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A rare giant renal angiomyolipoma in a 17-year-old male: A case report

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ABSTRACT

Renal angiomyolipoma is an uncommon benign tumor that mostly affects adults and is highly associated with tuberous sclerosis. It's very uncommon for it to affect adolescents. In this case, a 17-year-old male with abdominal pain and distension was diagnosed with sporadic giant renal angiomyolipoma. Histopathology confirmed the diagnosis after a successful nephrectomy. Though rare in teenagers, giant renal angiomyolipoma can be managed with surgery. Histopathology is a mainstay of confirming the diagnosis.

Conclusion: This case highlights the difficulties in managing big retroperitoneal tumors in teenagers and the necessity of taking sporadic angiomyolipoma into account when making a differential diagnosis.

1. Introduction

Rare giant mesenchymal tumors with variable ratios of smooth muscle cells, blood vessels, and adipose tissue make up renal angiomyolipoma.[1]. There are very few occurrences recorded in pediatric and adolescent populations; they are usually seen in adults from the second decade of life and are frequently linked to tuberous sclerosis complex, however 80 % of case are sporadic and usually affect adult group[2,3]. Tumors bigger than 10 cm in diameter are known as giant renal angiomyolipoma, they are rare and present serious management challenges because of the possibility of consequences like hemorrhage and rupture.[4]. We report a rare instance of a 17-year-old male with a giant renal angiomyolipoma.

2. Case presentation

A case of 17 years old male presented with a 4 years history of gradual painless hard surfaced and immobile abdominal mass which began solely on his left upper quadrant progressing further towards the midline associated with early satiety, loss of appetite and occasionally, non-projectile vomiting of food contents no history of constipation no blood in stool, no history of hematuria or change in urination habit, not epileptic and no history of mental illness reported, no history of the same condition in the family.

General examination: alert, not pale, not jaundice, slight dyspneic, no lymphadenopathy.

The vital signs were essentially within a normal range.

Per Abdominal Examination: massive distended abdomen with palpable mass on the left lumbar region which extends towards the left hypochondriac region, immobile and mildly tender on palpation, it was firm on consistency irregular in shape and does not cross the midline. Deep palpation was difficult due to the distention but there were no signs of hepatomegaly nor splenomegaly.

Laboratory results: urinalysis microscopic hematuria, CBC = Hg 11g/dl other parameters essential were normal, creatinine 112g/dl, BUN 4g/dl.

Abdominal CT-Scan: reviled a well-defined solid heterogeneous retroperitoneal mass originated from the left kidney containing markedly hypodense area with some necrotic components. no signs of local invasion to the near structures, hence strongly suggested angiomyolipoma (Fig. 1).

Diagnosis of renal angiomyolipoma was reached based on the history, clinical findings and abdominal CT-Scan with contrast, hence laparotomy was done, where a huge well capsulated mass measures (32x27 × 19cm) and weigh 4.8kg, originating from the left kidney was found with no invasion to nearby structure and no mesenteric lymph node identified, trans peritoneal left nephrectomy was done with total mass excision with minimal bleeding (Fig. 2.), drainage inserted and abdomen was closed. The whole mass was sent for histopathology. Patient was sent to general ward on day three the drain was removed and

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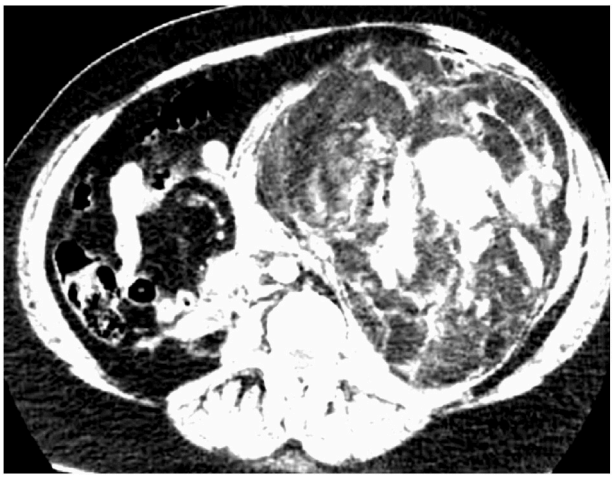

Fig. 1. Abdomina Ct-scan film shows the left renal Angiomyolipoma.

