CASE REPORT

Giant renal angiomyolipoma (RAML): A case report and literature review

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Abstract

Renal angiomyolipoma is a benign, rare type of tumour which consists of mature adipose tissue, thick-walled blood vessels, and smooth muscle cells. This tumour can grow to a large size, and is then defined as giant.

We are describing the case of a 34-year-old female with a giant renal angiomyolipoma, measuring 32cm x 28cm x 6cm, which caused her pain and constipation. The mass was removed successfully, and partial nephrectomy was done. The patient had an uncomplicated postsurgical clinical course, and renal function was preserved.

Renal angiomyolipoma is a hamartomatous, benign mass with variable components. It can occur sporadically or in association with genetic syndromes such as tuberous sclerosis. Symptoms include abdominal pain and constipation. Potentially dangerous sequelae include retroperitoneal haemorrhage and shock.

The treatment for these tumours is surgery with open nephrectomy. Other treatment modalities have also been developed. Diagnostic workup should be done for genetic syndromes whenever necessary.

Keywords: Angiomyolipoma, Renal angiomyolipoma, RAML, Tumour.

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Introduction

Angiomyolipomas (AMLs) are composed of three tissue elements: mature adipose tissue, thick-walled blood vessels, and smooth muscle cells. Studies have shown that renal AML can grow by 4cm each year in its maximum dimension.[1] AMLs are most commonly seen in the kidney, 80% of them are sporadic but 20% are related to tuberous sclerosis.[2] Although benign in nature, they have been shown to invade the renal vascular system.[3] Tumours greater than 10 cm (referred to as "giant" AMLs) are occasional.[4] Only a few reported cases had giant tumours larger than 20 cm.

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Reported here is the case of a patient with giant renal AML with subsequent management and treatment. Informed consent was obtained from the patient for writing about the case and publishing the findings.

Case Presentation

A 34-year-old female presented to the outpatient department of Jinnah Hospital, Lahore, in December 2023 with a four-month history of mass in the left lower abdomen, with associated pain and relative constipation.

General physical examination revealed a healthy-looking middle-aged woman, well oriented in time, place, and person lying comfortably in bed.

On inspection, the abdomen was protuberant in the centre, with an inverted umbilicus. Pfannenstiel scar mark was present. There were no visible striae or dilated veins. On palpation, the abdomen was soft and tender and bowel sounds were audible. A 25x15cm firm mass was felt in left iliac fossa. The mass was non-translucent. Cough impulse was negative.

Full blood count revealed no significant disturbance. Her haematocrit was 37.5% (Normal Range 36-44%). Chemical pathology report was positive for Anti-HCV but negative for HbsAg.

Ultrasonography revealed a large lumbar echogenic (fat containing), heterogenous lesion arising from the lower pole of the left kidney, with no surrounding haematoma. Right kidney was normal in size with no associated cyst, stones or hydronephrosis. Other abdominal organs were unremarkable.

On CT of the abdomen and pelvis with contrast, there was a large, exophytic, predominantly fatty mass in the lower pole area of the left kidney measuring approximately 22 x 15.7 x 13cm. The mass also contained multiple septate and non-fatty tissue areas. There was heterogenous post-contrast enhancement of the mass with septal enhancement. The kidney was displaced superiorly, and gut loops were displaced towards the right by the mass. Renal parenchymal defects were noted at the lower pole with parenchymal vascular pedicle and prominent intra-tumoural vessels. The right kidney was unremarkable.

Spleen was mildly enlarged and there were multiple aortocaval and para-aortic lymph nodes.

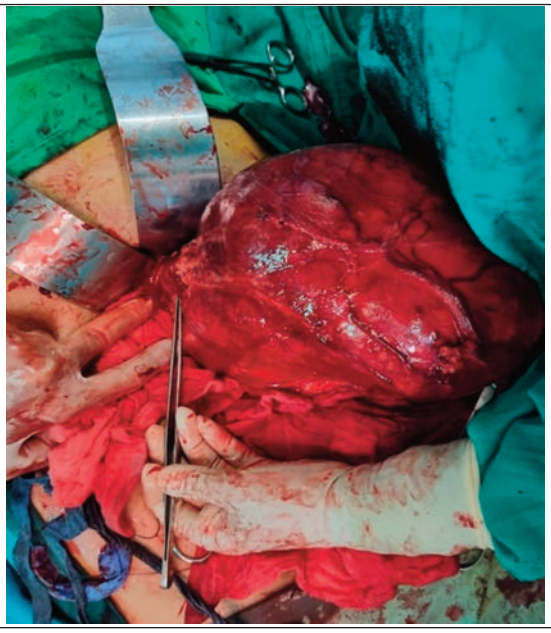

Figure-1: Cut section of the RAML revealing fatty tissue continuous with renal capsule.

There was no evidence of haemorrhage or other intra-renal fatty lesions. CT findings most likely suggested angiomyolipoma.

In CT angiography of the abdomen, all the major arteries were normal in calibre. Bilateral renal arteries were unremarkable. No evidence of stenosis, filling defect, aneurysm or vascular malformation was seen.

After pre-op work-up, the patient underwent surgery for removal of the tumour. Partial nephrectomy was done as CT scan showed evidence of angiomyolipoma, which is benign, and the tumour only involved the upper pole of the kidney, leaving the rest of the kidney normal. Transabdominal midline approach was used. The mass was successfully removed with the utmost attempt to preserve most of the renal function.

