Aggressive renal angiomyolipoma with renal vein and inferior vena cava thrombus: a case report

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Case presentation

A 58-year-old Chinese woman with good past health presented with a 2-day history of right lower abdominal pain. Routine biochemical blood tests revealed a white blood cell count of 9.1 × 10^9/L and normal renal function (creatinine level: 49 μmol/L). An urgent contrast-enhanced computed tomography scan revealed acute diverticulitis in the caecum. Incidentally, a solitary well-defined homogenous fat-density mass with unenhanced density of -63 Hounsfield units was evident at the right renal sinus, with extension into the right renal vein and inferior vena cava (IVC) [Fig 1]. Findings were suggestive of an aggressive renal angiomyolipoma (AML) with right renal vein and IVC thrombus. The patient was referred to an urologist. Subsequent robotic-assisted laparoscopic radical nephrectomy and IVC thrombectomy were performed uneventfully.

The surgical specimen provided by the pathologist demonstrated the right renal vein depicted as a purple red strip and fatty tumour (Fig 2a). Microscopic examination revealed all features of a typical AML including mature adipocytes, myoid spindle cells, and thick-walled blood vessels (Fig 2b). It was also positive on Melan A (Fig 2c) and HMB-45 stains (Fig 2d).

Discussion

Angiomyolipoma is a common benign renal mesenchymal neoplasm composed in variable proportions of adipose tissue, smooth muscle cells, and abnormal vessels. It is most commonly sporadic (80%) or associated with genetic syndromes such as tuberous sclerosis.[1] A renal mass with visualisation of macroscopic fat density on computed tomography is virtually diagnostic of AML. Therefore, visualisation of fat density in the intravascular compartment is indicative of tumour invasion.

Cases of aggressive AML with renal vein and IVC thrombus have been reported. A literature review[2] of 26 cases of invasive renal AML reported that a large size and right-sided location of the tumour may be contributing factors in AML with intravascular invasion. This may be related to the shorter and straighter course of the right renal vein. Two variants of renal AML, namely, classic and

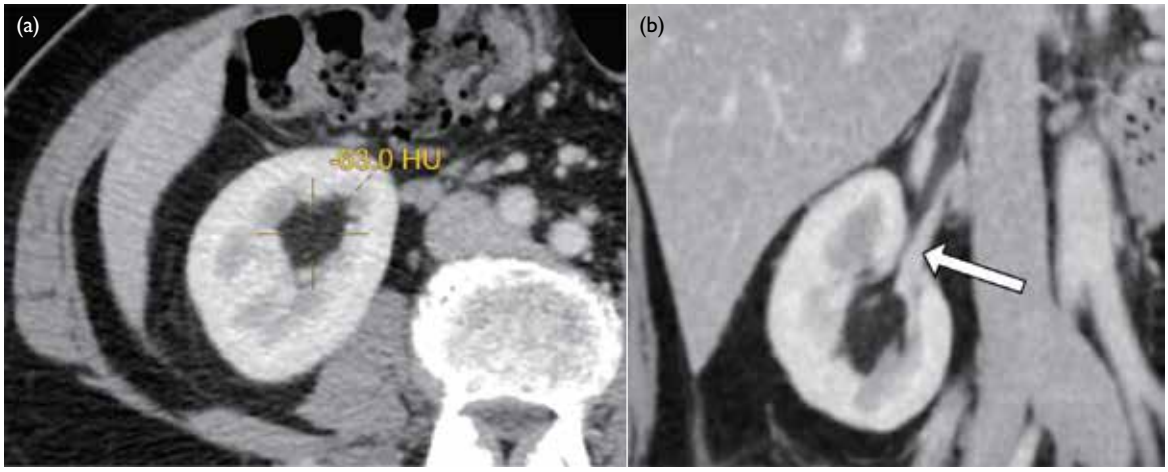

FIG 1. (a) Axial and (b) sagittal contrast-enhanced computed tomography images of the patient show a homogeneous fat-density mass of -63 Hounsfield units at the right renal sinus with extension into the right renal vein and inferior vena cava (arrow in [b])

