CASE REPORT

Benevolent Renal Angiomyolipoma with Intra-cardiac Extension—A Challenge in Diagnosis and Management

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Abstract While intra-caval and intra-cardiac extension of retroperitoneal tumors is extremely rare, it is almost unheard-of in benign tumors. We report the challenges in diagnosis and management of the first case of a renal angiomyolipoma (AML) with intra-ventricular extension in a young man who presented with pain and a lump in the abdomen.

Keywords Angiomyolipoma · Renal tumors · Intra-cardiac extension of renal tumor · Intra-cardiac extension of renal angiomyolipoma

Introduction

Angiomyolipoma is a benign tumor of the kidney classified under *PEComas* (*perivascular epitheloid cell*). They exhibit a characteristic presence of a perivascular epitheloid cell with mature adipocytes and thick-walled blood vessels [1]. They are more common in females due to hormonal influences [2]. They can range from microscopic lesions to very large tumors

with extension into the inferior vena cava (IVC) and heart [3]. Such extensive lesions pose diagnostic challenges; diligent preoperative work-up and multi-disciplinary approach in management yield best results.

Case Report

A 27-year-old young man presented with dull aching pain along with a slowly progressing lump in the upper abdomen for the last 4 years. There was no family history of tumors or tuberous sclerosis. Physical examination revealed a large lump in the right lumbar region extending into the right hypochondrium and epigastrium. A pansystolic grade III/VI murmur was audible in the tricuspid area. Laboratory investigations showed thrombocytopenia and polycythemia. Ultrasound revealed a hyperechoic mass related to the right kidney and tumor thrombus in the inferior vena cava (IVC). Computed tomography (CT) revealed fat attenuating mass lesion of 18 × 14 cm in the retroperitoneum, infiltrating into the renal hilum and extending through the renal vein into the IVC as well as reaching up to the right atrium and right ventricle (Fig. 1). Echocardiogram revealed a tumor thrombus extending into the right atrium and ventricle. CT-guided biopsy revealed fibro-adipose tissue without atypical cells.

The patient was planned for combined simultaneous mid-line laparotomy and sternotomy on cardiopulmonary bypass on *beating-heart* for *en masse* excision of the tumor and tumor thrombectomy. An intraoperative trans-esophageal echocardiography (TEE) confirmed the findings.

Under general anesthesia, a right femoral cannulation was performed for *lower* body drainage. The operation was started with a laparotomy immediately followed by a sternotomy along with an aortic and superior vena cava cannulation for *upper* body drainage.

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Fig. 1 Contrast-enhanced computed tomography. **a** Sagittal view showing the tumor thrombus extending into the IVC, right atrium, and right ventricle. **b** Coronal and **c** axial images showing tumor extent and infiltration

Intraoperatively, a $24 \times 17 \times 10$ -cm yellowish-pink tumor was seen in the right renal area and the kidney was not seen separately from the tumor. The IVC and right renal vein were distended with the tumor thrombus which was also felt in the right atrium and ventricle.

First, the abdominal tumor was partially mobilized to isolate the right renal vein, which was looped. The left renal vein, as well as the infrarenal IVC, was also looped for vascular control. Next, a right atriotomy was performed that revealed a dumb bell-shaped tumor in the right atrium extending into the right ventricle (Fig. 2a). The tumor thrombus was free

from the IVC vessel wall all through. The tumor was extracted in two portions—the *first portion* through the right atrium with its neck fractured at just beyond the IVC opening into the right atrium. The *second portion* of the tumor thrombus was delivered through an IVC venotomy *en masse* with the tumor proper (Fig. 2b). The IVC venotomy was repaired with a *pericardial patch*. Postcardiac pulmonary bypass TEE revealed no tricuspid valve regurgitation and the valve was coapting well. Postoperatively, the patient had an uneventful recovery and his hemoglobin and platelet count normalized shortly.

Fig. 2 **a** Large tumor thrombus visualized after cardiopulmonary bypass and right atriotomy. **b** Resected specimen showing the tumor proper along with the kidney and intra-caval (black arrow) and intra-cardiac

parts (white arrow). **c** Histopathological appearance (hematoxylin and eosin stain) suggestive of *classical variant* of angiomylipoma

Histopathology revealed a well-encapsulated classical variant of angiomyolipoma (AML) arising from the lower pole of the right kidney free from the adrenal gland and the thrombus also comprised of same tumor (Fig. 2c). At 9 months follow-up, patient is asymptomatic, with no recurrence on CT scan.

Discussion

Tumors with propensity to extend into the IVC and heart are usually of renal or adrenal origin, only rarely are these retroperitoneal sarcomas. Angiomyolipoma of the kidney is a benign mesenchymal tumor. It may be *sporadic* or *familial* (associated with tuberous sclerosis) in origin [1]. An *epitheloid variant* of AML is known to be more aggressive with malignant transformation and high recurrence rates after resection [4]. The *classical variant* of AML is far more common and usually benign with only rare cases of malignant transformation [5].

At presentation, most of the patients (90 %) of renal AML are asymptomatic. When present, pain and hematuria are common complaints. Life-threatening retroperitoneal hemorrhage (*Wunderlich syndrome*) is extremely rare (0.4 %), but risk is higher in larger tumors [6, 7].

Computer tomography or magnetic resonance imaging are the investigations of choice which typically show the presence of fat within the lesions. Renal cell carcinoma and retroperitoneal liposarcomas can have a similar radiological picture. MR diffuse tensor imaging (MR-DTI) may help to differentiate between a clear cell renal cell carcinoma from a low fat-containing AML [8].

The *classical variant* of AML rarely extends into the IVC, right atrium, and surrounding lymph nodes, which is probably due to its multifocal origin rather than by vascular invasion [1]. Riviere et al. in their review found that among patients with AML, 44 had IVC extension and most of them had large tumors (>4 cm) at presentation and more than 67 % of patients were symptomatic. The median age of presentation was

46.6 years, and only seven patients had right atrial extension and all of them were female [3]. Our patient is the only male with intra-cardiac extension and much younger compared to other patients and the only patient with intra-ventricular extension of renal AML.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interests.

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