
CASE REPORT

Aggressive Renal Angiomyolipoma: Radiological and Pathological Correlation

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ABSTRACT

Renal angiomyolipoma is usually regarded as a benign lesion with no malignant potential. This report is of a patient with aggressive renal angiomyolipoma with renal vein and inferior vena cava invasion in a middle-aged female. The imaging findings and the clinical utility of contrast enhanced multi-slice computed tomography and magnetic resonance imaging are discussed.

Key Words: Angiomyolipoma, Computed tomography, Inferior vena cava, Magnetic resonance imaging, Thrombus

CASE REPORT

A 61-year-old woman presented to the general surgical clinic with a 2-month history of dull right flank pain of insidious onset. There was no history of trauma, dysuria, haematuria, or pyrexia. Her past medical history was unremarkable except for mild hypertension (170/90 mm Hg). Physical examination of the abdomen and pelvis were normal. Routine biochemical blood tests including platelet count, blood clotting profile, and renal function were all normal. Abdominal and pelvic radiographs did not reveal any masses or renal tract calcification. Sonographic examination showed a 2.5 cm highly echogenic, well-circumscribed mass arising from the lower pole of the right kidney with extension into the renal pelvis and right renal vein. A phasic 'waving motion', synchronous with respiratory movement, was demonstrated, suggestive of tumour thrombus within the vein. The left kidney was normal.

As renal cell carcinoma was suspected clinically, the patient underwent further investigation with dynamic 2-phased contrast computed tomography (CT) scan of the kidneys. A solitary well-circumscribed homogenous

hypodense mass with attenuation of -85 HU was found in the medulla of the lower pole of the right kidney, with a lobulated tongue-like extension into the distended lumen of the right renal vein (Figure 1a). Oblique coronal reformatted images along the course of the right renal vein and inferior vena cava demonstrated extension into the infrahepatic inferior vena cava (IVC; Figure 1b). The imaging findings of a fatty tumour were consistent with an angiomyolipoma with extension into the right renal vein and IVC. To clearly delineate the upper extent of the tumour within the IVC, contrast-enhanced magnetic resonance imaging (MRI) was performed. This confirmed the fatty nature of the mass, its extension through the renal vein into the IVC, and the proximal limit of the tumour within the intrahepatic portion of the IVC (Figure 2).

A right radical nephrectomy with cavotomy was performed. A tumour was found within the medulla of the right kidney, extending into the right renal vein and inferior vena cava up to the level of the hepatic vein. The right kidney and tumour thrombus was removed en bloc through a cavotomy. No adhesion to the venous wall was noted intraoperatively. Histological examination of the tumour showed mainly mature adipocyte tissue intermixed with characteristic smooth muscle and vessels, in keeping with the radiological finding of renal angiomyolipoma (Figure 3). There was no histological evidence to suggest malignancy of the tumour. The patient had an uneventful recovery and remained

