

Giant Angiomyolipoma of the Kidney with Perinephric Extension: A Rare Case

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ABSTRACT

Angiomyolipomas (AMLs) are one group of benign tumours that are distinguishable radiologically. Because AMLs are composed of different tissues which include fat, muscle, vascular elements,

and even cartilage, the fat in particular, may be detected radiologically. Angiomyolipomas, particularly when they are small, warrant no treatment. They are resected only when they are over 4 cm or when they are symptomatic due to the risk of bleeding.

Key Words: Giant angiomyolipoma, Hamartoma, Fat attenuation, Computed Tomography, MRI

INTRODUCTION

AMLs are hamartomatous tumours which are composed of mature adipose tissue, thick-walled blood vessels, and sheets of smooth muscle. The amount of each component varies in each tumour. When an incidental solid renal mass is encountered, angiomyolipoma should be excluded. The imaging features are quite characteristic, that clinches the diagnosis. We are reporting a rare case of giant angiomyolipoma which extended into the perinephric space.

CASE REPORT

A 28-year old female presented to our department with the complaint of a gradually formed, painless abdominal swelling which involved the right lower abdomen, of a duration of about 25 days. Her vitals were stable and her physical examination revealed a large, firm, non tender swelling in the right lumbar region, which extended to the right iliac fossa.

Laboratory investigations revealed the following results: her routine serological tests showed an Hb% of 11.3gm, ESR of 3mm/Hr, RBCs-5.1 Million/Cc, WBCs-7400cells, Rbs-91gm, urea-25mg/dl, creatinine - 1.4 mg/dl, Na+ -142 (mEq/L), K+ -4.4 (mEq/L), chloride-102 (mEq/L) and Ca+ 9gm. The routine urine studies and microscopy were within normal limits. Her chest radiograph showed a normal picture.

Ultrasound of the abdomen revealed a hyperechoic lesion which extended from the right hypochondrium to the right iliac fossa, which displaced the kidney medially. Multiple small hyperchoic lesions were noted in the cortices of both the kidneys.

Contrast enhanced CT scan showed a predominant fat attenuation lesion which arose from the postero-lateral cortex of the right kidney, extending from the right hypochondrium to the right iliac fossa and displacing the kidney medially. Large vessels of ~ 7-8mm calibre were found to traverse the lesion.

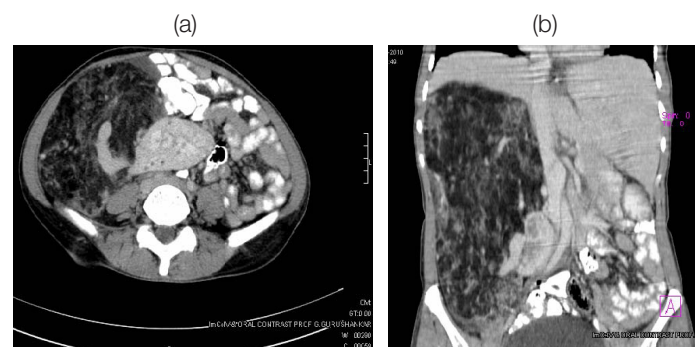
On MRI, the lesion showed hyperintensity on both the T1 and the T2 sequences and it showed signal suppression on the STIR sequence.

The intraoperative findings revealed a well encapsulated tumour which was adherent to the right kidney. The patient underwent

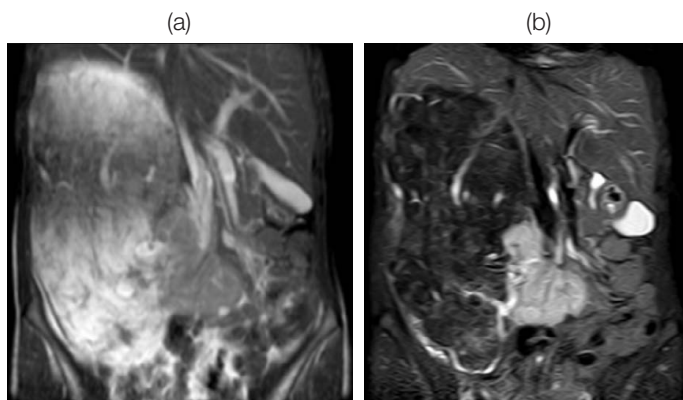
right nephrectomy with mass excision. The mass lesion measured 20(CC) x 17(TR) x 13(AP) cms in size and it weighed approximately 1500 gms. The cut section of the tumour mass was solid and it appeared bright yellow in colour. It contained prominent large blood vessels with no secondary changes. Histopathology and haemoxilyn - eosin staining revealed a non -capsulated, well margined lesion with abundant adipocytes, with smooth muscle and blood vessel components.



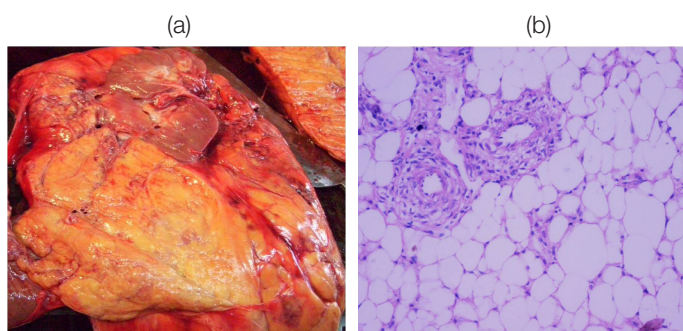
[Table/Fig-1]: Ultrasound image of the abdomen showing an ill defined hyperechoic lesion arising from postero-lateral cortex of right kidney displacing the kidney medially.



