Case Report

Giant renal Angiomyolipoma masquerading as a Wilms tumor

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ABSTRACT

Renal Angiomyolipoma (AML) is not commonly seen in the pediatric age group other than patients of tuberous sclerosis where in they can have renal AMLs within the first decade with bilateral in involvement. Diagnosis of renal AML can generally be made by the current radiological modalities in the appropriate clinical setting, but it can be mistaken for other tumors when it is large and has low-fat content. Herein we report a case of giant renal AML that was initially misdiagnosed as a Wilms tumor in a 12-year-old girl.

INTRODUCTION

Angiomyolipoma (AML) is the most common benign solid tumor of the kidney. They have an admixture of vascular, muscle, and fat components. AML occur sporadically in adult females preferentially.[1] In 20% cases, they may present with bilateral AML at an early age in patients who have tuberous sclerosis, VonHippel-Lindau syndrome and Neurofibromatosis type 1 (phakomatosis).[1] With this in mind, we present a case which was initially diagnosed as a Wilms tumor but was finally found to be a renal AML in a child without underlying phakomatosis.

CASE REPORT

A 12-year-girl was referred from another hospital with chief complaints of a lump in the left side of abdomen for the last 5 months. The mass had progressively increased in size over a period of 5 months to involve most of her abdomen and produced significant abdominal distension. There were no other complaints. An initial evaluation with ultrasonography and contrast-enhanced computed tomography (CECT) of abdomen and chest at the referring center showed that it was a left renal mass. Subsequently, she was started on chemotherapy for Wilms tumor with 3 drugs (vincristine, doxorubicin and actinomycin D). After receiving "week 6" of chemotherapy, she presented to us. At the time of presentation to us, she had a huge abdominal lump that was occupying the entire abdomen. The size of the mass had decreased subjectively as per the patient herself by 25% after initiation of chemotherapy. The initial CECT was reviewed at our center. It was found to be having a variegated appearance arising from the lower pole of the left kidney and occupying almost the whole of abdomen. No evidence of calcification or fat was identified. We repeated the CECT abdomen and chest which revealed a 21 cm × 17 cm × 12 cm solid mass with necrosis within it and this was arising from the left kidney [Figure 1].

There was no intravascular extension or lung metastasis. Fine-needle aspiration cytology (FNAC) was performed, but the aspirate contained only blood. Considering the rapid increase in size of the mass and the partial response to chemotherapy and the related necrosis, a repeat FNAC was

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Quick Response Code:	Website: www.indianjurol.com
	DOI: 10.4103/0970-1591.197325

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Received: 14.05.2016, **Accepted:** 12.10.2016

Financial support and sponsorship: Nil.

Conflicts of interest: There are no conflicts of interest.

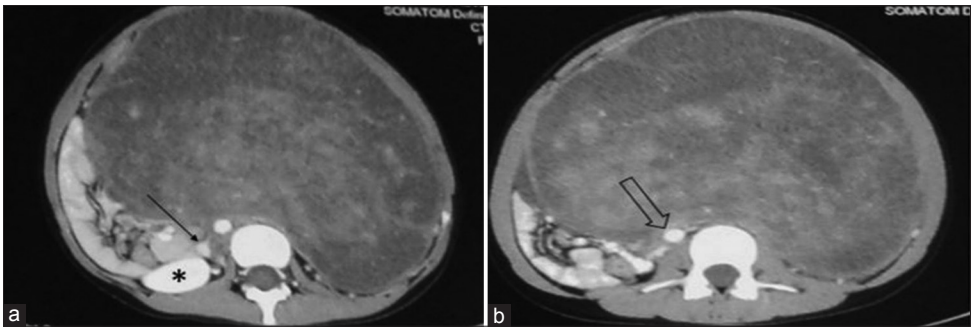

Figure 1: (a and b) Contrast-enhanced computed tomography scan images in the axial plane showing a large heterogeneous mass lesion arising from the left side of retroperitoneum replacing the entire left kidney. The mass was displacing the bowel loops, aorta (outlined arrow), and inferior vena cava (arrow) to the opposite side. There was no evidence of thrombus seen in the inferior vena cava. The right kidney (*) was normal and located in the right renal fossa

not ordered, and a working diagnosis of Wilms tumor was kept. She was administered 2 more weeks of chemotherapy and then taken up for resection.

