

## Case Report

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# A giant renal angiomyolipoma: a rare case report and review of literature

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## ABSTRACT

Renal angiomyolipomas (AMLs) are the most common benign tumours of the kidney that occur sporadically in 80% of the patients and are seen associated with genetic conditions such as tuberous sclerosis in rest of the patients. The diagnosis of renal AML pre-operatively have been made easier with the advent of good imaging modalities. The management of these tumours depends on factors such as size, clinical presentation, extent of parenchymal involvement and ranges from a wait and watch strategy to a radical nephrectomy. Herewith, we reported of a giant renal AML who presented with complaints of lump in abdomen with headache and palpitations. She was diagnosed to have a giant renal AML on imaging. The patient was managed with radical nephrectomy after renal AML. A brief case report with review literature was presented here.

**Keywords:** Giant renal angiomyolipoma, Renal angioembolization, Radical nephrectomy

## INTRODUCTION

Renal AMLs are benign tumours of the kidney composed of smooth muscle, adipose tissue and thick walled vessels. These tumours have a strong female predilection and affect middle aged women more commonly.<sup>1</sup> They are inherited as an autosomal dominant condition. AMLs are believed to originate from the perivascular endothelial cells, therefore are also referred to as PEComas.

AMLs are observed to increase in size every year by 4 cm in its greatest dimension. When AMLs grow beyond 10 cm in the greatest dimension, they are called giant AMLs.<sup>2</sup> These are known to present with complications such as compression of renal artery, haemorrhage. Hence the management of these tumours was of paramount importance. With the advent of advanced radiological scans, the identification of these tumours pre-operatively have been made easier which had led to effective

management of these cases. The decision regarding management of giant renal AMLs by RAE alone or surgical excision depended on clinical presentation, size and various other factors.

## CASE REPORT

A 34 year old female patient presented with pain in abdomen since 3 months and complaints of intermittent attacks of palpitations, headaches and giddiness since 2 months. The pain in abdomen was dull aching, more in the right lumbar region with no radiation or referral. She also had complaints of intermittent attacks of palpitations and headache which lasted for a period of 10-15 minutes, associated with giddiness. These attacks occurred 2-3 times a week and she was managed conservatively for the same.

On admission to the hospital, her vital parameters were stable.

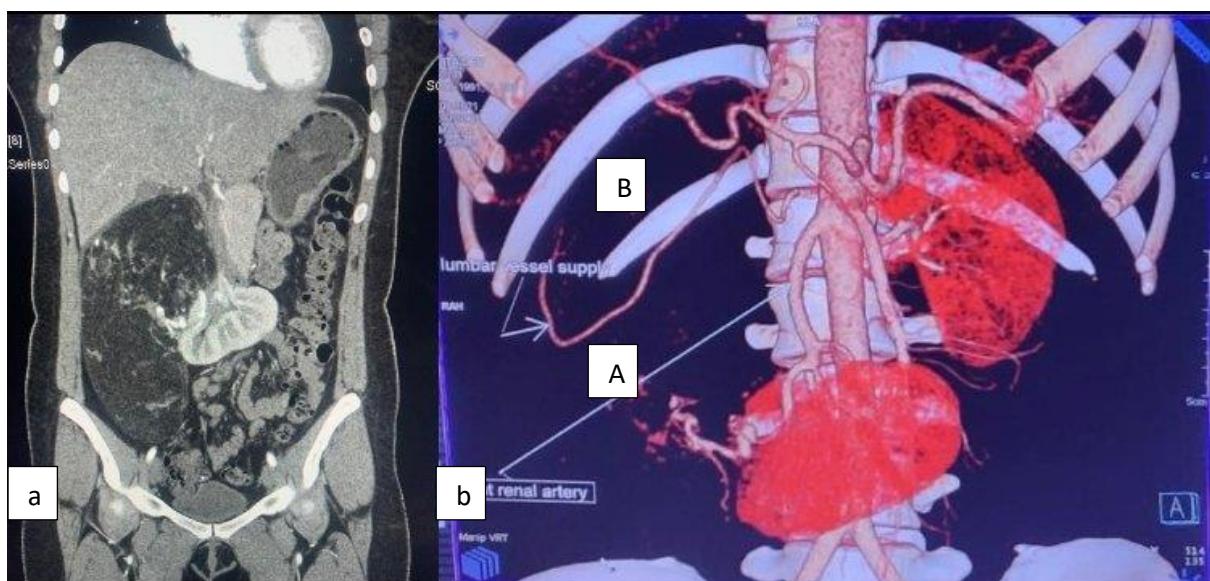
Abdominal examination revealed a firm, non-tender, ballotable mass in the right lumbar region measuring approximately 15x10 cm which moved with respiration.

