Case Report

Sporadic giant renal angiomyolipoma: A case report and literature review of clinical presentation, diagnosis, and treatment options

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Abstract

Giant angiomyolipoma (AML) is uncommon benign renal neoplasm that occurs sporadic or in association with tuberous sclerosis syndrome. There is no specific cutoff tumor size and/or weight to define giant AMLs. Ovarian stimulation hormones might cause a marked increase in its size and subsequent bleeding risk. Incidental findings are not common in those patients as mass-associated symptoms always present. Computed tomography scan is the standard diagnostic imaging study except for tumors with poor fat content. According to the clinical presentation, site and side of renal involvement giant sporadic AMLs have different treatment options, for example, active surveillance, selective renal artery embolization, nephron-sparing surgery, and/or radical nephrectomy. In the present case report, we present a 22-year-old female with huge right renal mass (29 cm × 23 cm × 21 cm) and treated with right radical nephrectomy which proved to be renal AML on pathology examination.

Keywords: Angiomyolipoma, giant, sporadic, treatment

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INTRODUCTION

Angiomyolipoma (AML) is the most common benign renal tumors and represents up to 3% of renal masses.[1] AMLs are usually diagnosed incidentally; however, with large-size AMLs (>4 cm) symptoms such as hemorrhage, pain, palpable mass, and mass-associated symptoms may present. A potentially life-threatening hemorrhage may occur due to abnormal vasculature, and/or rupture aneurysms require urgent intervention.[2]

Giant AMLs are extremely rare, with no specific cutoff tumor size or weight to define it. Few articles have reported cases of sporadic AMLs with a size of >20 cm.[3-10] In this article, we represent a case of sporadic AML with a size of 29 cm and weight of 9000 g treated at our urology department, with a thorough literature review of clinical presentation, diagnosis, and treatment options for sporadic AMLs (>20 cm).

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CASE REPORT

A 22-year-old female was referred to the King Saud Medical City Outpatient Urology Clinic for the assessment of large right abdominal swelling. She gave a history of right-sided abdominal swelling for the past 3 years which slowly increase in size and was associated with vague abdominal pain. Otherwise, she had no other urinary or gastrointestinal symptoms. She was diagnosed recently to have hypertension treated with angiotensin-converting enzyme inhibitor (ACEI).

On examination, the abdomen was soft with large abdominal swelling, smooth surface, solid in consistency, and not associated with tenderness, with superficial dilated veins in the anterior abdomen, lower back, and right lower limb [Figure 1a].

All laboratory investigations were within the normal values. Computed tomography (CT) abdomen and pelvis with contrast showed multilocular multicystic fluid containing mass arising from the right kidney, measuring 29 cm × 23 cm × 21 cm with no calcifications or soft-tissue enhancement, nor fat content [Figure 2]. There was no enlarged lymph node in the abdomen and pelvis. The chest X-ray was normal. Initial diagnosis was multilocular cystic nephroma, but renal hydatid cyst was suspected. Therefore, we started prophylactic albendazole 4 weeks before the surgery. An informed consent was obtained from the patient for the intervention and its possible complications. Actually, we have decided to perform the right radical nephrectomy based on preoperative images showing that the huge mass originates from the right kidney, as we believe that other treatment options such as partial nephrectomy (PN) and/or angioembolization would not be feasible treatment for such a huge mass. The right hemi-chevron approach with midline extension incision [Figure 1b] was performed with all precautions for hydatid cyst lesion.

Grossly, the mass was well circumscribed, measured 30 cm × 22 cm × 20 cm and weighted 9000 g with an attached 3 cm segment of dilated ureter [Figure 3a]. Cross-section cuts revealed tan-white-yellowish, homogenous gelatinous mass arising from the kidney parenchyma with no areas of hemorrhage or necrosis. Multiple dilated cystic spaces filled with clear yellowish fluid with smooth thin walls were seen [Figure 3b]. There was a thin normal cortex in the lower pole. Microscopic examination [Figure 4a and b] revealed a mass composed predominantly of smooth muscles within an edematous stroma, island of mature fat, and thick blood vessels are also noted throughout

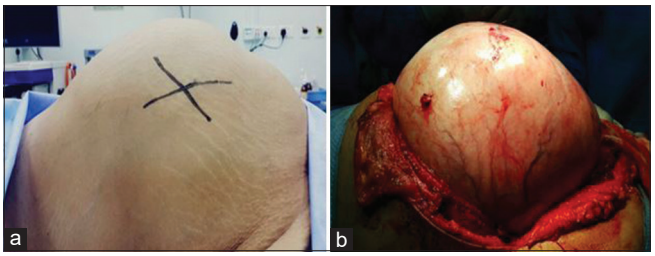

Figure 1: Large abdominal swelling (a) before "left side image" and (b) during the surgery "right side image"

