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Giant Epithelioid Angiomyolipoma of Kidney - A Rare Case Report

Dr. Sunanda Lakshmi GV[1], Dr. M. Vijaya Lakshmi[2], Dr. Kharidehal Durga[3], Dr. R. Madhuri[4], Dr. G. Divya Tejaswi[5]

[1,2,3,4,5]Department of Pathology, Narayana Medical College, Dr. YSR University of Health Sciences, Nellore, India.

Corresponding Author: Sunanda Lakshmi GV

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ABSTRACT

Renal angiomyolipoma is a benign mesenchymal neoplasm of kidney and composed of varied proportions of adipose tissue, smooth muscle cells- spindled or epithelioid and dysmorphic thickened or hyalinised blood vessels. Epithelioid angiomyolipoma is one of the rare variants of angiomyolipoma with high degree of complications, recurrence and metastasis. Whereas classic angiomyolipoma does not show metastasis and recurrence. Giant epithelioid angiomyolipoma means tumor size measuring greater than10 cm, very few cases have been reported in literature. Epithelioid angiomyolipoma of size greater than 20 cm is extremely rare.

Here we report a rare case of giant epithelioid renal angiomyolipoma of size greater than 20 cm in a 21-year-old female, presented with complaints of right flank pain and hematuria.

The patient underwent nephrectomy with probability of malignancy given in ultrasonography and specimen was sent for histopathological examination. Gross examination revealed enlarged kidney of size 22×18×10 cm and cut section shows grey white to yellow lesion measuring 21×17×10 cm almost occupying entire kidney. Microscopic examination revealed interlacing fascicles of spindle cells, lobules of adipocytes and thickened blood vessels. Foci of pleomorphic epithelioid cells with illdefined cell borders, pleomorphic vesicular nucleus and prominent nucleoli are noted.

Keywords: Epithelioid, angiomyolipoma, mesenchymal, giant, kidney

INTRODUCTION

Classical renal angiomyolipoma is a benign mesenchymal neoplasm of kidney and composed of varied proportions of adipose tissue, smooth muscle cells spindled or epithelioid and dysmorphic thickened or hyalinised blood vessels [1]. Angiomyolipoma comprises of 1 % of renal tumors. 80 % of AML are sporadic and remaining are familial, a part of tuberous sclerosis complex [2]. Total number of 160 epithelioid angiomyolipoma cases have been documented in literature till now. Epithelioid angiomyolipoma is a highly cellular tumor composed of sheets of epithelioid or polygonal cells with abundant cytoplasm, variable degree of nuclear atypia and minimal or no fat. In contrast to classic angiomyolipoma, epithelioid angiomyolipoma has high malignant potential and shows high degree of recurrence, metastasis and complications

CASE REPORT

Case history: A 21 years female presented to the Urology Department with complaints of dragging sensation in the abdomen, right side flank pain and hematuria since 5 months. Patient does not have any other skin or neurological manifestations and thus tuberous sclerosis has been ruled out.

Ultrasonography of abdomen and pelvis shows an ill-defined hyperechoic lesion of size about 18×10×10 cm in lower half of right kidney with possible differential diagnosis of Angiomyolipoma, pyelonephritis and Renal cell carcinoma.

CECT of whole abdomen shows ill-defined large hyperechoic lesion measuring 18 ×15 ×8cm in the lower pole of right kidney with internal multiple focal fatty lesions and engorged dilated vascular sinuses [Figure-1]. On contrast, significant heterogenous enhancement was seen. Impression was given as Giant angiomyolipoma of right kidney. Nephrectomy was done and the specimen was sent in 10 % neutral buffered formalin for histopathological examination.

Gross examination: Received nephrectomy specimen measuring 22 ×18 × 10 cm. Cut section shows grey white to grey yellow lesion of size 21×17×10 cm almost occupying the entire kidney without any corticomedullary differentiation [Figure-2].

Microscopy: Microscopic examination shows compressed renal parenchyma with adjacent foci showing lesion composed of interlacing bundles and fascicles of spindle cells, lobules of adipocytes, thickened and hyalinised blood vessels. At many foci sheets of pleomorphic epithelioid cells with poorly defined cell outlines, moderate eosinophilic cytoplasm, vesicular nucleus and prominent nucleoli are also noted. Mitotic figures of 1 / 10 HPF are noted. Atypical mitotic figures and areas of necrosis are not identified [Figure 3 and 4]. The diagnosis was given as Giant Epithelioid variant of angiomyolipoma of nonaggressive type.