Pathological features: Histopathological examination of the mass showed, a well-circumscribed mass measuring 32cm x 28cm x 6cm with renal pole at lateral surface measuring 5cm x 4cm in the gross examination. Cut sections revealed yellow fatty tissue continuous with the renal capsule [Figure 1].

H&E stained sections revealed a tumour comprising three tissue components, myoid spindle cells, mature adipose cells, and dysmorphic thick-walled blood vessels. Smooth muscle component appeared to originate from the vessel walls and was hyper-cellular. No granulomas or malignancy were seen in the given section.

According to the latest follow-up session, in March 2024, the patient was healthy with good renal function.

Discussion

AML is a benign hamartomatous lesion having varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels. Renal AMLs are benign tumours known to occur sporadically and in association with genetic syndromes like tuberous sclerosis and lymphangioleiomyomatosis.[5] Our patient's AML was not associated with any systemic manifestations.

The prevalence of AML is 0.44%; it is female predominant with female to male ratio of 2:1, suggesting that hormones could be contributing to the growth.[4]

Tumours may be asymptomatic, and the discovery is usually incidental on imaging. The main problems associated with AMLs are retroperitoneal haemorrhage and impingement of the tumour on the kidney and surrounding structures.[4] Up to 15% of the patients may present with retroperitoneal haemorrhage, also known as Wunderlich syndrome, which can lead to shock in one-third of the patients and can be life-threatening.[6]

In renal angiomyolipoma, patients may have variable clinical presentation according to the size of the lesion, its blood supply, and formation of pseudoaneurysms. The risk of formation of aneurysm is directly proportional to the tumour size.[7]

More than 50 cases of giant angiomyolipomas have been reported, most of which were unilateral. The literature was searched using PubMed and google scholar for the keywords 'giant angiomyolipoma' and 'angiomyolipoma of kidney' for the period 1979–2022.

Table-1: Giant renal angiomyolipoma in the indexed literature, by size.

Study	Year	Size (cm)	Side	Tuberous sclerosis	Bilateral
Katz et al[11]	1997	45×20×12	Left	Yes	No
Taneja et al[5]	2013	39×25×9	Left	No	No
Bains et al[12]	2020	35×20×12(R) 14×6 (L)	Both	Yes	Yes
Hussain et al[13]	2013	32×16×12 (R) and 27×21×16 (L)	Both	Yes	Yes
Current case	2022	32×28×6	Left	No	No
Barbará et al[14]	2004	31×19×10 (L) and 14×5.5 (R)	Both	Yes	Yes
Castillo et al[15]	1987	30×25×20	Left	No	No
Alshehri et al[16]	2020	30×22×20	Right	No	No
Tong et al[17]	1980	30		No	No
Zhou et al[18]	2015	28×20×14 (R) and 12×9×6 (L)	Both	No	Yes
Eason et al[19]	1797	28×16×9	Left	No	No
Hsu et al[20]	2002	24×17×10	Right	No	No
Cichocki et al[21]	2014	22.5×9.5×10.4	Left	No	No

The present case appears to be the fifth largest RAML in the world and largest RAML ever reported in Pakistan, measuring 32cm x 28cm x 6cm. (Table 1)

Ultrasonography, which is not reliable or sensitive in these tumours, shows a hyperechoic lesion with acoustic shadowing which cannot be differentiated from other kidney tumours. Computed tomography (CT) with contrast enhancement is the most frequently used radiologic tool to diagnose AML. It can distinguish AML from other kidney lesions. AML is the only benign renal tumour that is confidently diagnosed on cross-sectional imaging. The fat (confirmed on non-enhanced thin-cut computed tomography by a value of –20 [HU] or less) shown within a renal lesion on imaging is considered the diagnostic hallmark.[5]

On histopathology, AMLs are composed of three tissue elements: mature adipose tissue, thick-walled blood vessels, and smooth muscle cells. AMLs were originally classified as hamartomas but are now thought to belong to the family of perivascular epithelioid cell tumours (PEComas).[8] Immunohistochemically, AML is characterised by co-expression of melanocytic markers (HMB-45 and Mart-1/Melan A) and myoid markers (SMA and muscle-specific actin) which differentiate it from other tumours.[9]

Angiomyolipomas are at risk for spontaneous haemorrhage. It has been widely accepted that AML tumour size >4cm or intratumoral aneurysms >5mm is the recommended criteria to determine the risk of tumour rupture.[10]

Criteria for intervention is symptomatic lesions larger than 4cm, suspicion for malignancy, and presence in women of childbearing age.[11] Management options include embolization, radiofrequency ablation, and partial or total nephrectomy.

In this patient, as the tumour originated from the lower pole of the left kidney and was adherent to the capsule, partial nephrectomy was performed along with tumour resection to somewhat preserve renal function.

The patient was seen to be healthy at a follow-up visit and had no complications following treatment.

Conclusion

Renal angiomyolipoma (AML) is a benign tumour which usually occurs sporadically in the population. There are few cases of AML larger than 10cm in size, and even fewer reported cases of it being larger than 20cm. The tumour seen in this case appeared to be the fifth largest one ever reported. Tumours this size can lead to serious complications, such as haemorrhagic shock, and should be treated promptly, even if asymptomatic. Several therapies for this type of growth have been developed. Care must be taken to preserve as much renal function as possible whenever the lesion is being treated surgically. In this case, surgical treatment was promptly performed and partial nephrectomy was deemed the best option. Diagnostic workup was done for tuberous sclerosis as this is a possibility in patients with these tumours and should always be considered.

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Conflict of Interest: None.

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AFM: Data analysis, reviewing and editing.
HUA: Drafting, found references and added citations.
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