

Giant Angiomyolipoma Associated With a Dilated Vessel Prone to Hemorrhage

Shagufta Wahab,¹ Rizwan Ahmad Khan,² Manjari Thapa,¹ Arif Wahab,³ Ibne Ahmad¹

¹Department of Radiodiagnosis, Jawaharlal Nehru Medical College Hospital, Aligarh, India

²Division of Pediatric Surgery, Department of Surgery, Jawaharlal Nehru Medical College Hospital, Aligarh, India

³Center of Cardiology, Jawaharlal Nehru Medical College Hospital, Aligarh, India

Angiomyolipomas are benign neoplasms of the kidney which can clinically mimic renal malignancy. The imaging features are quite characteristic, and diagnosis can be clinched solely based on radiological investigations. Although mostly asymptomatic, they can be a cause of significant morbidity due to size and hemorrhage. We report a case of giant angiomyolipoma with a dilated vessel susceptible to hemorrhage.

IJKD 2009;3:168-71
www.ijkd.org

Keywords. hamartoma, angiomyolipoma, tuberous sclerosis, computed tomography

INTRODUCTION

Hamartoma is a benign tumor consisting of disorganized, yet mature, tissue. Renal hamartomas include angiomyolipoma, leiomyoma, lipoma, myolipoma, fibroma, and mesoblastic nephroma.¹ Although angiomyolipoma is rare, it is the most frequently reported renal hamartoma.¹ These are mostly asymptomatic and are discovered incidentally on imaging with ultrasonography, computed tomography (CT), or magnetic resonance imaging.² We present a giant exophytic angiomyolipoma with a visible prominent and dilated vessel, suggesting the risk of severe hemorrhage. The rarity of the size and the fact that we were able to predict the risk of hemorrhage on CT scan prompted us to report the case.

CASE REPORT

A 48-year-old woman presented to the outpatient department of our cardiology center with complaints of palpitation, swelling in the lower limbs, vague abdominal pain, and tenderness on the left side of the abdomen for 2 months. Routine investigations were carried out including ultrasonography, which showed a large echogenic mass with posterior acoustic shadowing appearing to arise from the left kidney. Contrast-enhanced CT revealed a well-

defined large exophytic mass sized approximately 12 × 10 × 15 cm arising from the anterolateral cortex of the kidney with CT-attenuation value of fat (Figure 1). Based on the CT image, a diagnosis of large angiomyolipoma was made.

The patient then underwent magnetic resonance imaging, and the mass was found to be hyperintense on both T1- and T2-weighted images with suppression on short tau inversion recovery sequences. The signal intensity of the lesion matched



Figure 1. Contrast-enhanced computed tomography scan shows well-defined exophytic mass in the left kidney with an attenuation value of fat with a prominent vessel traversing the lesion.

with that of mesenteric and subcutaneous fat. The lesion appeared to be very well defined, growing in an exophytic pattern from the anterolateral cortex of the kidney (Figures 2 to 4). Also seen were multiple enhancing vessels with a diameter of approximately 7 mm, traversing through the mass, which indicated that the mass was likely

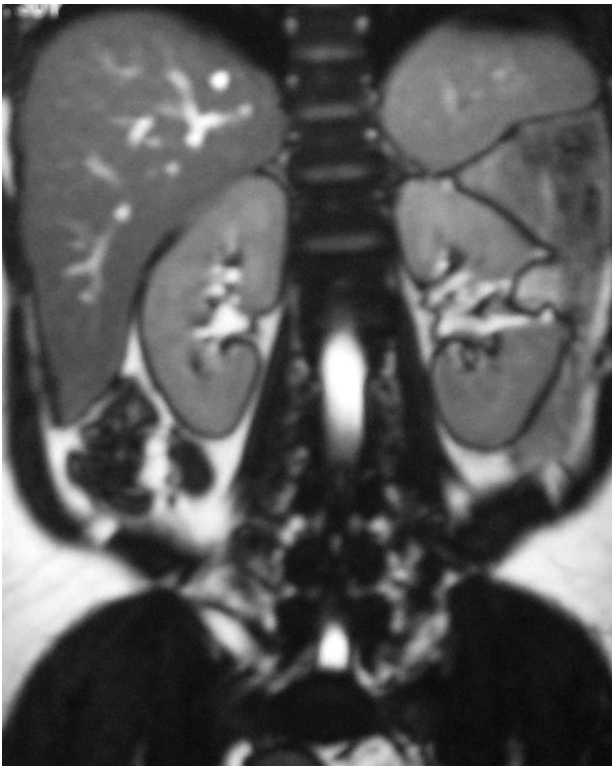


Figure 2. Coronal magnetic resonance image shows the exophytic lesion arising from the anterolateral cortex of the left kidney.