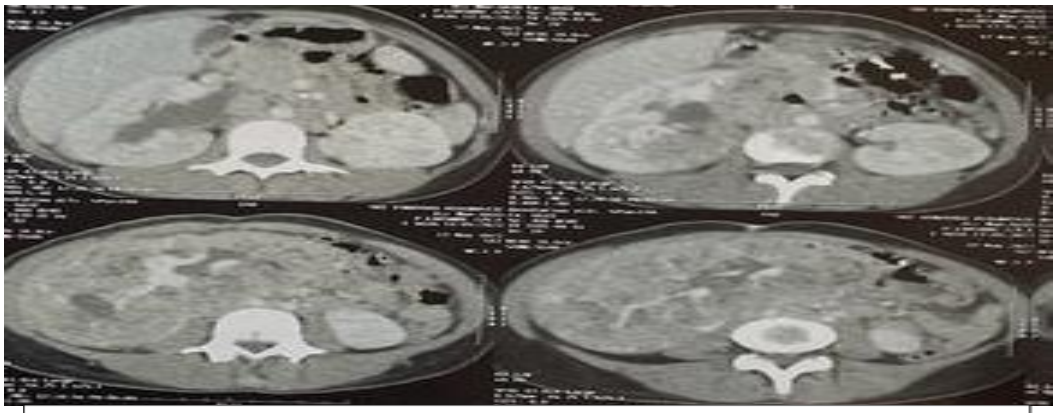

Figure 1: CECT shows ill-defined hyperechoic lesion of size 18 ×15×8 cm in lower pole of right kidney.

Figure 2: Gross examination showing Grey white to yellow mass 21×17×10 cm almost occupying entire kidney.

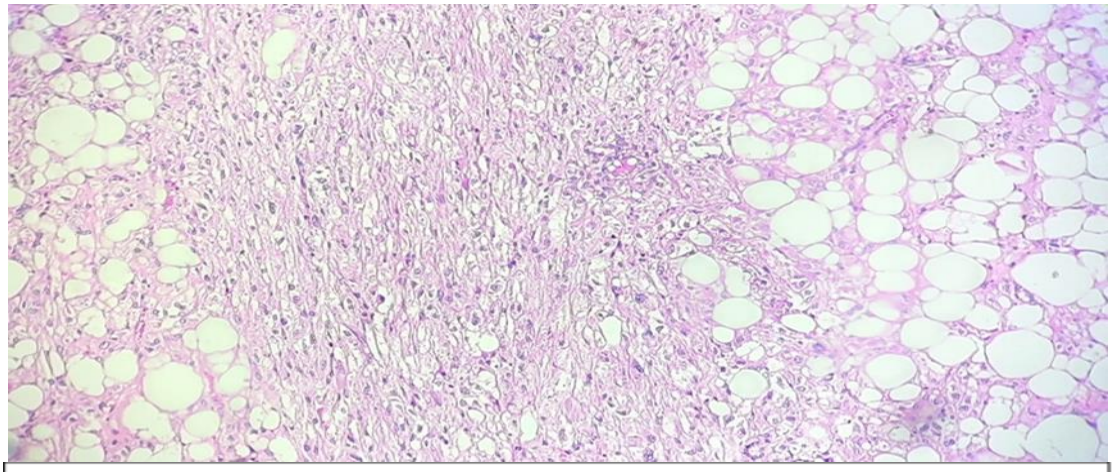

Figure 3: Microscopic examination shows adipocytes , smooth muscle cells and few blood vessels. Foci of epithelioid cells with mild nuclear pleomorphism also noted. [H&E- X 100]