The operative findings confirmed that the mass was arising from the lower pole of the left kidney, and it was occupying the entire abdomen and pushing the peritoneal contents to the right paracolic gutter [Figure 2a]. The contralateral kidney and other organs were normal. A standard left nephroureterectomy with lymph node sampling was performed. The specimen measured 27 cm × 22 cm × 18 cm and weighed 5.5 kg [Figure 2b].

Postoperative recovery was uneventful. The histopathological report was suggestive of AML. Immunohistochemical analysis on the specimen showed negative pattern for WT-1, MIC-2, desmin, NSE, synaptophysin, S-100 andchromogranin. There was a positive reaction towards HMB45 [Figure 3] and trace positivity towards vimentin. Consultation from the Departments of Pediatrics and Ophthalmology were sought to rule out phakomatosis. She was evaluated and found not to be having any such features, and hence, she has been put on annual follow-up.

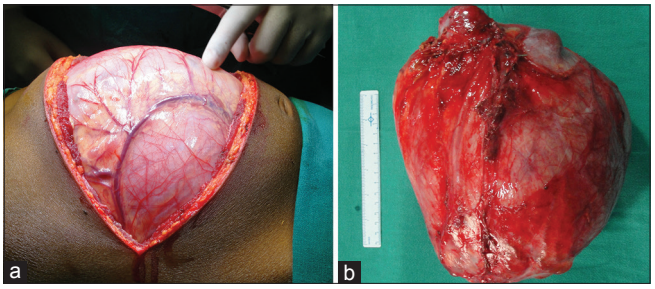

Figure 2: (a) Intra-operative image showing the lump occupying almost the entire abdominal cavity with colon stretched over it and pushed to the right of the patient (b) The excised specimen showing that it arose predominantly from the middle and lower pole of the left kidney

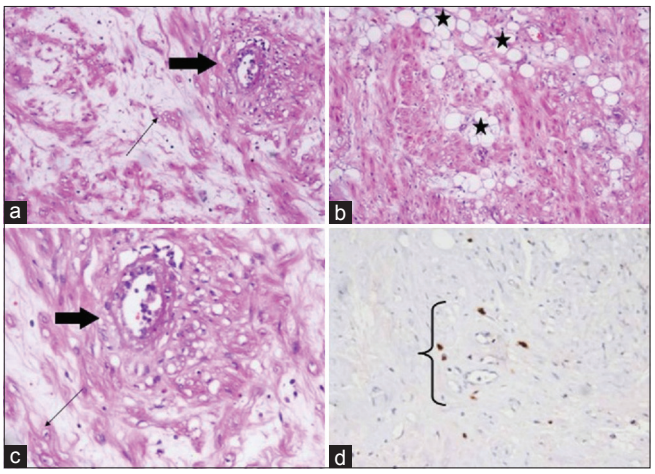

Figure 3: Photomicrograph of the tumor shows features of anangiomyolipoma with a variable mixture of smooth muscle bundles (thin arrow), blood vessels (bold arrow), and adipose tissue (star) (a and b: H and E, ×40 and c: H and E, ×200). Focal immunopositivity for hydroxy beta-methyl butyric acid-45 was also noted (parentheses) (d: HMB 45, ×200)

DISCUSSION

The histopathological report of this being an AML was a surprise. AML is an uncommon benign tumor more so in children. AML comprises haphazardly arranged vascular, smooth muscle and fatty tissues. The histologic picture resembles a hamartoma, but currently, they are considered as benign neoplasms.[2] There is a 4:1 female predominance and the mean age at presentation is around 40 years.[1] Eighty percent of children with tuberous sclerosis develop these lesions by the age of 10 years.[2] In children, renal AML are rare without tuberous sclerosis.