Ultrasound of the abdomen done showed an echogenic fat containing lesion measuring 21x10 cm occupying the right lumbar and right hypochondriac region with medial and inferior displacement of the right kidney. The lesion showed internal vascularity with a few vessels extending from the superior pole of the right kidney into the lesion.

Plasma free metanephrenes, urine 24 hour metanephrenes and urinary vanillylmandelic acid was done which was within normal limits.

Contrast enhanced computed tomography (CECT) of the abdomen and pelvis was done which showed a well-

defined circumscribed solid mass with predominant fatty density, measuring approximately 9.7x15.2x19.8 cm (APxTRAxCC) in the right suprarenal and right subhepatic space. The mass was seen to be displacing the kidney inferomedially with the kidney now placed in the midline in the umbilical region (Figure 1a). Soft tissue component of the lesion showed post contrast enhancement. Cortical breach was noted at the upper pole of the right kidney with part of the lesion extending into it. Right suprarenal gland was not seen separately from the lesion. The vascular pedicle supplying the tumour was seen passing through the renal parenchyma into the tumour through the capsular breach (Figure 1a). This vessel was seen to be a direct branch of the right renal artery (Figure 1b). Based on the above finding, a probable diagnosis of renal AML was made.



**Figure 1: CECT abdomen and pelvis images; (a) Coronal section of CECT showing a large tumour with HU of 65 HU suggestive of fatty component in the lesion; (b) 3D reconstruction of the coronal section of the CECT image showing the vascular supply of the tumour; (A) right renal artery; (B) lumbar artery and the tortuous intra parenchymal course of its vascular pedicle; the white arrow indicates the feeder vessel.**

Patient then underwent angioembolisation of the tumour. On doing a right renal angiogram, a hypervascular mass was found in the right kidney with feeders directly arising from the right renal artery (Figure 2). The major dysplastic feeder was selectively cannulated and embolization was carried out using PVA particles.

Within 48 hours of angioembolisation, patient was taken up for surgical excision of the tumour under general anaesthesia. Intra-operatively, it was observed that the tumour involved the superior pole of the right kidney (Figure 3). It was receiving its major blood supply through a hypertrophied vessel originating from the right renal artery. This main vascular supply was seen entering the tumour through the breech in the renal capsule. Involvement of the renal parenchyma was seen intra

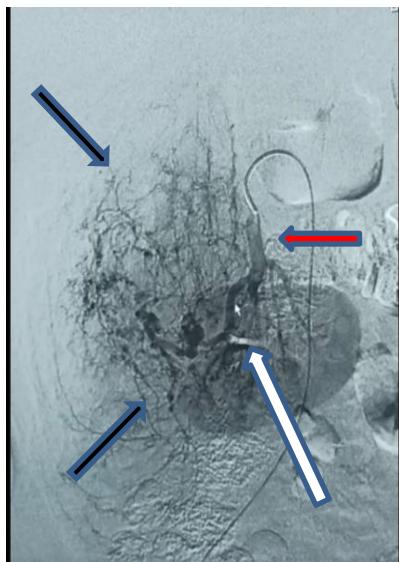
operatively with multiple tiny collateral entering the parenchymal part of the lesion. Hence, intra-operative decision was taken to proceed for a radical nephrectomy.

The excised tumour measured 20 cmx16 cmx6 cm (CCxTRxAP) and weighed 450 gms.

On gross examination of the tumour, there were large fatty areas with focal and central areas of haemorrhage within (Figure 4a). It was confirmed that the feeding vessel to the tumour was passing through the renal parenchyma into the tumour through a capsular breech (Figure 4b).

Post operative period was uneventful.

