Case Report

Renal angiomyolipoma

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ABSTRACT

Renal angiomyolipoma (AML) is an uncommon benign tumor of kidney. It is composed of abnormal thick walled blood vessels, smooth muscles, and adipose elements. It has an incidence of 0.3–3% among kidney tumors. These tumors are isolated in 80% of cases and are associated with tuberous sclerosis in 20% of the cases. Here, we report a case of renal AML in a 57-year-old female with review of literature.

Key words: Angiomyolipoma, benign tumor, kidney

INTRODUCTION

Renal angiomyolipoma (AML) is an uncommon benign mesenchymal tumor of the kidney. It contains fat, smooth muscles, and tortuous blood vessels in varying proportions. It was first described by Bournville in 1880.[1] It is often considered to be hamartoma or choristoma. Morgan and Associates first used the term AML.[2] It has a incidence of 0.3–3% among kidney tumors.[3] AMLs are more commonly associated with the genetic disease tuberous sclerosis, in which most individuals will have several AMLs affecting both kidneys. It is associated with these diseases or sporadic, AMLs are caused by mutations in either the TSC1 or TSC2 genes, which govern cell growth and proliferation. It is derived from the perivascular epitheloid cells which are HMB-45 positive.[5]

CASE REPORT

A 57-year-old woman presented with abdominal pain and mass in the right side of the abdomen since 4 months duration. On clinical examination, there was a firm, irregular nontender mass palpable in the right lumber region. Ultrasonography revealed a large hypoechoic mass originating from midpole of right kidney's scan demonstrated a lobulated mass with hypodense lesion arising from midpole of right kidney. Right sided nephrectomy was done and specimen was sent for histopathological examination.

Gross

Received a specimen of kidney covered all around by perirenal fat of size 10 cm × 9 cm × 4 cm. On one side ureter of 4 cm length was seen. Cut surface shows a part of the kidney with central area occupied by tumor mass having brownish colored [Figure 1].

Microscopic examination confirmed the diagnosis of renal AML with characteristic picture of tumor composed of mature fat cells, irregular thick and thin walled blood vessels, and smooth muscle proliferations [Figure 2].

DISCUSSION

Renal AML is a benign mesenchymal tumor of kidney. Two forms of this tumor are recognized clinically. First associated with tuberous sclerosis[2] where the tumor is usually small, multiple, bilateral, and has an equal sex distribution.

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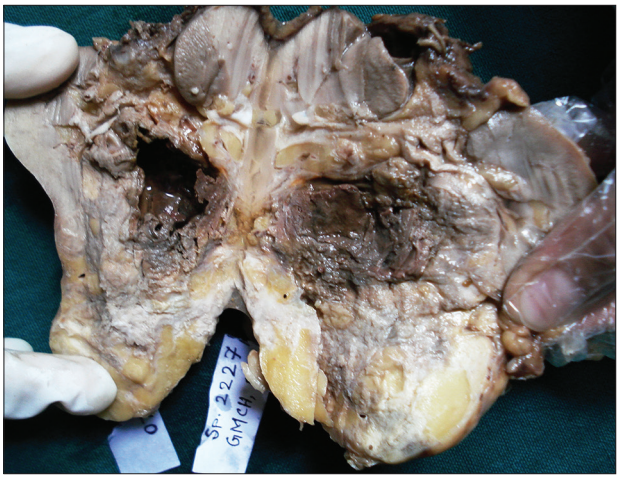

Figure 1: A specimen of kidney covered all around by perirenal fat of size 10 cm × 9 cm × 4 cm

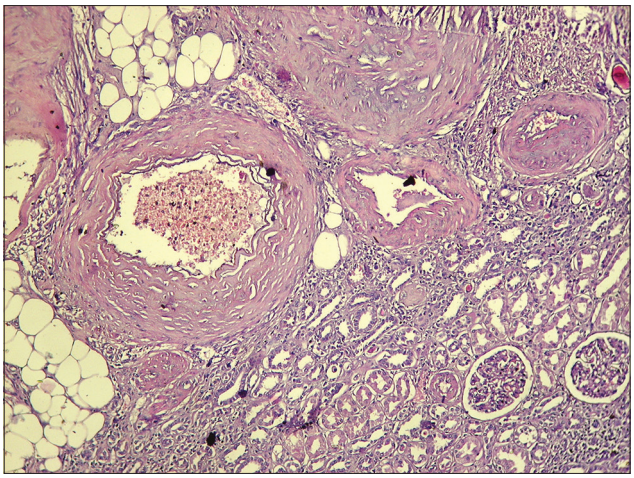

Figure 2: Tumor composed of mature fat cells, irregular thick and thin walled blood vessels, and smooth muscle proliferations

The second form that occurs sporadically accounts for 80% of the tumor and is often seen in woman and is not associated with tuberous sclerosis. Here, the tumor is often large, single, and unilateral.[4]

Mean age of presentation is 43-year-old and 4 times more common in woman than in men with involvement of right kidney in 80% of the cases.[5] The patients present with abdominal mass, hematuria, and flank pain. The solitary sporadic tumors may cause acute abdomen and shock as a result of spontaneous hemorrhage in the tumor. [6] The most common site involved is the kidney, but it is also seen in extra renal sites such as liver, lungs, lymph nodes, and retroperitoneal tissues.

Angiomyolipoma is benign neoplasm with size varying from few millimeters to 20 cm. On gross, these tumors are lobulated. Cut surface yellowish to gray. Histologically three components are seen. Mature adipose tissue, tortuous thick walled blood vessels, and smooth muscles. It shows positivity for MHB-45, Gp100, Melan-1.

Major complications are retroperitoneal hemorrhage. Rare cases with sarcomatous transformation with subsequent distant metastasis have been reported.[7]

CONCLUSION

The current management options include observation, embolization, and partial or total nephrectomy. Tumors <4 cm tend to be asymptomatic hence require regular follow-up with no intervention. Tumors more than 4 cm may undergo rupture so they should be resected to avoid complications like hemorrhage.

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Conflict of interest

There are no conflict of interest.

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