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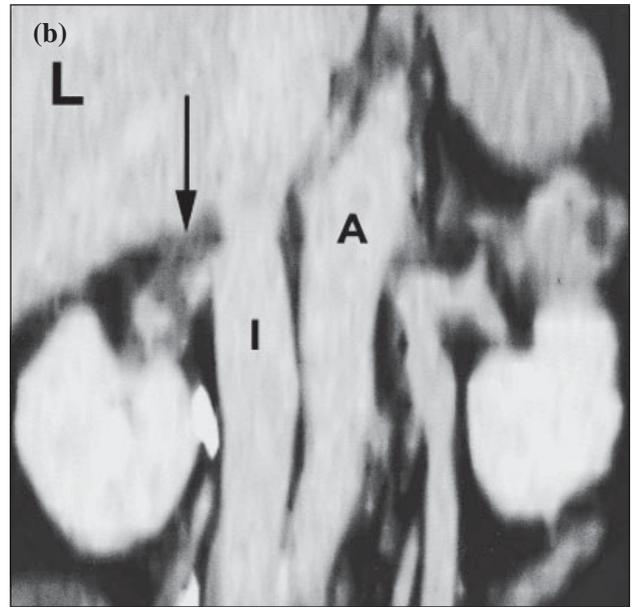
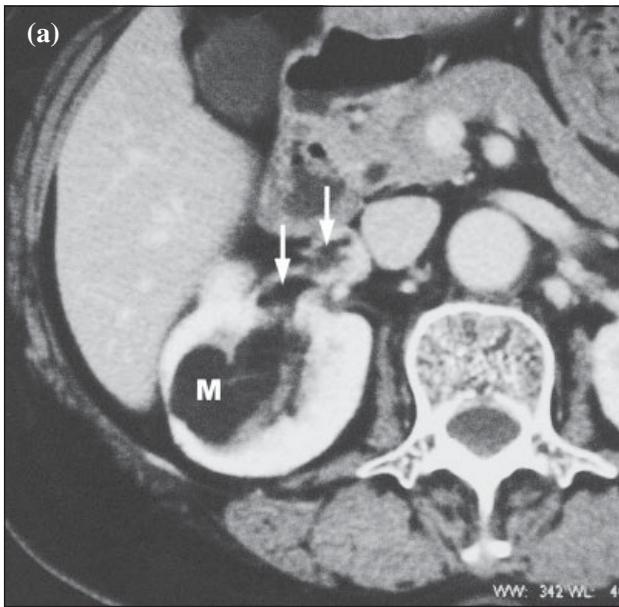


Figure 1. (a) Post-contrast multi-slice computed tomography scan during venous phase demonstrating a hypodense mass (M) arising from the medulla of the right kidney with extension into the distended right renal vein (arrows); (b) oblique coronal reformatted image along the course of right renal vein and inferior vena cava shows tumour extension (arrow) into the inferior vena cava (I) inferior to the liver (L). 'A' denotes the aorta.

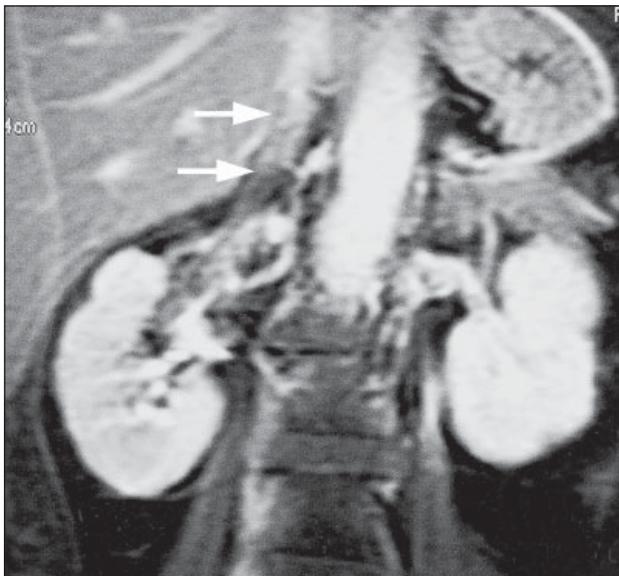


Figure 2. Magnetic resonance angiogram showing the proximal extent of the tumour thrombus (arrows) within the intrahepatic portion of the inferior vena cava.

in good health at follow-up consultation, with no evidence of tumour recurrence noted on computed tomography (CT) scan performed 6 months after surgery.

DISCUSSION

Renal angiomyolipoma is considered a benign hamartomous lesion composed of an admixture of mature adipose tissue, smooth muscle, and abnormal thick-walled blood vessels. Two distinct groups of patients have been identified with this entity. The first group

