

Large Aneurysm in Renal Angiomyolipoma

Ateeque Ahmed Khan, Samiya Saquib, Rabia Hafeez and Sarosh Kadri

ABSTRACT

Large aneurysms seen in renal angiomyolipoma are usually more common in patients with tuberous sclerosis in comparison with sporadic cases and they are more prone to hemorrhage ranging from renal hematoma to life-threatening perirenal hemorrhages. We present a case of 13-year female patient who was referred to Civil Hospital, Karachi, with bilateral flank pain, cutaneous nodule and low intelligence. Her color doppler ultrasound revealed pseudoaneurysm in left kidney on the background of bilateral renal angiomyolipomas. Contrast enhanced CT scan confirmed various radiological manifestations of tuberous sclerosis including subependymal tubers, bilateral angiomyolipomas, and pseudoaneurysm associated with perinephric hemorrhage in left kidney.

Key Words: *Angiomyolipoma. Pseudoaneurysm. Perinephric hemorrhage. Colour doppler ultrasound. CT scan.*

INTRODUCTION

Angiomyolipoma is the most common mesenchymal renal neoplasm and has been classified under the group of perivascular epithelioid cell tumors.¹ Angiomyolipomas are seen sporadically in approximately 80% of patients, the rest being seen as part of tuberous sclerosis.²

In about half cases of angiomyolipomas measuring more than 4 cm, there are associated vascular complications with risk of clinically significant hemorrhage.³

However, pseudoaneurysms are rare vascular complication found within angiomyolipomas associated with hemorrhage. We report a rare case of pseudoaneurysm formed within angiomyolipoma associated with perirenal hematoma in a patient of tuberous sclerosis.

CASE REPORT

A 13-year female presented to the Radiology Department, Civil Hospital, Karachi, with bilateral flank pain and cutaneous nodules on the face for the last 2 years. There was no history of hematuria. Patient had history of seizures at the age of 1 year and had low intelligence. On physical examination, multiple cutaneous nodules and shagreen patches were noted over face (Figure 1A). Rest of the examinations were normal. Her routine hematological and biochemical investigations were within normal limits.

She was referred to Radiology Department for ultrasound abdomen which showed bilateral multiple echogenic lesions in kidneys, located in both cortex and medulla,

and similar lesions in both lobes of liver (Figure 1B). There was a well define rounded cystic lesion seen in upper pole of left renal cortex measuring about 1.7 cm. Color flow doppler sonography showed flow within the cystic lesion and communication with the renal parenchymal vessels (Figures 1C and 1D).

Subsequently, her CT scan of brain and abdomen were performed for further evaluation, using 16 Slice Toshiba Activion Scanner.

Contrast enhanced CT scan of abdomen done in both arterial and venous phases, which showed bilateral multiple fat attenuated (-16 HU) lesions in kidneys and liver consistent with angiomyolipomas. There was well defined pseudoaneurysm, which showed contrast enhancement noted at cortex of upper pole of left kidney. It was surrounded by a large perirenal hematoma measuring approximately 4.0x6.5 cm (Figures 2A and 2B). CT scan of the brain revealed bilateral multiple foci of periventricular subependymal calcification consisted

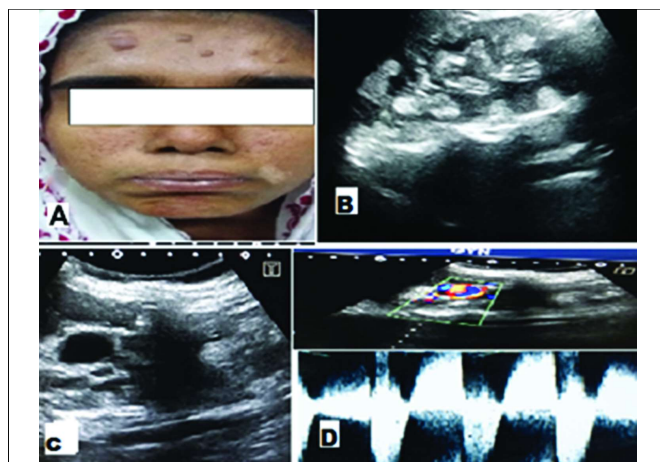


Figure 1: (A) Multiple small nodules on facial skin of patient confirmed to be facial angiofibromas. (B) B-mode ultrasound shows right renal angiomyolipomas. (C) Grey scale ultrasound image shows echogenic mass in upper pole of left kidney with central anechoic cystic lesion. (D) Color Doppler image shows central dual color flow within cystic lesion and spectral analysis shows bidirectional type of spectrum.

Department of Radiology, Civil Hospital and Dow University of Health Sciences, Karachi.

Correspondence: Dr. Rabia Hafeez, Resident, Department of Radiology, Civil Hospital and Dow University of Health Sciences, Karachi.

E-mail: doc_rabi24@yahoo.com

Received: December 12, 2017; Accepted: March 07, 2018.

with subependymal hamartomas (Figure 2C). Bone window showed multiple sclerotic foci in axial skeleton (Figure 2D). She, subsequently, underwent surgery after embolisation.