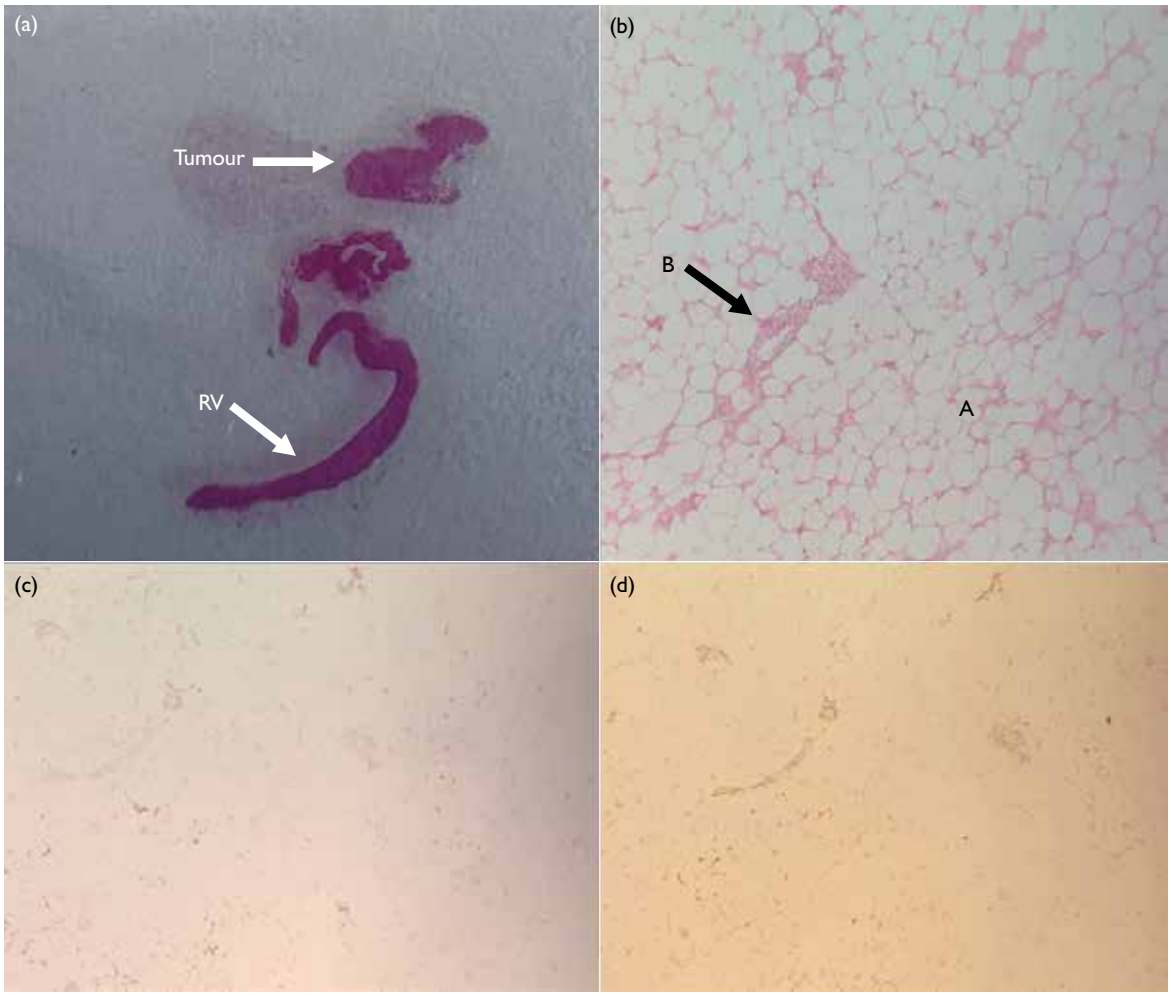

FIG 2. (a) The surgical specimen of the patient on glass slide (haematoxylin and eosin [H&E] staining, not under microscope) includes the renal vein (RV) and fatty tumour (Tumour). (b) The high-powered H&E stained slide (×100) shows the tumour containing all features of angiomyolipoma including mature adipocytes (A), myoid spindle cells, and thick-walled blood vessels (B). Immunohistochemical staining shows tumour cells positive for Melan A (c) and HMB-45 (d) [both ×100]

epithelioid, have been described. The epithelioid variant has been reported to exhibit aggressive behaviour with IVC thrombus.[3] In this case, the patient had a classic subtype of renal AML with mature adipocytes (Fig 2b) and no epithelioid cells.

The optimal treatment is robotic nephrectomy and IVC thrombectomy irrespective of tumour size, since IVC thrombus can be life threatening with higher risks of vessel occlusion and tumour embolus. A multi-institutional study in 2016 retrospectively reviewed 32 cases of robotic radical nephrectomy and IVC thrombectomy and demonstrated a favourable outcome with adequate robotic experience including less blood loss, earlier patient discharge, and fewer complications compared with open nephrectomy.[4] Inferior vena cava thrombectomy follows similar surgical principles to that for renal cell carcinoma with IVC thrombus. Proper visualisation and mobilisation of the kidney, renal vein and IVC allow selective vascular clamping, namely, the infrarenal and suprarenal IVC as well as the renal vein. The IVC is then exposed and the tumour is dissected away from the IVC. The cavotomy is then closed and nephrectomy is performed.

We describe a classic subtype of renal AML extending into the renal vein and IVC, managed by robotic-assisted laparoscopic nephrectomy and IVC thrombectomy. Computed tomography and other imaging modalities are essential to diagnose AML and for proper surgical planning. With proper visualisation of the kidney, renal vein and IVC, a safe and successful outcome of robotic nephrectomy and IVC thrombectomy is achieved.

Author contributions

Concept or design: PHW Lo, HM Kwok, FH Ng, JKF Ma.
Acquisition of data: PHW Lo, HM Kwok, FH Ng, HY Lo.
Analysis or interpretation of data: PHW Lo, HM Kwok,

HY Lo.
Drafting of the manuscript: All authors.
Critical revision of the manuscript for important intellectual content: All authors.

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

All authors have disclosed no conflicts of interest.

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Ethics approval

This study was approved by the Kowloon West Cluster Research Ethics Committee of Hospital Authority, Hong Kong [Ref No.: KW/EX-23-010(180-06)]. The patient has given a consent statement for publication of this case report.

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