CASE REPORT

An unusually large renal angiomyolipoma peeping into the right atrium

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SUMMARY

Angiomyolipomas (AMLs) are benign hamartomatous tumours of the kidney that occur sporadically or in association with tuberous sclerosis. Although they are benign lesions, larger tumours can, rarely, behave aggressively and have extrarenal extension. Extension of an AML into the renal veins and inferior vena cava (IVC) is rare. We report a rare case of an AML with extension into the renal vein and IVC up to the right atrium that was successfully managed surgically.

BACKGROUND

Angiomyolipomas (AMLs) are benign hamartomatous tumours of the kidney, with angiomatous, lipomatous and myomatous tissues present in varying amounts. They can occur sporadically (80–90%) or in association with tuberous sclerosis. Syndromic AMLs, seen in up to 80% of patients with tuberous sclerosis, can be multiple and bilateral (15%).¹ Although they are benign lesions, larger tumours can, rarely, behave aggressively and have extrarenal extension.² Extension of an AML into the renal veins and inferior vena cava (IVC) is rare, unlike renal cell carcinomas. We report a rare case of a 50-year-old woman who presented with a large AML of the left kidney, with extension into the renal vein and IVC, up to the right atrium.

CASE PRESENTATION

A 50-year-old woman presented with vague dull aching pain in the left flank for the past 3 months. Her general condition was stable and general examination was unremarkable. On abdominal examination, a vague mass, felt in the left flank, was moving with respiration and ballotable.

INVESTIGATIONS

Laboratory investigations were normal except for mild anaemia. Ultrasonography revealed a large, predominantly echogenic mass lesion arising from the left kidney (figure 1). The left renal vein was expanded with echogenic thrombus. The thrombus was seen extending to the IVC up to the cavoatrial junction. Contrast-enhanced CT (CECT) of the abdomen showed a large (12×10×6 cm) mass arising from the left kidney, encasing the renal hilar vessels and extending to the left renal vein (figure 2). It showed extensive areas of fat attenuation. On MRI, the tumour was seen as hyperintense on T1-weighted and T2-weighted images and showed suppression of signals on sequences with fat saturation indicating macroscopic fat. The tumour showed extrarenal extension to reach the

undersurface of the spleen. There was mild obstructive lower polar caliectasis. There was extension of tumour thrombus with similar signal characteristics into the IVC, up to the cavoatrial junction, and it was seen protruding into the right atrium (figures 3–5). There was no locoregional lymphadenopathy and no distant metastasis. The right kidney was normal.

DIFFERENTIAL DIAGNOSIS

The differential consideration for macroscopic fat-containing tumours of the retroperitoneum include AML, lipoma, liposarcoma, teratoma, renal cell carcinoma and adrenal myelolipoma.

Demonstration of calcification favours other neoplasms over AMLs. Extension of a tumour into the IVC is commonly seen in renal cell carcinoma but can also, rarely, be seen in AMLs, depending on the tumour's histology (vide infra).

Based on the findings mentioned above, a diagnosis of left renal AML with extension into the perinephric space, left renal vein, IVC and right atrium was made.

TREATMENT

The patient underwent left radical nephrectomy with IVC thrombectomy (figure 6). The post-operative period was uneventful.

OUTCOME AND FOLLOW-UP

Subsequent histopathology revealed a triphasic tumour comprised of a mixture of adipocytes, thick blood vessels and smooth muscles, consistent with AML. During a 6-month follow-up on ultrasound and CECT, no residual/recurrent lesion was present.

DISCUSSION

AML is a benign hamartomatous renal tumour with a higher prevalence in middle-aged women.³ Most of these tumours are asymptomatic and detected incidentally. Larger tumours (>4 cm) can be symptomatic with flank pain and haematuria, or following retroperitoneal haemorrhage from intratumoural vessels. There are two types of AMLs: common, classic (triphasic) and rare epithelioid. Epithelioid variants of AML can be aggressive, with local extension, distant metastasis, higher recurrence and mortality.^{4 5}

Classical AMLs, rarely, show extension into the renal vein or outside the renal capsule, especially when they are large. An extensive literature review by Riviere *et al*³ showed 44 cases of AMLs with renal vein or IVC extension in which almost all the

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Figure 1 (A) Ultrasonography showing poorly defined echogenic mass arising from left kidney with caliectasis in its residual lower pole; (B and C) showing echogenic tumour in inferior vena cava (arrow) extending to right atrium (star).

Figure 2 Axial sections (A and B) of non-contrast CT of upper abdomen showing large mass with fat attenuation (star) arising from left kidney with tumour thrombus in inferior vena cava (arrow).

Figure 3 MR images in coronal plane: T2W (A), T1W (B) and T1W fat suppressed (C) images showing fat containing mass (star) arising from left kidney with thrombus of same intensity in IVC (arrow). IVC, inferior vena cava; T1W, T1-weighted images; T2W, T2-weighted images.

Figure 4 MRI T2W image in axial plane: (A) left kidney mass with extension into left renal vein (arrow) and (B) with thrombus of same intensity in IVC (arrow). IVC, inferior vena cava; T2W, T2-weighted images.

Figure 5 MRI, T2W image in sagittal plane: (A) left kidney mass causing hydronephrosis in residual kidney (star) and (B) with thrombus of same intensity in IVC reaching right atrium (arrow). IVC, inferior vena cava; T2W, T2-weighted images.

Figure 6 Gross specimen showing nephrectomy specimen with removed thrombus from IVC and right atrium. IVC, inferior vena cava.

lesions were larger than 4 cm. Similarly, the tumour in our case was large.

The extension of the tumour into the renal vein and IVC necessitate tumour thrombectomy along with nephrectomy. Thoracotomy is needed when the tumour reaches the atrium. Moreover, the higher chances of tumour thromboembolism into the pulmonary bed necessitate close observation and early surgery in these patients.

Imaging plays a crucial role in characterising the tumour as well as revealing its local extent. AML is seen as a well-defined echogenic mass on sonography. The echogenicity of the tumour is because of its fat content and multiple acoustic interfaces from numerous inner vessels. The tumour thrombus also shows a similar appearance. On CT, the tumour shows areas of fat attenuation (-10 HU or lower). On CT scan, presence of haemorrhage can obscure the fat component in larger tumours. MRI also can detect the presence of fat in these tumours. Fat suppression techniques such as chemical saturation and inversion recovery can show the lipid components even with the presence

of haemorrhage. MRI can show the abnormal tumoural vessels and helps in accurately delineating the local extent. Multiplanar reformatted images in various planes show the cephalic extent of IVC tumour thrombus precisely. The fat component is not detectable in ~5% of AMLs and it is hence difficult to differentiate these from other neoplasms. To conclude, different imaging modalities, acting complementary to each other, play a

pivotal role in the detection, diagnosis, management planning and preoperative preparation in patients with AML. Aggressive tumours with IVC and right atrial extension require close observation and surgical removal after proper preoperative planning.

Competing interests None declared.

Patient consent Obtained.

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Learning points

- ▶ The differential diagnoses of fat-containing lesions involving the renal fossa include angiomyolipoma, lipoma, adrenal myelolipoma and retroperitoneal liposarcoma.
- ▶ Presence of macroscopic fat in the absence of calcification in a renal mass is virtually diagnostic of angiomyolipoma.
- ▶ Inferior vena cava and perinephric extension may, rarely, occur in angiomyolipomas, necessitating extension of surgical technique.

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