The imaging characteristics of renal AML can vary considerably based on the amount and type of histologic elements present. CT scan images are diagnostic when macroscopic fat is found within the mass. Although fat is occasionally found even in Wilms tumor, the diagnosis of renal AML is generally straight forward when correlated with other clinical features. T chaprassian *et al*. had described three cases of pediatric renal AML (age range 11–15 years) that posed a diagnostic challenge.[3] In all 3 cases, the diagnosis was made only after nephrectomy. The misdiagnosis was attributed to the large size of the tumor with poor fat content, in an age group where renal AML is usually not common. Renal AML is the only benign renal tumor that can be categorically diagnosed by radiological

studies alone. Despite this, the imaging in our case did not indicate towards AML because of low-fat content. Lipopenic AML comprises only about 5% of all AMLs, and in this situation, the diagnosis can be difficult to clinch with radiology alone.[4]

Wilms tumor accounts for 87% of all pediatric renal masses. Although 75%–80% of the patients present before 5 years of age, the age range is quite wide, and cases have been reported even in adolescence and adults.[1,5] In fact, in the Society of Pediatric Oncology (SIOP) 93-01 study, thirty patients out of 962 patients enrolled with Wilms tumor were older than 16 years.[5] In the SIOP study, Wilms tumor in children between 6 months to 16 years were diagnosed by means of radiological studies alone.[5] With this background, the diagnosis of Wilms tumor was initially considered in our patient. Because the size of the mass had decreased on chemotherapy further made us to believe that this mass was in fact a Wilms tumor.

FNAC is also helpful in diagnosing renal AMLs. Typical cytological features described in conventional AML are admixture of adipocytes, smooth muscle cells, endothelial cells, foam cells, and giant histiocytes with a backdrop of inflammatory cells.[6] However, in our case, it was non contributory. A FNAC or needle biopsy before initiating chemotherapy probably would have been helpful in diagnosing it early.

Tuberous sclerosis is a neurocutaneous syndrome which has an autosomal dominant inheritance pattern. Tuberous sclerosis complex has been characterized by Vogt's triad, comprising mental retardation, adenoma sebaceum, and seizures. Renal AMLs in the setting of tuberous sclerosis may reach giant proportions although there is no consensus on when to term an AML as "giant."[7-9] The largest reported case of bilateral renal AML measured 30 cm × 21 cm × 13 cm and 30 cm × 18 cm × 10 cm with a total tumor burden of 7843 g that resulted in respiratory failure.[10] Our patient was later evaluated for any markers for phakomatosis but none were found. After reviewing the literature, it is our belief that our case has by far the largest pediatric sporadic AML reported in literature. The largest sporadic renal AML reported till date weighed 7500 g in a 49-year-old lady.[11]

Management is conservative if the size of the lesion is <4 cm but larger lesions need to be surgically removed. Nephron-sparing surgeries are advocated, especially in tuberous sclerosis patients in whom bilateral affliction may occur. In our case, the absence of a pre-operative diagnosis precluded a nephron-sparing approach although in the hindsight, renal preservation would have been a difficult technical proposition considering the size and extent of involvement of the kidney.

To conclude, a rare case of giant sporadic pediatric renal AML has been reported that was mistaken for a Wilms tumor. This case highlights the importance of obtaining a tissue diagnosis before initiating chemotherapy, especially when the age of the patient is not in the standard age bracket for Wilms tumor.

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How to cite this article: Dhua AK, Ranjan A, Agarwala S, Bhatnagar V, Mathur SR, Devasenathipathy K. Giant renal Angiomyolipoma masquerading as a Wilms tumor. Indian J Urol 2017;33:73-5.