[Table/Fig-2]: Contrast enhanced CT scan (a) axial section and (b) coronal reconstruction showed a predominant fat attenuation lesion arising from the postero-lateral cortex of right kidney, extending from the right hypochondrium to right iliac fossa displacing the kidney medially. Large vessels of ~ 7-8mm caliber were seen traversing the lesion.



[Table/Fig-3]: Coronal T2WI MRI (a) shows a well defined hyperintense lesion arising from the postero-lateral cortex of right kidney, extending from the right hypochondrium to right iliac fossa displacing the kidney medially. STIR coronal MRI with contrast (b) lesion shows signal suppression confirming the fat content. Multiple blood vessels are seen to course through the lesion.



[Table/Fig-4]: Post nephrectomy cut section of the gross specimen (a) shows the large yellow colored fatty lesion in which the small right kidney is embedded superiorly and to the right; Microscopy with Haematoxylin & Eosin staining of the specimen shows multiple thick blood walled vessels, abundance of fat globules and a few spindle shaped cells representing the angio, lipo and myo components of the AML. No atypical and malignant features were identified

DISCUSSION

Angiomyolipomas are the most frequent mesenchymal benign neoplasms of the kidney, with a prevalence of 0.3–3%. They occur more commonly in women than in men. Angiomyolipomas, particularly when they are small, warrant no treatment, while they are resected only when they are over 4 cm or symptomatic, due to the risk of bleeding.

Angiomyolipomas contain smooth muscle and vascular, lipomatous, and myeloid elements in different proportions. When an incidental solid renal mass is encountered, angiomyolipoma should be excluded [1].

The overall female: male ratio is approximately 4:1, which suggests a hormonal component to the tumour growth [2]. It can occur sporadically, or it can be part of the tuberous sclerosis complex (TSC). Sporadic angiomyolipomas account for 80 percent of the AML, they are usually solitary and they occur almost exclusively in women in the fourth to fifth decades of life (mean age 43 years) [3].

Most of the small angiomyolipomas are asymptomatic and they are found incidentally during radiological studies. Research has shown that 25-50 percent of the patients have some or all of the features and the stigmata of TSC. The classical presentation of AML is a palpable tender mass (47%), flank pain (53%) and gross haematuria (23%); this is known as 'Lenk's triad' [4].

However, it is rare to have a diagnosed AML with this triad. The less frequently associated symptoms include nausea or vomiting, fever, anaemia and blood pressure alteration [5].

IMAGING FEATURES

Small angiomyolipomas (less than 3cm in diameter) appear typically as hyperechoic, sharply marginated, and homogeneous lesions on the gray-scale ultrasound (US) [6]. Large angiomyolipomas (more than 3cm in diameter) may reveal a homogeneous or heterogeneous bright appearance on the US due to solid, adipose and haemorrhagic components [7].

Angiomyolipomas consist of variable quantities of blood vessels, smooth muscle, and fat. Most of the angiomyolipomas can be diagnosed by identifying portions of the mass which have an attenuation of -10 HU or less, which are indicative of fat [8]. However, approximately 4%–5% of the angiomyolipomas may either not contain any fat cells or they may contain an insufficient amount of fat cells to allow a diagnosis which is based on imaging [9,10]. The MR imaging examination should include the application of the T1- and T2-weighted sequences and of a frequency-selective fat-suppressed dynamically acquired T1-weighted sequence before and after intravenous gadolinium chelate administration. The fat suppression techniques generally are unhelpful in detecting fat in the angiomyolipomas with minimal fat, because such masses contain little or no fat and they often appear isointense to the renal parenchyma on the T1-weighted images [9]. If a mass exhibits areas of signal suppression, a fat-containing angiomyolipoma should be considered as a strong possibility. Most of the angiomyolipomas can be diagnosed with confidence by identifying the presence of the fat cells within a noncalcified renal mass [10].

AMLs can vary in size from a few millimeters to larger than 20 cm [11]. It is unusual to see an AML which is larger than 10 cm and therefore many studies have demonstrated that any AML which measures over that size is considered as a "giant".

The two major morbidities which are associated with renal AMLs are retroperitoneal haemorrhage and impingement of the AML on the normal tissue, which can impair the renal function [2,12]. Enlarging angiomyolipomas can develop micro- and macro-aneurysms that can rupture. These can be sudden and painful, and occasionally life-threatening. Several studies have demonstrated that the frequency of the symptoms and the risk of bleeding (rupture) increases with the size of the AMLs. The reported risk of haemorrhage varies markedly, ranging from 3 to 50 percent. In a review on angiomyolipomas which were ≥ 4 cm, 82 to 94 percent were found to be symptomatic and 50 to 60 percent were found to bleed spontaneously [13].

The management of angiomyolipomas historically, has been correlated with their symptoms. Most of the patients with small tumours (less than 4 cm) that tend to be asymptomatic are managed conservatively, with periodic ultrasonography [14]. However, patients with tumours which are larger than 8 cm, generally tend to be asymptomatic. It is these larger tumours that are at a greater risk of spontaneous or traumatic ruptures, resulting in haemorrhagic complications. These patients are therefore treated with angiography and selective arterial embolization as a first line treatment. Partial or radical nephrectomy is indicated if there is persistent haemorrhage, a suspicion of malignancy, or failed embolization.

Atypical angiomyolipomas with an absent fat component are about 5% and they usually appear iso or slightly hyperdense when they are compared to the adjacent kidney on unenhanced CT, due to the smooth muscle components, and the homogeneously enhancing masses after the contrast administration.

A similar pattern may be observed in complicated benign cysts (haemorrhagic, protein-rich or gelatinous), renal metastasis and RCC. The evidence of hyperdense and fat masses in the kidney at the same time is diagnostic of multiple angiomyolipomas [1].

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