

# Giant Renal Angiomyolipoma: Unusual Cause of Huge Abdominal Mass

Rajesh Taneja, Dig Vijay Singh<sup>1</sup>

Departments of Urology, Indraprastha Apollo Hospitals, New Delhi, <sup>1</sup>Mohandai Oswal Hospital, Ludhiana, Punjab, India

**Address for correspondence:**

Dr. Dig Vijay Singh,  
 Department of Urology, Mohandai Oswal Hospital, Ludhiana - 141 010, Punjab, India.  
 E-mail: [digvijayaiims@yahoo.com](mailto:digvijayaiims@yahoo.com)



Received: 12-08-2013  
 Accepted: 21-10-2013  
 Published: 28-11-2013

## ABSTRACT

We present the imaging and histopathological characteristics of a giant renal angiomyolipoma (AML) in a 49-year-old female patient, who presented with bloating sensation in the abdomen and a steadily increasing abdominal girth for about 3 years. Contrast enhanced computed tomography (CECT) scan films of abdomen revealed that a large fat containing tumor had replaced the left kidney while displacing the rest of the abdominal contents toward the other side of the midline. Intraoperatively the left kidney was completely replaced by a fat containing tumor. The recovered surgical specimen measured 39 cm × 25 cm × 9 cm and weighed 7500 g. Histopathological investigation with immuno-histochemical staining of the specimen with hydroxy beta-methylbutyric acid-45 confirmed this lesion as AML. CECT scan of the head did not show any lesion suggestive of tuberous sclerosis. The giant tumor of the present case is the heaviest AML in both syndromic and sporadic categories and largest by dimensions as sporadic AML ever reported in the literature.

**Key words:** Angiomyolipoma, giant kidney tumor, heaviest renal angiomyolipoma, sporadic tuberous sclerosis

## INTRODUCTION

Renal angiomyolipoma (AML) is an infrequent tumor that, in most cases, follows a benign course and has clearly defined radiological and histological characteristics.<sup>[1]</sup> We present a case of giant renal AML, which is the heaviest ever reported, encountered in a tertiary care hospital in India.

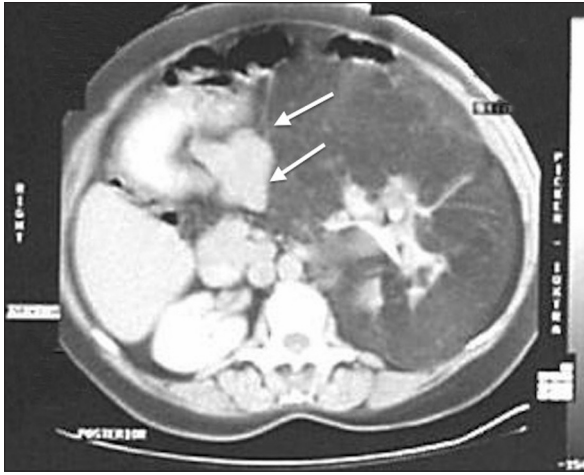
## RADIOLOGIC FEATURES

A 49-year-old lady, a native of Afghanistan, presented with a 3-year history of steadily increasing abdominal girth and bloating sensation in the abdomen. She also experienced intermittent dull ache in the left flank. She underwent investigation in another institute and was found to have a huge abdominal mass. Review of contrast enhanced computed tomography (CECT) scans of abdomen revealed a large fat containing tumor measuring 40 cm × 24 cm × 10 cm (<–20 Hounsfield Units), replacing the left kidney and displacing the rest of the abdominal contents toward the other side of the midline [Figure 1]. There was absence of enhancement, inhomogeneity, necrosis, or calcification ruling out the possibility of malignancy. A provisional diagnosis of benign renal

Access this article online	
Quick Response Code:	Website: <a href="http://www.clinicalimagingscience.org">www.clinicalimagingscience.org</a>
	DOI: 10.4103/2156-7514.122326