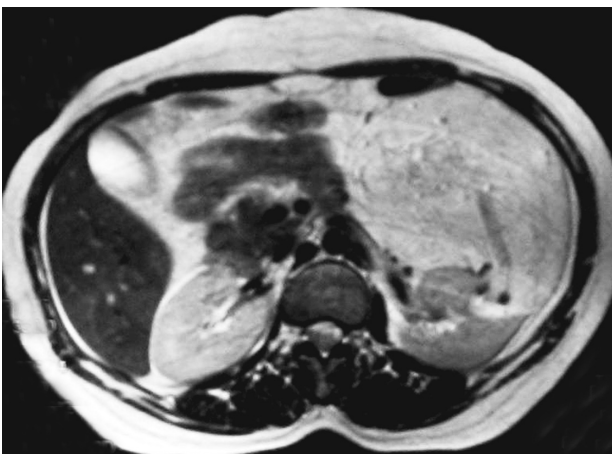


Figure 3. Axial T2-weighted magnetic resonance image shows the signal intensity of the lesion similar to the mesenteric and subcutaneous fat tissues. The signal void due to the prominent vessel can be well appreciated in the image.



Figure 4. Contrast-enhanced fat saturation magnetic resonance image shows suppression of the signal intensity of the lesion, confirming its fatty nature.

to present with hemorrhage in the future. The patient was referred to the department of surgery for further evaluation and management, where she underwent total excision of the tumor (nephron-sparing of the kidney). The histopathology report of the lesion revealed nonencapsulated but well-margined lesion with abundant adipocytes on hematoxylin-eosin staining. Adjacent to the tumor, varying amounts of smooth muscle, blood vessels, and adipocytes were seen.

DISCUSSION

Angiomyolipoma was named so by Morgan and colleagues in 1951, as they found it a complex of mature adipose tissue, smooth muscle cells, and thick-walled vessels.³ With an overall prevalence of approximately 0.3% to 3%, angiomyolipoma of the kidney is quite uncommon.¹ This tumor can develop in 2 forms that are otherwise histologically similar. Almost 80% of the cases develop sporadically, typically in middle-aged women, and the other 20% are associated with tuberous sclerosis, also known as *Bourneville disease*.¹ Tuberous sclerosis is an autosomal dominant disease that classically diagnosed as a triad of mental retardation, seizures, and adenoma sebaceum, with angiomyolipomas seen in 80% of the cases.¹ Angiomyolipoma in tuberous sclerosis tends to occur in younger ages (mean of approximately 17 years); are larger, multiple, and bilateral; and often require surgical management as they are more likely to bleed. Both progesterone and estrogen receptors have been discovered in angiomyolipoma found in

females and in patients of tuberous sclerosis first by L'Hostis and colleagues.⁴ This is consistent with the female predominance in the sporadic form of angiomyolipoma and aggressive nature of the lesion in tuberous sclerosis. This also perhaps explains the rapid growth of angiomyolipomas and higher propensity to hemorrhage during pregnancy.

The abnormal blood vessels within angiomyolipomas may appear thick, but they lack normal elastin, resulting in formation of aneurysm and have been linked with increased incidence of hemorrhage.⁵ Calcification and necrosis is rare. Most of the angiomyolipomas are smaller than 4 cm, usually asymptomatic, and very unlikely to bleed.^{6,7} However, 82% to 94% of angiomyolipomas equal to or greater than 4 cm in diameter are symptomatic, and 50% to 60% of these result in spontaneous hemorrhage.⁸ The usual symptoms are associated with the effects of the mass or hemorrhage in the form of abdominal or flank pain, palpable mass, nausea, vomiting, hematuria, anemia, hypertension, urinary tract infection, shock, or even kidney failure.

Often angiomyolipomas can be diagnosed by various imaging modalities. On ultrasonography, they appear as well-defined and echogenic mass, irrespective of the fat content,⁹ often with posterior acoustic shadow; however, they cannot be confidently differentiated from renal cell carcinoma.¹⁰ Nonetheless, the characteristic detection of fat within the lesion by CT is diagnostic for angiomyolipoma. Thus, CT is a noninvasive technique with high diagnostic specificity that avoids unnecessary surgical intervention. Magnetic resonance imaging can also demonstrate the intratumoral fat, especially by fat suppression technique. Angiographic findings include cluster of saccular microaneurysms and macroaneurysms, hypervascularity, tortuous vessels, and the absence of normal tapering of the vessels.¹¹ Rarely, the absence of fat, involvement of surrounding structures, and invasion of the inferior vena cava and the right atrium may be seen in angiomyolipoma, making it difficult to be differentiated from renal cell carcinoma. Other rare fat-containing renal lesions such as lipoma, myolipoma, liposarcoma, oncocytoma, and Wilms tumor may sometimes mimic angiomyolipoma.