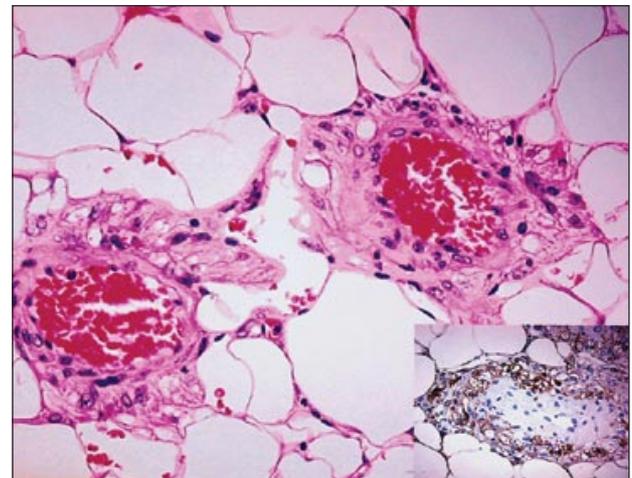


Figure 3. Microscopic appearance of the tumour thrombus showing typical mixture of mature adipocytes with thick-walled vessels surrounded by a collarette of spindle-shaped smooth muscle cells, which stain positive for human melanoma block-45 (insert).

consists of people with tuberous sclerosis complex in whom the angiomyolipoma is a manifestation of this complex neurocutaneous syndrome, which includes epilepsy, mental retardation, and adenoma sebaceum. Tumours in this group are frequently bilateral and multiple, while lesions found in the second group with angiomyolipoma are usually solitary and occur almost exclusively in women in the fourth to fifth decade of life. Seventy five percent of all reported cases of aggressive angiomyolipoma occur as sporadic isolated cases²⁻¹³ with only 25% related to tuberous sclerosis.

Discovery of a renal angiomyolipoma is usually an incidental finding during a sonographic or CT scan for an unrelated clinical indication. Most patients with a small tumour (defined as tumours less than 4.0 cm) tend to be asymptomatic, therefore, a conservative approach is generally advocated.¹ Those with tumours of more than 8 cm have a greater risk of spontaneous or traumatic rupture resulting in haemorrhagic complications.¹

Symptomatic patients with complicated large angiomyolipoma should be treated with nephrectomy, either partial or radical, or by transarterial embolisation. Intravascular extension into venous structures, in particular the renal vein and IVC, demonstrates an 'aggressive' element and distinguishes an angiomyolipoma from the usual cortical renal angiomyolipoma.^{2,3} The first report of such a tumour was in 1982⁴ and 16 patients with angiomyolipoma with intravascular extension have been reported in the world literature to date.²⁻¹³ Fifty percent of patients complained of mild loin pain at the site while the rest complained of other unrelated or minor symptoms such as vague abdominal pain. Half of the patients had a solitary tumour with 70% originating on the right side. There seems to be no relationship between aggressive behaviour and tumour size.^{2,5}

Sonography was the preliminary radiological investigation for 85% of patients, but CT scan confirmed the diagnosis for all patients. The characteristic sonographic findings were of a highly or mixed echogenic renal tumour, with tongue-like extension into the renal vein. As renal cell carcinoma could have a similar sonographic appearance, CT remains the imaging tool of choice for radiological confirmation. The demonstration of a large portion of tissue consistent with fat within the tumour at CT scan is virtually diagnostic of angiomyolipoma. The present case highlights the advantage of dynamic contrast-enhanced CT assessment of this entity, whereby the fatty nature of the tumour is succinctly demonstrated and tumour extension into the renal vein and IVC is visualised. Both the coronal reformatted CT images and the MR angiogram for this patient was extremely helpful to the surgeons for their surgical planning for cavotomy as the cephalic extent of the tumour thrombus was clearly shown, facilitating placement of the

vascular clamp during surgery.⁶ Renal venacavography can be used to confirm the position of the intravenous extension⁷ although this may not be clearly determined. This procedure also carries a potential risk of dislodging the tumour thrombus with catastrophic cardiopulmonary consequences.

To conclude, renal angiomyolipoma can display an apparent aggressive presentation with intravenous invasion into the renal vein and IVC. Such cases should be managed as a separate entity, with prompt surgical intervention. Contrast-enhanced multi-slice CT with reformation or MR imaging will confirm the diagnosis of angiomyolipoma with clear demonstration of the cephalic extent of the tumour thrombus.

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