Copyright: © 2013 Taneja R. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

This article may be cited as:  
 Taneja R, Singh DV. Giant Renal Angiomyolipoma: Unusual Cause of Huge Abdominal Mass. J Clin Imaging Sci 2013;3:56.  
 Available FREE in open access from: <http://www.clinicalimagingscience.org/text.asp?2013/3/1/56/122326>



**Figure 1:** 49-year-old lady with complaints of bloating sensation in the abdomen and a steadily increasing abdominal girth, which was subsequently diagnosed as angiomyolipoma of left kidney. Contrast enhanced computed tomography scan axial section film of abdomen reveals a lipomatous neoplasm 40 cm × 24 cm × 10 cm in dimension (HU < -20 HU) replacing the entire left kidney, while displacing the rest of the abdominal contents (arrows) toward the other side of the midline.

neoplasm, possibly AML causing pressure effects was made. Hence, CECT scan of the head was done, which excluded any lesion suggestive of tuberous sclerosis (TS) [Figure 2] and decision of to perform nephrectomy was taken.

## PATHOLOGIC FEATURES

Under intubated general anaesthesia, an abdominal midline incision was made extending from xiphisternum to just above pubic symphysis. The left kidney was seen to be completely replaced by a fat containing tumor. Simple nephrectomy was performed with intact tumor retrieval and minimal blood loss, without necessitating any blood transfusion.

The recovered surgical specimen measured 39 cm × 25 cm × 9 cm and weighed 7500 g with compressed renal hilar vessels and ureter [Figure 3]. Grossly, it was well-circumscribed and with a glistening yellow ("fatty") appearance. Histopathological examination of the specimen revealed majority of the cells were mature adipocytes associated with thick-walled blood vessels and a few epithelioid stromal cells. There was a pattern with typical fat and perivascular epithelioid cells arranged around a blood vessel, suggesting it to be AML [Figure 4]. Immuno-histochemical staining of the tissue by beta-hydroxy beta-methylbutyric acid-45 (HMB-45) (a melanosome-associated protein) indicated the presence of epithelioid component [Figure 5].

The patient recovery was uneventful and she was discharged from the hospital on the 5<sup>th</sup> post-operative

day. At 6 months post-operative follow-up, no evidence of tumor recurrence was found.

