

Giant Angiomyolipoma Masquerading as Perinephric Abscess: A Diagnostic Conundrum

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Abstract

Renal angiomyolipomas are the most common benign tumours of the kidney accounting for up to 1% of all renal masses. Giant angiomyolipomas which have a size greater than 10 centimetres are rare entities with few cases reported in literature. Small angiomyolipomas are usually a symptomatic and increasing size correlates with symptomatology. These are usually incidentally detected or when symptomatic may present with an abdominal lump, flank pain or hemorrhage. Herein, we report a rare case of 45-year-old lady with giant angiomyolipoma with clinical presentation indistinguishable from perinephric abscess. The case is rare with regards to the large size of tumour and the discordant presentation unusual for an angiomyolipoma.

Keywords

Angiomyolipoma, Giant Angiomyolipoma, Perinephric Abscess

1. Introduction

Renal angiomyolipomas are the most common benign tumours of the kidney accounting for up to 1% of all renal masses. Giant angiomyolipomas which have a size greater than 10 centimetres are rare entities with few reported cases in literature. Small angiomyolipomas are usually asymptomatic and increasing size correlates with symptomatology. These are usually incidentally detected or when symptomatic may present with an abdominal lump, flank pain or hemorrhage. Here, we present a case that was misdiagnosed as perinephric abscess due to its peculiar presentation. The case is rare with regards to the large size of tumour and the discordant presentation unusual for angiomyolipoma.

2. Clinical Presentation

A 45-year-old lady presented to the emergency department with complaints of

fever, left flank pain and burning micturition for duration of one week. She had been on thyroxine 50 mcg for hypothyroidism for 8 years and was euthyroid. She had no other significant past medical or family history. Physical examination revealed pallor and a tender renal lump in left lumbar region. She had haemoglobin level of 8.4 g%, leukocyte count of 28,600/mm³, blood urea of 90 mg% and serum creatinine of 2.1 mg%. Urine routine microscopy showed 8 - 10 pus cells/hpf. Urine culture was sterile. Ultrasound of the abdomen showed a heterogeneously hypoechoic collection of dimensions 9.5 × 5.6 centimetres in left perinephric region visualised separately from the left kidney. An initial diagnosis of left perinephric abscess with acute kidney injury was made and management was instituted for the same. Patient was admitted for a course of IV antibiotics and therapeutic drainage of the abscess. Upon attempted diagnostic aspiration, only 5 millilitre of clotted blood was obtained. The initial diagnosis was called into question in this new light. Patient's kidney function improved with adequate hydration. A contrast enhanced computed tomography (CECT) scan was done for confirmation following normalisation of kidney function. CECT revealed giant left renal angiomyolipoma with large intra-tumoral bleed and an arterial feeder arising from left kidney (**Figure 1**).

Patient underwent selective arterial embolization which was unsuccessful. The patient was then planned for open exploration. Left simple nephrectomy was performed as nephron sparing nephrectomy was not deemed possible in this case due to large size of tumour encasing whole of the left kidney and intraoperative bleeding. The tumour measured 27 × 8.5 × 6 centimetres and weighed 2000 grams. On gross examination of cut section of the tumour specimen, a large area of intra-tumoral haemorrhage was identified (**Figure 2**).

Histopathology reiterated angiomyolipoma of the left kidney as the definitive diagnosis. Postoperative period was uneventful and patient was discharged on postoperative day 5 (**Figure 3**). Patient followed up in outpatient department for 6 months and was found to be doing well. Patient gave her full and informed consent for publication of case report.

3. Discussion

Angiomyolipoma (AML) is a tumour consisting of thick-walled poorly organised blood vessels, smooth muscle s and varying levels of mature adipose tissue [1]. Renal angiomyolipoma (AMLs) account for 1% of all renal masses [2] with overall incidence in general population of 0.07% - 0.3% [3]. Giant angiomyolipoma which have a size greater than 10 centimetres [4] are rare entities with few reported cases in literature of AMLs measuring greater than 20 centimetres.