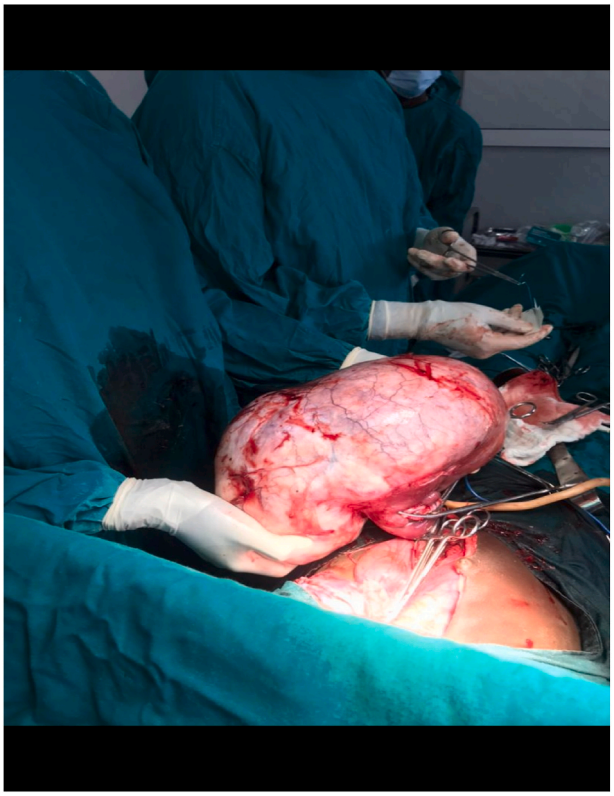

Fig. 2. Giant left renal mass.

discharged home, follow up was done for six months (creatinine monitoring and urine output) on outpatient clinic with no any significant complication reported.

Histological findings: multiple sections of tissues lined by fibrous capsule, stroma composed of triphasic with spindle cells, mature adipose tissue and thick-walled blood vessels. Focal areas with hypercellular of epithelioid shaped. Extensive areas with hyalinization changes were also seen. Conclusion of angiomyolipoma were reached (Figs. 3 and 4).

3. Discussion

Renal angiomyolipoma is the commonest being renal condition contribute about 0.3–3% of all benign renal masses, most affect 2nd and 5th decade of life[4,1].

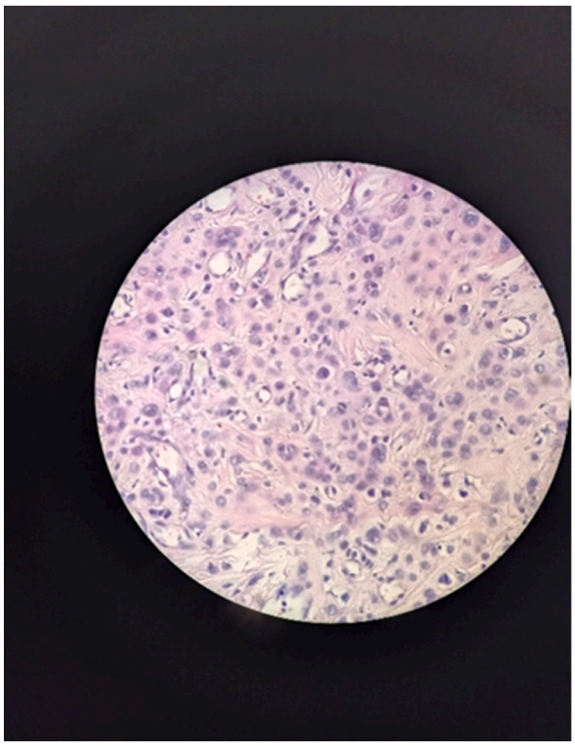

Fig. 3. Tissues lined by fibrous capsule; stroma composed of triphasic with spindle cells. stroma composed of triphasic with spindle cells, mature adipose tissue and thick-walled blood vessels.