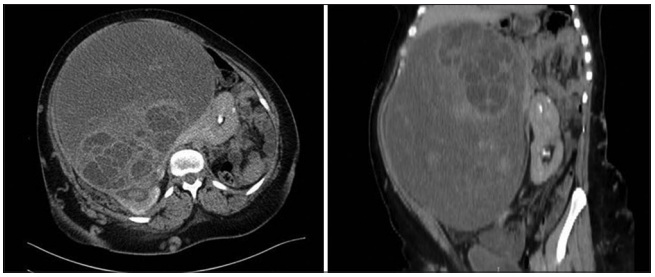

Figure 2: Abdominal and pelvic computed tomography scan with contrast showing multilocular multicystic fluid containing mass arising from the right kidney, measuring 29 cm × 23 cm × 21cm with no calcifications or soft-tissue enhancement, nor fat content

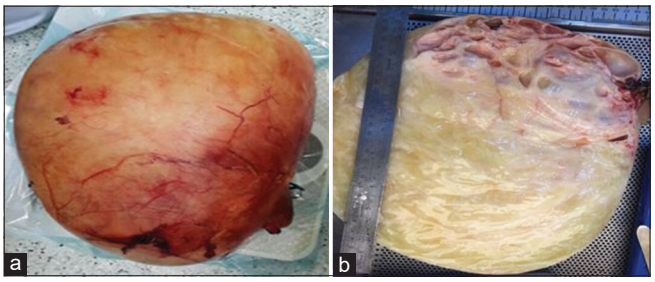

Figure 3: Gross picture pathology (a) specimen weight 9000 gram "left side image" and (b) cross cut section of the mass "right side image"

the tumor, with no evidence of malignancy or epithelioid elements. Tissue immunohistochemical staining showed a strong positivity of smooth muscle actin, caldesmon, and estrogen receptor. No definite staining is seen with human melanoma black (45) and CD117. CAE1/AE3 highlighted entrapped dilated tubules. These features are consistent with AML. Tumor is entirely excised with no evidence of malignancy seen.

Postoperatively, she was doing fine and had a normal renal function during follow-up, as well as, she no longer use ACEI for the treatment of hypertension. Radiologically, there was no evidence of tumor recurrence at a 15-month follow-up.

DISCUSSION

AML is uncommon benign renal neoplasm which composed a variable mixture of mature adipose tissues,

blood vessels, and fully differentiated smooth muscles. AML is possibly derived from perivascular epithelioid cells, it predominates in female, and recent evidences suggest strong tumor positivity to female hormones with increasing its size during pregnancy[11] and oral contraceptive therapy.[12] More recently, Watanabe *et al*. have reported a massive increase in AML size from 1 to 18 cm for a 40-year-old female after 10 months of treatment with ovarian stimulation hormone.[13]

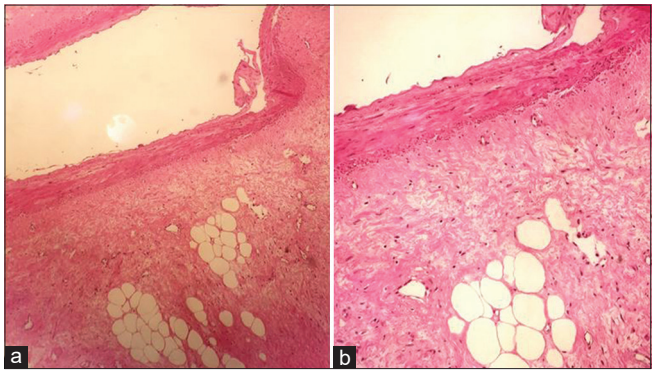

Figure 4: Microscopic examination (H and E) of the tumor with medium and high power on the left (a) and right images (b), respectively

Similar to small-sized AMLs, giant AMLs (>20 cm) might occur in sporadic form[3-10] or in association with genetic syndromes like tuberous sclerosis complex (TSC).[14,15] To the best of our knowledge, there is no specific cutoff tumor size and/or weight to define giant AMLs. Previous case reports showed that AMLs may reach size up to range of 25–39 cm and weight of 3818–17,900 g. Of note, our patient had a tumor size of 29 cm in largest dimension and weight of 9000 g.

According to our literature review [Table 1], it seems that the incidental finding of sporadic AMLs (>20 cm) is not reported since all patients have symptoms and signs of large abdominal mass effect.[3-10] Patients commonly present with abdominal swelling and fullness, dull aching,

Table 1: Clinical presentation, diagnosis, and treatment options of angiomyolipomas