Up to 80% AMLs are sporadic. The classical sporadic presentation is of a middle-aged lady, suggesting a hormonal component to tumour growth, with a single asymptomatic tumour as was in this case [5]. AML may occur with tuberous sclerosis complex (TSC) in 20% - 30% patients and approximately 50% of patients with TSC develop angiomyolipoma [6].

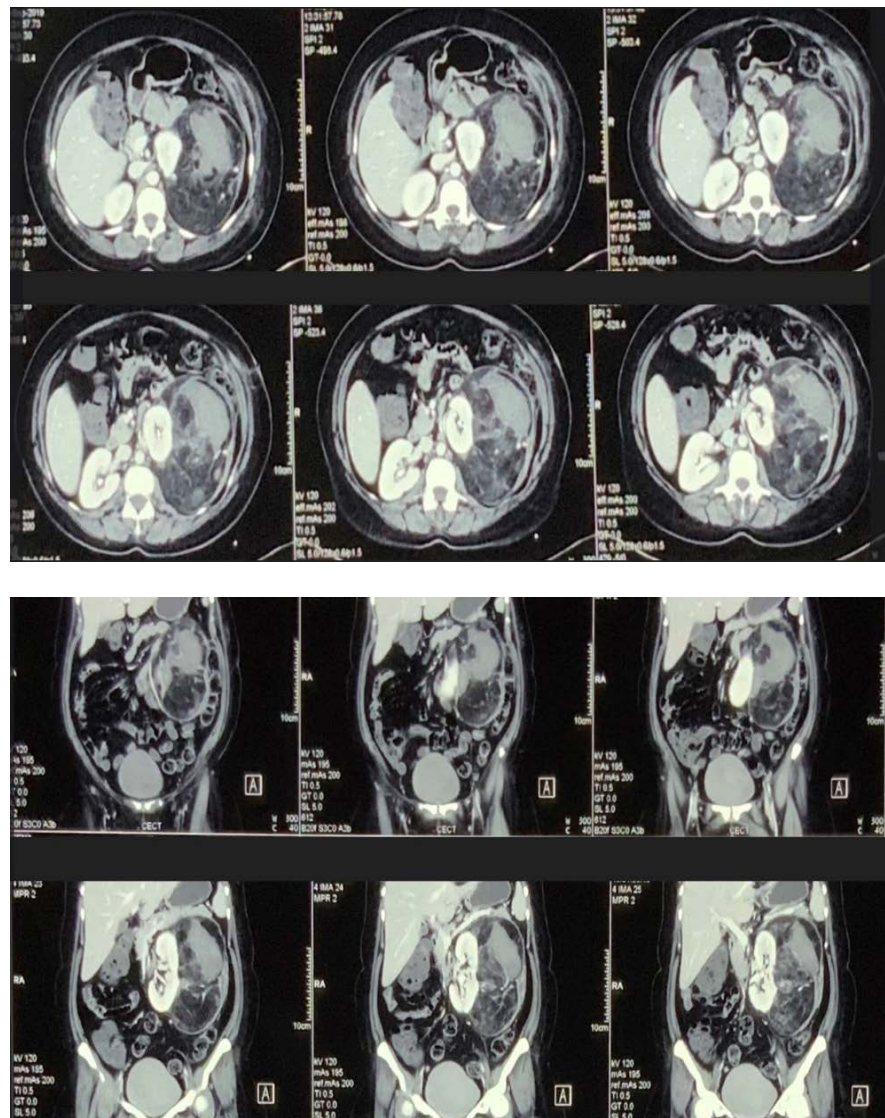


Figure 1. Axial and coronal views of contrast enhanced computed tomography showing large mass arising from left kidney laterally indenting its surface with negative 20 Hounsfield values (suggestive of fat) with a large area of hemorrhage within the tumour.

Asymptomatic tumours are usually less than 4 centimetres in size. Larger tumours tend towards symptomatic end of the spectrum which may include an abdominal mass, flank pain, haematuria, anaemia and haemorrhage. Life threatening intra-tumoral and retroperitoneal haemorrhage (Wunderlich syndrome) leading to shock may occur with large sized angiomyolipoma [7]. As seen in the presented case, wherein a giant angiomyolipoma presented with Wunderlich syndrome.