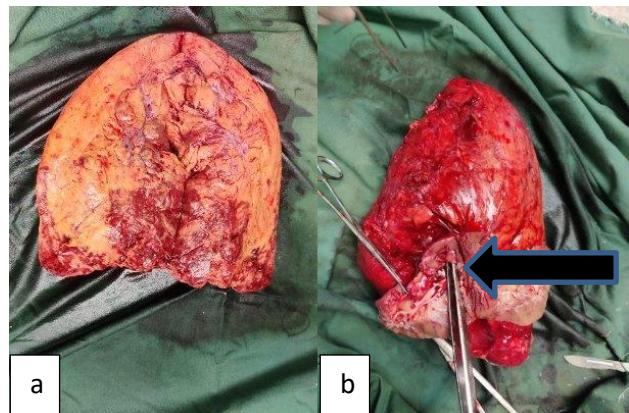
Histopathology of the tumour showed features of a renal angiomyolipoma with dysmorphic vessels, myogenic spindles and adipocytes. Tumour cells were positive for human melanoma black-45, smooth muscles actin, desmin and focally positive for melan A.



**Figure 2: Right renal angiogram; the red arrow indicates the right renal artery; the white arrow indicates the vascular pedicle of the tumour that was selectively cannulated and embolized; the black arrow indicates the extensive vascular blush seen within the tumour indicating that the major blood supply of the tumour was from a direct branch of the right renal artery.**



**Figure 3: Tumor is seen involving the upper pole of the right kidney.**



**Figure 4: Gross sections of the resected tumour; (a) gross section of the tumour showing excessive fatty areas with areas of central haemorrhage within; (b) gross image of the tumour after dividing the kidney showing the course of the vessel supplying the tumour; the black arrow indicates the forceps that is inserted into the vessel.**

The patient was discharged with advice to follow up with sonography of the abdomen after 3 months.

## DISCUSSION

The prevalence of renal AMLs vary from 0.2% to 0.6% with 0.6% in the female and 0.28% in the male population.<sup>3</sup> The tumors, though rare, may be aggressive and may present with complications, hence their management was of paramount importance.

A large proportion of renal AMLs were detected incidentally on imaging and were silent. Tumors less than 4 cm were usually managed conservatively with periodic evaluation, while tumors of 4-8 cm were managed with close monitoring for symptoms and intervention was done when there was suspicion for progression in size and malignancy.<sup>4</sup> Indications for intervention included tumors more than 8 cm, increasing size ( $>4$  cm), symptomatic cases, haemorrhage and associated decreased renal function.<sup>5</sup>

In the index case, the initial symptom complex led us to making a clinical diagnosis of an adrenal tumour. However, radiological imaging and normal plasma metanephhrines led us to the final diagnosis of a renal AML.

The two main strategies to manage this tumour were surgery and renal angioembolisation (RAE). Various randomised control studies have been conducted comparing the outcomes of the above two strategies. While none of them defined clear guidelines for management, it had been observed that the requirement for repeated ablative procedures have been observed in a few cases managed by RAE alone. A randomised control study was conducted in 2011 on 59 patients with a renal

angiomyolipoma, out of which 17 patients underwent RAE alone. It was observed that the tumour decreased in size by 50-60% only on follow up and 14% of these patients required repeated RAE sessions.<sup>5</sup> The incidence of haemorrhage following RAE was also a recognized complication. RAE was also observed to be effective in patients presenting with acute haemorrhage or with a single feeding vessel to the tumour as compared to cases with multiple feeding branches or collaterals to the tumour.<sup>5</sup>

Similar studies also concluded that surgical excision of the tumour was superior to RAE as it offered complete removal of the tumour, less chances of a recurrence and lower rates of repeated procedures.<sup>6,7</sup> A systemic review of the data on management of renal angiomyolipomas from 1990-2017 was done which concluded that recurrence occurred in 4-15% of patients treated by surgery and in 6-39% of patients treated by selective arterial embolization (SAE). According to the systematically reviewed data, the most frequently reported effective treatment was surgery (31%), particularly nephron sparing surgery (NSS), followed by SAE (17%). Both have similar morbidity, but there were more recurrences and need for secondary treatment in patients treated by SAE alone. (0.85% in surgery versus 31% in SAE).<sup>7</sup>