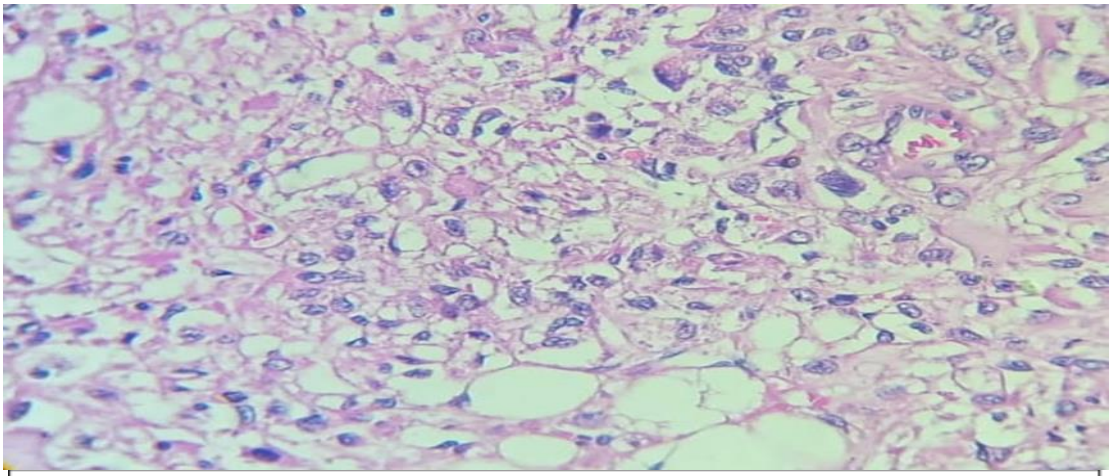

Figure 4 : Microscopic examination shows pleomorphic epithelioid cells with small foci of mature adipocytes. [H& E-x 400]

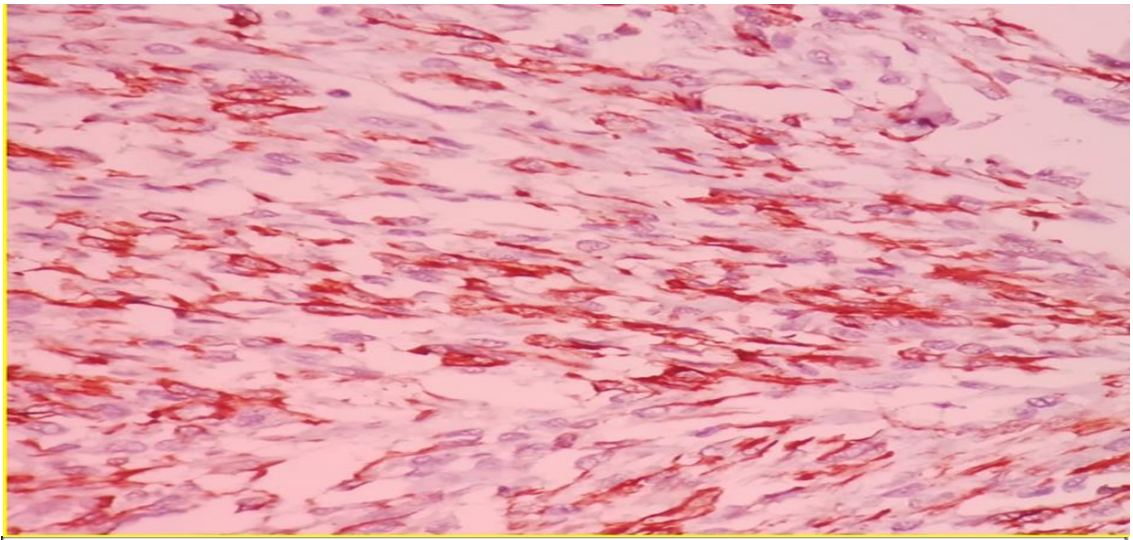

Figure 5 : Immunohistochemical picture showing HMB 45 positive tumor cells[IHC-x 400]- Confirmatory diagnosis of angiomyolipoma.

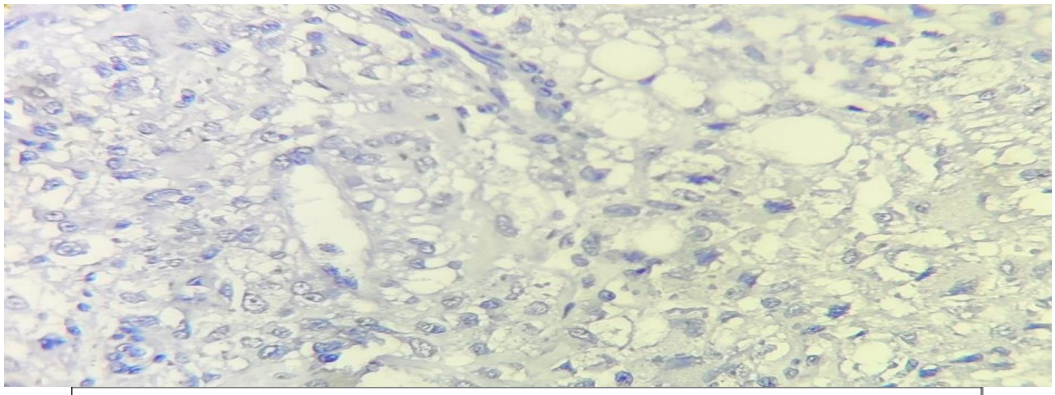

Figure 6 : Immunohistochemical picture showing CK 7 negative tumor cells[IHC-x 400]