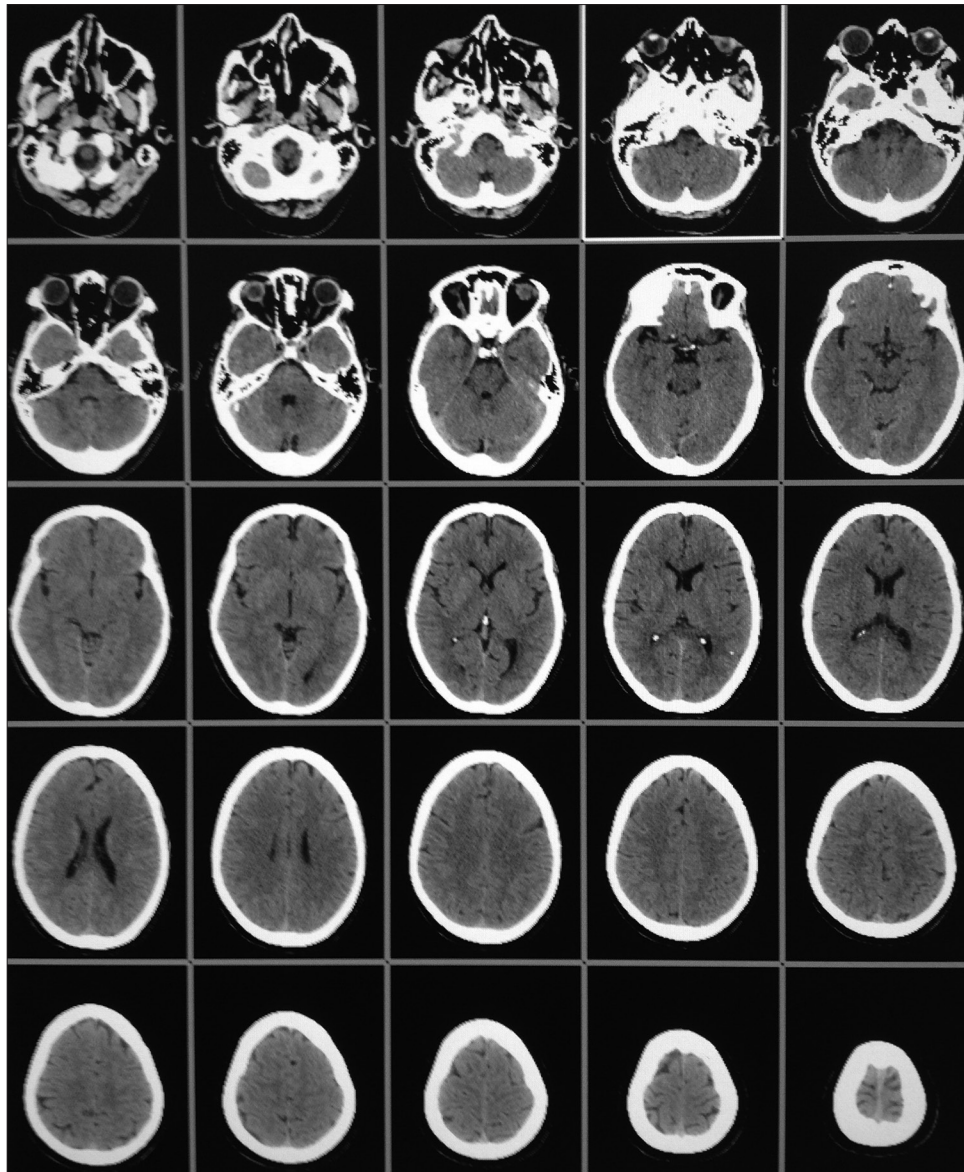
## DISCUSSION

AML is a benign hamartous lesion consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels. Renal AML are benign tumors known to occur sporadically and in association with genetic syndromes like TS and lymphangioleiomyomatosis.<sup>[2]</sup> However, in our patient there were no clinical or radiological signs suggestive of any systemic syndrome.

According to Vitaly et al., an estimated 20-30% of AMLs are found in patients with TS syndrome, which is an autosomal dominant disorder characterized by mental retardation, epilepsy, and adenoma sebaceum. Conversely, approximately 50% of patients with TS develop AMLs. In these cases, mean age at presentation is 30 years, and there is a 2:1 female-to-male predominance. However, 70-80% of patients with AML who do not have TS, there is a more pronounced female predominance and most patients present later in life, during the fifth or sixth decade,<sup>[3]</sup> as was in the present case. Although, renal AML in patients with TS can be associated with significant morbidity, mostly related to complications from bleeding, sporadic cases do occur mostly as incidental lesions. The detection of sporadic cases has increased due to greater use of abdominal imaging for the evaluation of a wide variety of non-specific complaints. Occasionally, the increase in abdominal swelling<sup>[4]</sup> or epigastric fullness<sup>[5]</sup> may be the presenting feature, as in our case.

AML is the only benign renal tumor that is confidently diagnosed on cross-sectional imaging. The presence of fat (confirmed on non-enhanced thin-cut computed tomography by a value of -20 [HU] or less) seen within a renal lesion on imaging is considered the diagnostic hallmark. Findings of more than 20 pixels with attenuation less than -20 HU and of more than 5 pixels with attenuation less than -30 HU have been shown to have a positive predictive value of 100%.<sup>[3]</sup> Histopathologically, AML consists of mature adipocytes, thick-walled blood vessels, and epithelioid stromal cells in various proportions. Usually it displays as a pattern of typical fat and perivascular epithelioid cells arranged around a blood vessel.<sup>[1]</sup> Positive immunoreactivity for HMB-45, a monoclonal antibody raised against a melanoma-associated antigen, is characteristic for AML and can be used to differentiate this tumor from sarcoma and other tumors.<sup>[3]</sup>