Various case reports of similar cases confirmed that a pre-operative angioembolisation of the tumour was beneficial in terms of decreased blood loss intra operatively.<sup>7,8</sup> Also, there was formation of edema surrounding the tissues post embolization, which facilitated easy dissection and hence decreased operative time. However, occurrence of post embolisation tumour lysis syndrome in these patients should be kept in mind. Pre-operative angioembolisation evoked immune-modulation and proliferation of cytotoxic T cells.<sup>9</sup> This led to tumour lysis with resultant systemic circulation of tumour antigens.<sup>9</sup> Tumour lysis syndrome occurred as a result of an exaggerated immune system response to the exposed tumour antigens. The symptom complex included fever, vomiting, severe flank pain, tachycardia and elevated inflammatory markers. They were usually managed with NSAIDs and anti-pyretic agents. Hence, surgical intervention should be planned prior to occurrence of tumour lysis syndrome.

The diagnosis in the index case was that of a giant renal AML with compression on the renal pedicle and adrenal gland causing unusual symptoms such as episodic headache and palpitations. Hence, decision to perform a surgical excision of the tumour with pre-operative SAE was taken.<sup>5</sup> Studies confirmed that RAE led to inflammation of the tissues surrounding the tumour. This resultant tissue damage led to adhesion formation between the tumour and the surrounding tissues. Also, recanalization of the embolised vessel or collateral formation to the tumour had been observed in various studies. The above two mechanisms begin after 72 hours of embolisation and was maximum during 10 days to 2

weeks.<sup>10</sup> Hence, the surgery was planned within 48 hours of embolisation of the tumour.

Patients with renal AMLs can be managed surgically with nephron sparing surgery or simple nephrectomy depending on various pre operative and intra-operative factors.<sup>11</sup> The primary deciding factor regarding the line of management depended on the preoperative imaging techniques. Advanced imaging techniques offered information about the size, fat content, vascular supply of the lesion and extent of involvement of the renal parenchyma and renal vein visible.

Pre-operative factors favouring nephron sparing surgeries included tumour with increased fat components on computed tomography (CT), minimal breach in renal capsule, adequate renal function and a single discrete vascular supply that can be easily dissected off from the renal parenchyma and the renal pedicle. Patients with bilateral giant AMLs were also managed with nephron sparing surgery preferably to preserve adequate renal function.<sup>12</sup> AMLs with epithelial components and showing minimal fat on CT were known to be malignant AMLs, hence radical nephrectomy was advised in these cases.<sup>13,14</sup> Radical nephrectomy was also performed in patients with a single non-functional kidney or in cases where there was an increased involvement of the renal parenchyma.<sup>15</sup> Radical nephrectomy was also performed in cases where the tumour was seen to receive a complex vascular supply making it difficult to perform a nephron sparing surgery.

In this case, although the renal function was adequate, imaging showed that the vascular pedicle supplying the tumour was seen arising as a direct branch of the right renal artery. This feeding vessel was seen passing through the renal parenchyma, through the breach in the renal capsule into the tumour. Multiple small collaterals were seen from this major vascular pedicle entering the tumour. Intra-operatively, it was also noted that the tumour involved the renal parenchyma through the breach at its upper pole. Hence, decision was taken to perform a radical nephrectomy in view of the above pre operative and intra operative findings.

Guidelines about management of giant AMLs were still being studied. Giant AMLs were best managed with SAE followed by surgical excision. Pre-operative imaging was the most critical factor in management of renal AMLs. The anatomical course of the vascular supply and the extent of involvement of renal parenchyma seen intra-operatively was vital in deciding on preservation of the involved kidney.

## CONCLUSION

Renal AMLs are common benign tumours of the kidney which commonly occur sporadically. Giant AMLs (>10 cm) are diagnosed 95% of the times with advanced radiological imaging techniques. RAE alone without

surgery in these cases have a high risk of haemorrhage, recurrence and require repeated sessions. Hence the ideal management of giant AMLs is SAE followed by surgical excision of tumour with or sparing of the involved kidney. The management of these cases depend on pre operative imaging and intra operative findings, while the functioning of the involved kidney is also of utmost significance.

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