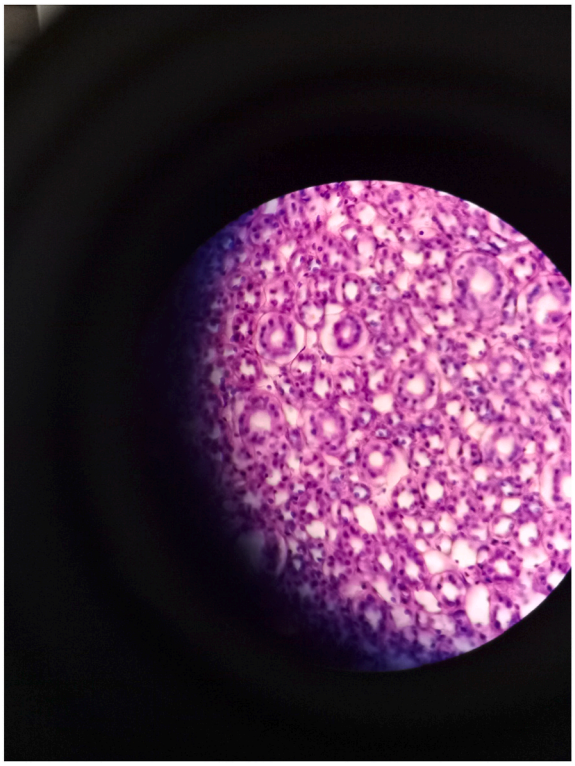

Fig. 4. Focal areas with hypercellular of epithelioid shaped. Extensive areas with hyalinization changes.

Sporadic type is commonly affecting 4th decade of life and often less aggressive,[5], very rare in adolescent age.[3]. The classic clinical presentation are abdominal distension, hematuria and frank pain, however most of the tumors are asymptomatic.[6].

The diagnosis of angiomyolipoma mostly is by abdominal CT-Scan with contrast or MRI, supported by urinalysis which shows hematuria, in some advanced setting genetics study is done[7],[6],.[2]. Management can either be emergency for retroperitoneal bleeding tumors, radical nephrectomy for larger invasive unilateral tumors, embolization for small tumors <4cm and expectant management for smaller asymptomatic tumor[8].

This case report explains a rare case of giant unilateral solitary sporadic giant angiomyolipoma in 17 years old boy, this was diagnose based on clinical history, examination, investigations done. However, our patient doesn't have any stigma for tuberos-sclerosis hence was more suggestive of sporadic type.[8] Was schedule for elective laparotomy, intraoperative findings were a complex giant mass 32x27 × 19 cm, weigh 4.8kg, originate from the posterior edge of the left kidney with no invasion to the surrounding structures, hence left total nephrectomy was done as indicated in the other literature as the mode of managements[3],[9].

Giant sporadic angiomyolipoma is often a rare in younger age, however for adolescence is mostly extremely less reported,[5], sporadic type commonest preset with small mass of approximately <4cm, giant sporadic angiomyolipoma is often very rare in adolescent age.[2]. Definitive management of giant Angiomyolipoma is nephrectomy which possess less recurrence rate and good prognosis (p. 7), in the Morden advanced setting, selective artery embolization therapy can be opted for, which has shown success for even patients with large masses and those with a higher risk of hemorrhage,[10], In a patient with tuberous sclerosis, mTOR inhibitors remain the treatment of choice.[11].

However, with this finding of giant sporadic angiomyolipoma in adolescent age raise awareness and suspicious to think when dealing with giant retroperitoneal tumors originating from the renal and when encountered nephrectomy is still a management of choice at limited resource settings.[9].

4. Conclusion

Giant sporadic renal angiomyolipoma are uncommon tumors in children and adolescents. This case highlights the need to consider them in diagnosing giant renal masses in adolescents with flank pain and swelling. Early diagnosis and proper surgical treatment are crucial for effective management for big masses and preventing complications.

This work has reported in line with the SCARE criteria[12].

CRediT authorship contribution statement

Fred Laizer: Writing – original draft, Resources, Funding acquisition, Formal analysis, Data curation, Conceptualization. **Hanson Nkini:** Writing – review & editing. **Heri Babu:** Supervision, Methodology. **Erasto Medard:** Writing – review & editing, Validation, Supervision. **Amon Bamanyis:** Investigation. **Teodros Veronesi:** Writing – review & editing.

Consent

Written informed consent was obtained from the patient's parents for the publication of this case report and accompanying images. A copy of this consent form is available for the editor-in-chief of this journal to review upon request.

Ethical approval

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Conflict of interest

N/A.

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