The indication for surgical treatment is usually a symptomatic AML or an incidental AML of size larger than 4 cm. In cases



**Figure 2:** 49-year-old lady with complaints of bloating sensation in the abdomen and a steadily increasing abdominal girth, which was subsequently diagnosed as angiomyolipoma of left kidney. Contrast enhanced computed tomography scan axial section films of the head reveals no lesion suggestive of tuberous sclerosis.

of bilateral lesions, as in TS, nephron-sparing surgery (by either selective embolization or open or laparoscopic/robotic partial nephrectomy) must be performed.<sup>[3]</sup> In our case, the tumor was so large that there was no option but to excise the whole lesion in totality.

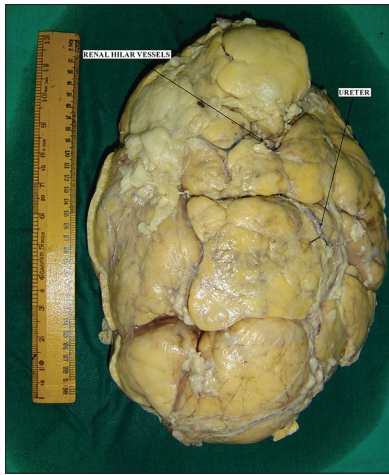
On review of cases in the literature, we find our case represents the heaviest AML (7500 g) ever reported, both in TS and sporadic categories. This giant lesion surpasses the AML mass weighing 6300 g in a case associated with TS reported by Rosselló Barará et al.,<sup>[6]</sup> and the lesion (weighing 3500 g each) in the cases reported by Tsutsumi et al.,<sup>[5]</sup> and Katz and Poster.<sup>[7]</sup> The only other AML, which has dimensions larger than the present one (39 cm × 25 cm × 9 cm) has been reported by Katz and Poster, where the mass measured

45 cm × 20 cm × 15 cm and weighed 3500 g (weight less than 7500 g measured in the present case), and was associated with TS.<sup>[7]</sup> Thus the present case is the largest sporadic renal AML in size and heaviest renal AML in both syndromic and sporadic categories ever reported in the literature.

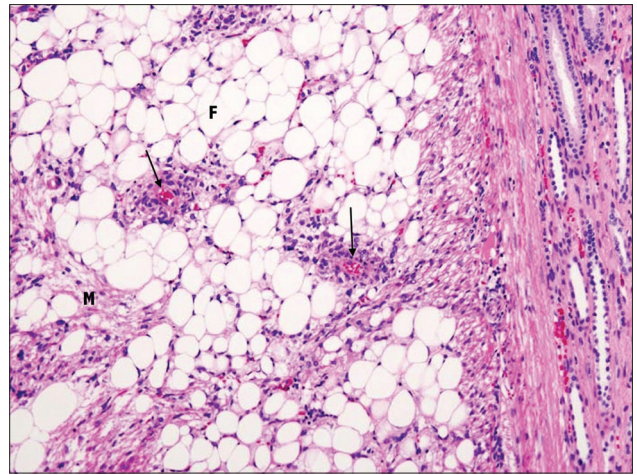
## CONCLUSION

AML can be identified when on CT image more than 20 pixels show attenuation less than -20 HU and if more than 5 pixels show attenuation less than -30 HU. Histologically, AML lesions show a pattern of typical fat and perivascular epithelioid cells arranged around a blood vessel. Positive immuno-histochemical staining by HMB-45 (a melanosome-associated protein) for epithelioid