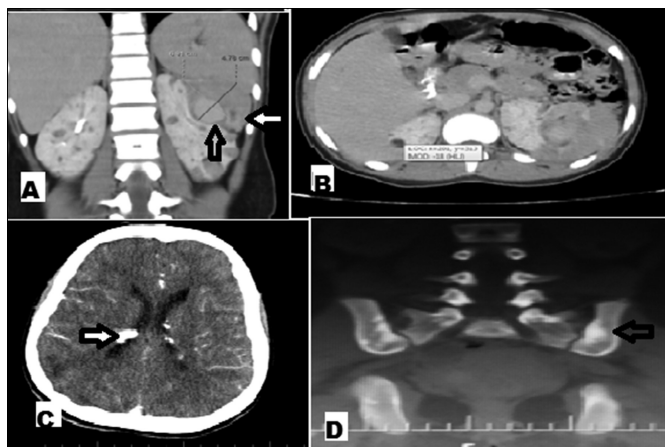


Figure 2: Coronal (A) and axial (B) CT scan nephrogram phase images show hyperdense pseudoaneurysm (up arrow) in upper pole of left kidney surrounded by heterogenous perinephric hematoma indenting its upper border (white arrow). (C) Axial CT brain shows multiple subependymal calcification in ependymal region (arrow). (D) Coronal CT bone window shows multiple sclerotic foci in axial skeleton (arrow).

DISCUSSION

Tuberous sclerosis is an autosomal dominant neurocutaneous syndrome and the diagnosis is confirmed by the presence of multi-organ involvement. Most common various radiological manifestations include neurological, renal, retinal and cardiac, especially when associated with skin lesions.⁴

Renal angiomyolipoma is a benign vascular lesion originating from perivascular epithelial cells and composed of adipose tissue, angiogenic and smooth muscle tissues in varying proportion. The blood vessels in angiomyolipoma are different from normal vessels being lack of elasticity and are thick walled. Due to their characteristic feature, they are more prone to form pseudoaneurysm and subsequently rupture to cause hemorrhages.⁵

Renal angiomyolipoma is seen in about 0.3-3% of cases.⁶ In tuberous sclerosis, angiomyolipomas are bilateral and multiple in about half of cases. They have a higher growth rate and commonly associated with hemorrhage.^{7,8}

In renal angiomyolipoma, patients usually presents clinically with variable presentation depending upon measurement of lesion, its blood supply and formation of pseudoaneurysm. Risk of formation of aneurysm increases as tumour size enlarges.⁶ In previous reported cases of aneurysm, the sizes have been usually not more than 2 cm. Yamakado done a series of 23 patients,⁹ the maximum size reported was 1.33 cm. In our case, the size of aneurysm was 1.7 cm.

Spontaneous hemorrhage from angiomyolipoma limited within renal capsule and surrounding perinephric fat may be rarely, the first presenting complain. This has been termed Wunderlich's syndrome.¹⁰

Mode of treatment depends upon size of lesion. Lesions measuring less than 4 cm, usually treated by less invasive procedure of embolisation after selective angiography. It has better prognosis and without any need of surgical intervention. While in lesions larger than 4 cm, surgery remains the treatment of choice after angiography, especially in cases of tuberous sclerosis.¹¹⁻¹³

In conclusion, angiomyolipoma can be associated with vascular complication such as formation of arterial pseudoaneurysm with hemorrhage. Aneurysm within angiomyolipoma associated with hemorrhage are at risk to rebleed, and they are life-threatening requiring early treatment. Color-flow doppler ultrasound plays a key role and remains a firstline modality for diagnosis.

REFERENCES

1. Bharwani N, Christmas TJ, Jameson C. Epithelioid angiomyolipoma: imaging appearances. *Br J Radiol* 2009; **82**:e249-52.
2. Halpenny D, Snow A, McNeill G. The radiological diagnosis and treatment of renal angiomyolipoma: current status. *Clin Radiol* 2010; **65**:99-108.
3. Lapeyre M, Correas JM, Ortonne N. Color-flow doppler sonography of pseudoaneurysms in patients with bleeding renal angiomyolipoma. *AJR* 2002; **179**:145-7.
4. Umeoka S, Koyama T, Miki Y. Pictorial review of tuberous sclerosis in various organs. *Radiographics* 2008; **28**:e32.
5. Magabe PC, Otele W, Mugambi LM. Large aneurysm in renal angiomyolipoma causing life-threatening retroperitoneal hemorrhage. *Afr J Urol* 2014; **20**:193-6.
6. Zhang JQ, Fielding JR, Zou KH. Etiology of spontaneous perirenal hemorrhage: a meta-analysis. *J Urol* 2002; **167**:1593-6.
7. Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. *J Urol* 2002; **168**:1315-25.
8. Künzi T, Walther F, Marti HP, Frey FJ, Vogt B. Intrarenal arterial aneurysms with haematuria in a patient with tuberous sclerosis complex. *Nephrol Dial Transplant* 2005; **20**:2268-70.
9. Yamakado K, Tanaka N, Nakagawa T. Renal angiomyolipoma: relationships between tumor size, aneurysm formation, and rupture. *Radiology* 2002; **225**:78-82.
10. Albi G, del Campo L, Tagarro D. Wunderlich's syndrome: causes, diagnosis and radiological management. *Clin Radiol* 2002; **57**: 840-5.
11. De Luca S, Terrone C, Rossetti SR. Management of renal angiomyolipoma: a report of 53 cases. *BJU Int* 1999; **83**:215-18.
12. Rimon U, Duvdevani M, Garniek A. Large renal angiomyolipomas: digital subtraction angiographic grading and presentation with bleeding. *Clin Radiol* 2006; **61**:520-6.
13. Ramon J, Rimon U, Garniek A. Renal angiomyolipoma: long-term results following selective arterial embolization. *European Urology* 2009; **55**:1155-61.

