forms of this tumour are recognised clinically: the first associated with tuberous sclerosis where the tumours are usually multiple and bilateral with an equal sex distribution, and the second a sporadic form which is usually single and more commonly found in women. Eighty per cent of angiomyolipomas are sporadic.

Oesterling *et al* reviewed 602 cases of angiomyolipoma in the world literature from 1948 to 1985². Overall, 39.5% of lesions had haemorrhagic complications; of tumours less than 4cm in diameter, 13% bled, while of those more than 4cm, 51% bled. The increasing availability of ultrasound and CT scan will improve the ability to diagnose these tumours when they present in an acute fashion with haemorrhage. CT scanning was found to be the most sensitive in diagnosing the aetiology in patients with spontaneous perinephric haemorrhage³. The detection of even small amounts of fat differentiates between a renal cell carcinoma and an angiomyolipoma. This is important because the latter may be treated by embolisation or partial nephrectomy with conservation of renal parenchyma.

The treatment of these tumours depends on the symptomatology. Tumours greater than 4cm in diameter are more likely to be symptomatic and to bleed. We feel that all tumours which are symptomatic especially those with evidence of previous haemorrhage, should be treated aggressively, as a second haemorrhage may be fatal. Selective embolisation should be considered as the first choice of treatment, but if unavailable, nephrectomy should be performed. If the tumours are located at one pole, a partial nephrectomy conserving as much renal tissue as possible should be done, especially if the tumours are bilateral.

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