With the increased use of cross-sectional imaging over 80% of AMLs are now discovered incidentally, with haemorrhage at presentation (Wunderlich syndrome) seen in less than 15% and shock less than 10%. The classic triad of symptoms associated with renal masses of flank pain, palpable mass and haematuria were historically found in 37% - 41% of patients with AML [8].

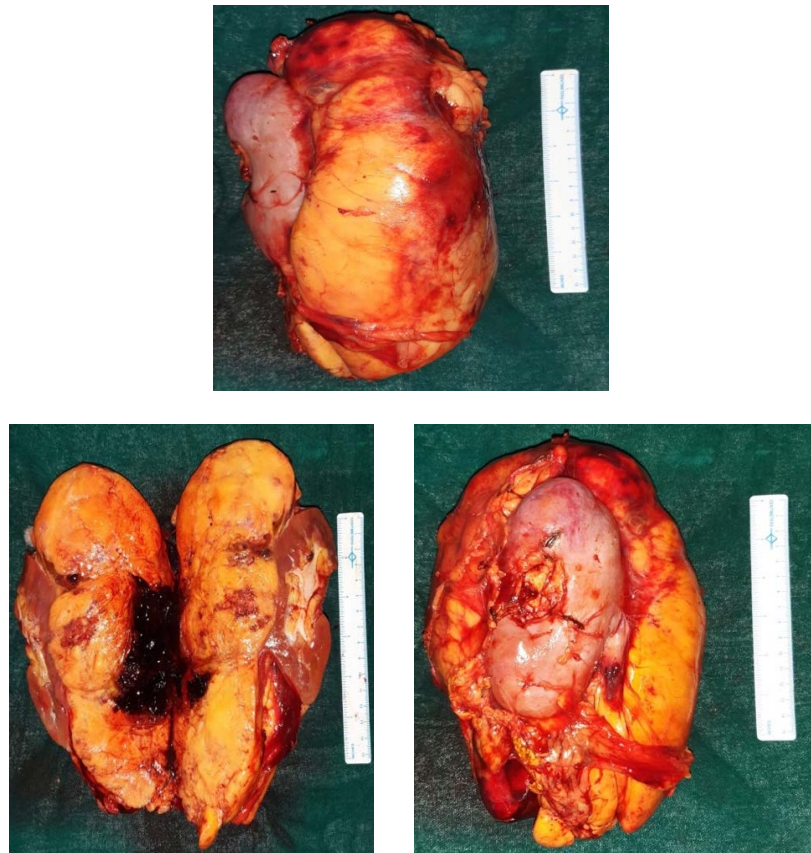


Figure 2. Left nephrectomy specimen showing giant angiomyolipoma arising from post-erolateral aspect of left kidney measuring $27 \times 8.5 \times 6$ centimetres, tumour is seen indenting the lateral surface. Cut-section through the kidney and tumour revealed a large area of intratumoural hemorrhage.

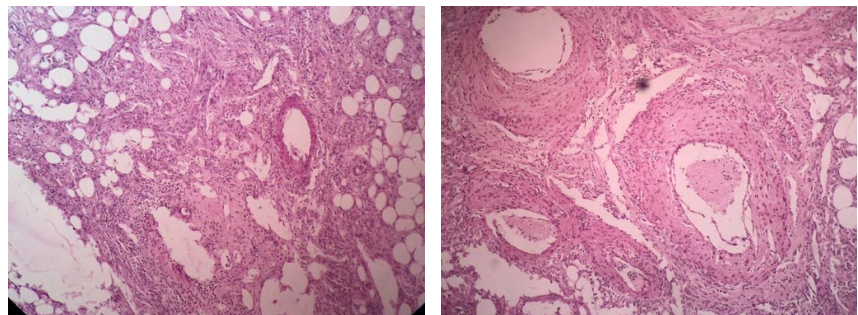


Figure 3. Histopathology sections demonstrating tumour composed of adipose tissue, thick walled blood vessels and smooth muscle. Vessel walls show prominent sclerosis.