Management of the lesions depend upon the size of the tumor, associated risk of hemorrhage,

and whether the lesion is sporadic or associated with tuberous sclerosis. Lesions smaller than 4 cm can be managed conservatively with biannual follow-up examinations with ultrasonography, CT, or magnetic resonance imaging. Elective resection is indicated if the risk of hemorrhage appears to be increasing during the follow-up period. However, larger lesions need to be followed more frequently or require intervention in the form of intra-arterial embolization, nephron-sparing tumor resection, or depending upon their size and location, total nephrectomy.^{6,8,12} Treatment with selective arterial embolization of the lesion is especially indicated in patients with tuberous sclerosis, where there may be inadequate number of functioning nephrons owing to substitution of the renal parenchyma by multiple cysts and angiomyolipomas.^{12,13} More aggressive follow-up and treatment are also necessary during pregnancy, due to hormonal influence on tumor growth and increased risk of hemorrhage.⁵

CONFLICT OF INTEREST

None declared.

REFERENCES

- Hajdu SI, Foote FW, Jr. Angiomyolipoma of the kidney: report of 27 cases and review of the literature. *J Urol*. 1969;102:396-401.
- Elhence V, Mehta B, Jain M, Gupta RK. Renal angiomyolipoma: a clinico-pathological study or eleven cases. *Indian J Cancer*. 2002;39:55-60.
- Morgan GS, Straumfjord JV, Hall EJ. Angiomyolipoma of the kidney. *J Urol*. 1951;65:525-7.
- L'Hostis H, Deminiere C, Ferriere JM, Coindre JM. Renal angiomyolipoma: a clinicopathologic, immunohistochemical, and follow-up study of 46 cases. *Am J Surg Pathol*. 1999;23:1011-20.
- Logue LG, Acker RE, Sienko AE. Best cases from the AFIP: angiomyolipomas in tuberous sclerosis. *Radiographics*. 2003;23:241-6.
- Vallejo BJ, Herrera TE, Domenech CA, Lafuente PM, de Ramirez TI, Robles MJ. [Renal angiomyolipoma: presentation, treatment and results of 20 cases]. *Actas Urol Esp*. 2008;32:307-15. Spanish.
- Limaiem F, Mekni A, Chelly I, et al. Renal epithelioid angiomyolipoma: a case report and literature review. *Pathologica*. 2008;100:31-5.
- Takebayashi S, Horikawa A, Arai M, Iso S, Noguchi K. Transarterial ethanol ablation for sporadic and non-hemorrhaging angiomyolipoma in the kidney. *Eur J Radiol*. 2008.
- Heckmann M, Heinrich M, Humke U, Bautz W, Uder M. [Differential diagnosis of focal lesions of the kidney in CT and MRT]. *Rontgenpraxis*. 2008;56:219-40. German.

10. Prasad SR, Surabhi VR, Menias CO, Raut AA, Chintapalli KN. Benign renal neoplasms in adults: cross-sectional imaging findings. *AJR Am J Roentgenol.* 2008;190:158-64.
11. Winterkorn EB, Daouk GH, Anupindi S, Thiele EA. Tuberous sclerosis complex and renal angiomyolipoma: case report and review of the literature. *Pediatr Nephrol.* 2006;21:1189-93.
12. Park HK, Zhang S, Wong MK, Kim HL. Clinical presentation of epithelioid angiomyolipoma. *Int J Urol.* 2007;14:21-5.
13. Iarrobino G, Capasso L, Buonincontro S, et al. [Preservative surgery of the kidney for haemorrhagic angiomyolipoma: a report of two cases]. *Chir Ital.* 2008;60:587-93. Italian.

Correspondence to:
Rizwan Ahmad Khan, MD
4/817-F, Sir Syed Nagar, Mallah Nagla Road, Aligarh- 202002,
India
Tel: +91 94 1021 0281
E-mail: drrizwanahmadkhan@yahoo.co.in

Received March 2009
Revised May 2009
Accepted June 2009