Author	Sex	Age (years)	Side	Largest dimension (cm)	Weight (g)	Clinical presentation	Diagnostic imaging	Treatment option
Nepple *et al*., 2010[3]	Male	53	Right	35	17,900	Progressive weight gain, with increasing abdominal girth. Early satiety with postprandial nausea and emesis	CT: Cystic renal mass without fat density	Radical nephrectomy
Taneja and Singh 2013[4]	Female	49	Left	39	7500	Bloating sensation in the abdomen and a steadily increasing abdominal girth	CT: A large fat containing tumor	Radical nephrectomy
Cavicchioli *et al*., 2014[5]	Male	50	Bilateral	27.5 (left) 28.5 (right)	NA	No history of pain, hematuria, or other symptoms, but only complained of abdominal enlargement	CT: Hypointense masses with no significant contrast enhancement	Active surveillance
Zhou *et al*., 2015[6]	Female	50	Bilateral	28 (right) 12 (left)		Flank mass and abdominal fullness	CT: Right large AML and multiple left AMLs	Right radical nephrectomy. left preoperative SAE, then PN
Kori *et al*., 2015[7]	Female	30	Left	25	8000	Dull pain and palpable lump in left side of the abdomen	CT: Well-defined mass with multiple enhancing vessels and fat predominance (−69 HU)	Radical nephrectomy
Albersheim-Carter *et al*., 2016[8]	Female	61	Left	30	3818	Hemodynamic instability from a retroperitoneal bleeding	CT: Mass containing few small vessels, mixed fatty and soft-tissue elements	First, IV fluid resuscitation, transfusion and SAE. Later on PN
Chen *et al*., 2017[9]	Female	34	Left	29	NA	Progressive bloating sensation in the abdomen	CT: Large mass with mixed density	Radical nephrectomy
Sharma *et al*., 2018[10]	Female	42	Left	25	NA	Abdominal pain and fullness. The pain is mild, dull aching, localized and continuous with a gradual increase in intensity over a period of time	CT: Fat density rounded lesions in the left kidney. A large perirenal mass showing prominent vessels	Enbloc excision of the retroperitoneal mass with a left nephrectomy and a segmental descending colectomy
Current study 2019	Female	22	Right	29	9000	Abdominal swelling slowly increase in size and associated with vague abdominal pain	Multilocular multicystic fluid containing mass, with no calcifications or soft-tissue enhancement, nor fat content	Radical nephrectomy

CT: Computed tomography, AMLs: Angiomyolipomas, SAE: Selective renal artery embolization, PN: Partial nephrectomy, IV: Intravenous, HU: Hounsfield units, NA: Not available

or vague abdominal pain; the bloating sensation which might associated with nausea and vomiting. The clinical presentation usually has an insidious onset and slowly progressive course. A serious presentation of giant AMLs is bleeding either retroperitoneal or into the collecting system which if massive might associated with hemodynamic instability and require resuscitation with urgent surgical intervention.[8]

AMLs have characteristic findings on radiological evaluation to differentiate it from renal cell carcinoma (RCC). Ultrasound usually reveals a hyperechogenic well-circumscribed mass with shadowing. CT with contrast is diagnostic of AML by the presence of fat (confirmed by a value of –20–80 HU on imaging).[16] The absence of fat attenuation usually makes it difficult to differentiated AML from RCC, as in our case, the CT scan did not show fat content in the mass (only microscopic visible fat). Similarly, Nepple *et al*. have reported a large cystic renal mass (35 cm) without fat density on CT that on histopathological examination was found to be a fat-poor AML with predominant epithelioid cells.[3] Of note, about 5% of AMLs do not show fat attenuation on imaging studies.[16]

The treatment decision of sporadic giant AMLs is greatly determined if one or both renal units are involved, presence of normal renal tissue to preserve it, as well as, the patient clinical presentation. Most of cases of giant AMLs are symptomatic and treated with radical nephrectomy due to involvement of the whole renal unit[3-7,9,10] and that was the treatment option we offered to our patient. In some occasions, if the patient has no symptoms active surveillance might be an option. Cavicchioli *et al*. have reported a successful active surveillance protocol with serial magnetic resonance imaging follow-up every 4–5 months for the treatment of male patient with bilateral giant AMLs (the right mass size was 28.5 cm, and the left mass size was 27.5 cm), with abdominal enlargement is the only complaint and no pain, hematuria, or other related symptoms.[5] In patients who developed severe bleeding that affects the hemodynamic status, urgent intervention is required. Albersheim-Carter *et al*. have reported on a 60-year-old female patient with left side giant AML (30 cm) who presented with shock due to massive retroperitoneal bleeding secondary to hemorrhage from the tumor and was treated with intravenous fluid replacement, blood transfusion, and immediate selective renal artery embolization (SAE). Four days after SAE, they offered her a successful PN to avoid any future bleeding attack.[8] Since preservation of postoperative renal function is one of the main goals to achieve during the treatment of giant AMLs, PN should be an option whenever it is possible. More recently, Zhou *et al*. have treated a 50-year-old female patient with bilateral AMLs (the right mass size was 28 cm, and the left mass size was 12 cm), with right radical nephrectomy and left SAE followed by PN after reduction of the tumor size.[6] From our review, it seems that recurrence after surgical treatment of AMLs (>20 cm) are uncommon, and all patients were tumor-free at their last follow-up.

In summary, giant AMLs are extremely rare condition that occurs sporadic or in association with TSC. Female hormones might cause a marked increase in its size and subsequent bleeding risk. Patients usually symptomatic and have large abdominal mass-related symptoms. CT scan is diagnostic in majority of cases due to its fat attenuation effect. Treatments of those giant masses include different treatment modalities such as active surveillance, SAE, PN, and radical nephrectomy according to the clinical scenario.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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