Rare reports of angiomyolipoma mimicking renal cell carcinoma, Wilm's tumour and retroperitoneal liposarcoma are found in literature [9] [10] [11]. A few rare reports are present in which patients with ruptured angiomyolipomas presented with unexplained fever [12] [13]. In these cases, patients were known cases of bilateral angiomyolipomas with tuberous sclerosis and rupture of angiomyolipoma could be suspected. Angiomyolipoma mimicking a perinephric abscess as in our case report has not been reported in literature previously.

The diagnosis is usually apparent and definitive based on contrast enhanced computed tomography (CECT) scan with presence of fat in the lesion (confirmed by a value of -20 HU or less) being a diagnostic hallmark [14]. Possibility of a fat containing renal cell cancer (RCC), fat-poor angiomyolipoma and liposarcoma need to be considered and may warrant further evaluation using MR or percutaneous biopsy in doubtful cases [15]. MRI can be used to identify the fatty tissue. However, because the presence of bleeding in any renal tumour can mimic the typical pattern of angiomyolipoma, MRI should not be considered the diagnostic method of choice [4].

With regards to the present case report, the definitive diagnosis was established following CECT. The diagnosis of angiomyolipoma was missed on USG with haemorrhage within the tumour being misdiagnosed as perinephric abscess. CECT was deferred initially to correct acute kidney injury in the patient which was possibly due to hypovolemia associated with reduced intake.

Giant AMLs, as in our case, have been deemed to be at a higher risk of rupture due to formation of intralesional aneurysms with the size of intralesional aneurysms being a strong factor predictive of haemorrhage [8].

Treatment should be individualised depending on patient factors, tumour size and symptomatology. There is no one fits all modality with regards to angiomyolipomata management.

Small and asymptomatic tumours maybe managed conservatively with active surveillance. High risk of haemorrhage associated with large angiomyolipoma warrants intervention.

Conventionally, the criteria for intervention have been symptomatic lesions, size > 4 cm, suspicion for malignancy, women of childbearing age, an associated aneurysm size > 5 mm, concomitant TSC and poor access to follow up or emergency care as additional considerations for treatment [8].

Modalities may include partial or radical nephrectomy and/or selective embolization. Surgery in AML has progressed from initially recommended nephrectomy to nephron sparing surgery. This is especially important in AML associated with TSC as lesions are multifocal, bilateral and recurrent. Radiofrequency ablation is another modality which has shown some promise [8].

Newer modalities have emerged to the fore with enhanced and evolving understanding of molecular basis for angiomyolipomas. Mammalian target of rapamycin (mTOR) pathway inhibitors have emerged as a novel modality. There is a use for mTOR inhibitors in tuberous sclerosis patients with giant AMLs not amenable to other treatments or patients with less remaining renal reserve; however, questions as to the durability of responses, duration of treatment and impact of toxicity from chronic therapy remain. Additionally, the role of mTOR inhibitors in the management of patients with sporadic associated giant AMLs, as in the presented case, remains to be determined [8] [16] [17].

In our case, intervention following optimisation was warranted in view of the large sized tumour and evidence of haemorrhage. Selective angioembolisation was attempted preoperatively with the intention to minimise intraoperative

bleeding. However, embolization was unsuccessful. The patient underwent left simple nephrectomy.

Partial or nephron sparing nephrectomy and radio ablation could not be undertaken owing to the tumour encasing the whole kidney and intraoperative haemorrhage. There have been case reports of treating giant angiomyolipomas with nephron sparing surgery or excision of the tumour especially in cases of bilateral disease or solitary kidney in the settings of tuberous sclerosis [18]. Since there is further need to evaluate these approaches and our patient didn't have these special circumstances, we proceeded with the standard protocol.

4. Conclusion

To summarize, renal angiomyolipoma is a rare benign tumour mostly discovered as an incidental finding. The giant sized angiomyolipoma masquerading as perinephric abscess in our case report is a unique presentation, not reported previously. There is also established a need for further evaluation of other modalities of treatment in sporadic cases of giant AML with better outcomes and maximal organ preservation.

Acknowledgements

None.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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