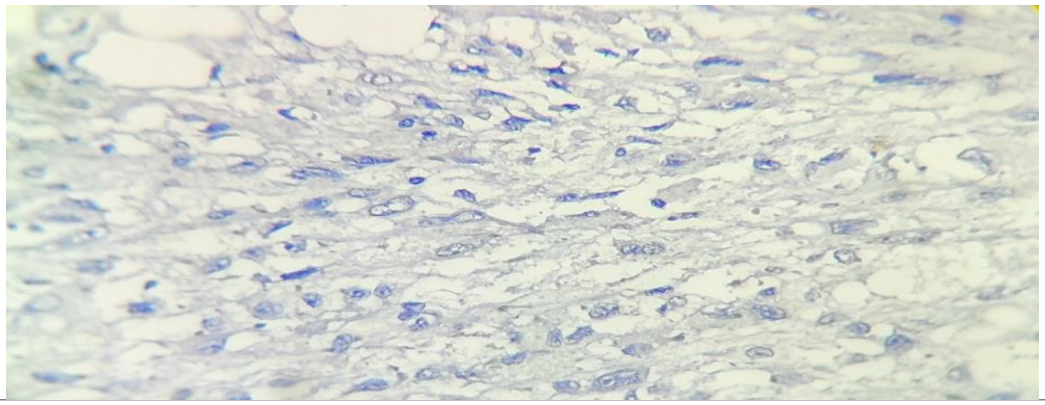

Figure 7 : Immunohistochemical picture showing Ki 67 staining of tumor cells[IHC-x 400] – Low index.

DISCUSSION

Classical renal angiomyolipoma is a benign mesenchymal neoplasm of kidney. Females are more frequently affected than males and M: F 1:4 [3,4,5]. Many of the cases are asymptomatic and symptoms appear as the size of the tumor increases. Hematuria, palpable mass and flank pain are more common clinical manifestations [3,4,5]. In the present case patient presented with flank pain, hematuria and dragging sensation in abdomen.

Epithelioid angiomyolipoma is a rare subvariant of angiomyolipoma and was first described by Pea et al in 1998 [6]. Epithelioid angiomyolipoma accounts for 4.6% of all AML [7]. Giant angiomyolipoma is of size greater than 10 cm and very few cases have been reported in literature till now [8,9]. De Bree et al also reported a case of giant Epithelioid angiomyolipoma though most commonly seen in kidney, it may involve liver, lung, pancreas, bladder, prostate, uterus, ovary, vulva, vagina and bone [9] 2004 WHO classification states that in contrast to classic angiomyolipoma, epithelioid angiomyolipoma has malignant potential and shows high degree of recurrence, local invasion and distant metastasis even after resection [10]. In the literature aggressive angiomyolipoma is characterised based on four histological features greater than 70% of atypical epithelioid cells, > 2 mitosis / 10 HPF, atypical mitotic figures and necrosis [11,12]. In our case there is 30 % of atypical epithelioid cells and 1/10HPF mitosis without atypical mitotic figures and no areas of necrosis, so the final diagnosis was Giant epithelioid angiomyolipoma of non-aggressive type. The closest differential diagnosis of epithelioid angiomyolipoma is renal cell carcinoma where histopathology and immunohistochemistry are helpful in differentiating both lesions. HMB 45 is positive in epithelioid angiomyolipoma and epithelial markers like CK 7 and EMA are negative [3]. In the present case HMB 45

was positive, CK 7 negative and Ki 67 low index thus renal cell carcinoma was rule out and definitive diagnosis of Giant Epithelioid variant of renal angiomyolipoma of nonaggressive type was given.

Outcome and follow-up:
On follow up the patient is doing well.

CONCLUSION

Diagnosis of Epithelioid angiomyolipoma is very important and this entity require regular follow-up for detecting recurrence and metastasis whereas in classic angiomyolipoma nephrectomy is sufficient. Histopathology and immunohistochemistry help in confirmatory diagnosis of epithelioid angiomyolipoma and helps in differentiating it from renal cell carcinoma.

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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