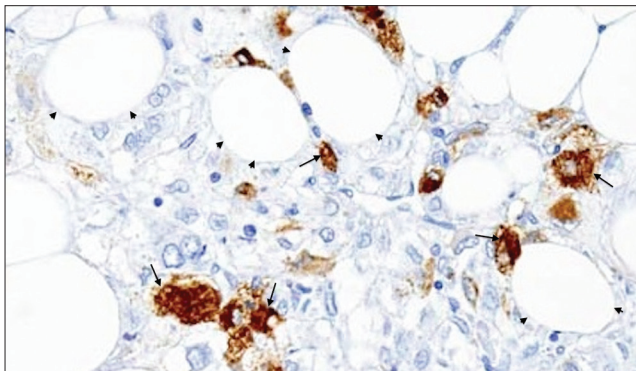




**Figure 3:** 49-year-old lady with complaints of bloating sensation in the abdomen and a steadily increasing abdominal girth which was subsequently diagnosed as angiomyolipoma (AML) of left kidney. Excised specimen shows a giant AML 39 cm × 25 cm × 9 cm in dimension and weighing 7500 g, replacing the whole kidney. Renal hilar vessels and ureter are compressed, and emerging out of the mass.



**Figure 4:** 49-year-old lady with complaints of bloating sensation in the abdomen and a steadily increasing abdominal girth, which was subsequently diagnosed as angiomyolipoma of left kidney. Histopathology slide of the excised mass shows admixture of tortuous thick-walled blood vessels (arrows), sheets of mature adipose tissue (F) and bundles of smooth muscle fibres (M) (Stain used hematoxylin and eosin, ×100).



**Figure 5:** 49-year-old lady with complaints of bloating sensation in the abdomen and a steadily increasing abdominal girth, which was subsequently diagnosed as angiomyolipoma of left kidney. Photomicrograph (×40) of immunohistochemical stained specimen shows mature adipocytes (arrow heads) and strong staining of the epithelioid cells with the beta-hydroxy beta-methylbutyric acid-45 (arrows).

component confirms the lesion as AML. We present a case where the patient presented with a huge abdominal mass in the left kidney diagnosed as AML and was managed by left nephrectomy. The giant tumor is the heaviest AML ever reported in the literature. At 6 months post-operative follow-up, there was no evidence of tumor recurrence.

## REFERENCES

1. Serrano Frago P, Del Agua Arias Camisón C, Gil Sanz MJ, Allué López M, Gonzalvo Ibarra A, Plaza Mas L, et al. Controversies related to epithelioid variant of renal angiomyolipoma: A review of the literature. *Urology* 2006;67:846.e3-5.
2. Hsu TH, O'Hara J, Mehta A, Levitin A, Klein EA. Nephron-sparing nephrectomy for giant renal angiomyolipoma associated with lymphangioleiomyomatosis. *Urology* 2002;59:138.
3. Vitaly M, Surena FM, Christopher GW. Benign renal tumors. In: Campbell-Walsh, *Urology*. 10<sup>th</sup> ed. Philadelphia: Saunders Elsevier; 2012. p. 1498-501.
4. Lee YC, Huang SP, Liu CC, Wu WJ, Chou YH, Huang CH. Giant extrarenal retroperitoneal angiomyolipoma: A case report and literature review. *Kaohsiung J Med Sci* 2003;19:579-82.
5. Tsutsumi M, Yamauchi A, Tsukamoto S, Ishikawa S. A case of angiomyolipoma presenting as a huge retroperitoneal mass. *Int J Urol* 2001;8:470-1.
6. Rosselló Barbará M, Mus Malleu A, Rebassa Llul M, Casals Armada J. Giant renal angiomyolipoma disclosed by mild lumbar trauma and treated with nephrectomy: Report of a case. *Actas Urol Esp* 2004;28:133-7.
7. Katz DS, Poster RB. Massive renal angiomyolipoma in tuberous sclerosis. *Clin Imaging* 1997;21:200-2.

**Source of Support:** Nil, **Conflict of Interest:** None declared.