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Pararenal Retroperitoneal Angiomyolipoma: A Rare Case Report

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Abstracts: ***Angiomyolipomas are benign mesenchymal tumors that usually occur in kidneys. In contrast, pararenal angiomyolipomas are very uncommon tumors. We report a rare case of pararenal retroperitoneal angiomyolipoma with no evidence of tuberous sclerosis. A 33 years old woman presented with left sided flank pain. Abdominal computerized tomography (CT) revealed a large angiomyolipoma of 9.3x6.1x7.7centimeters sized arising from the lower pole of left kidney. During surgical procedure mass was found to be at pararenal retroperitoneal location. Histopathological examination and immunohistochemistry revealed the mass to be pararenal angiomyolipoma.***

Keywords: Angiomyolipoma, Pararenal, Retroperitoneal, Tuberous sclerosis.

1. Introduction

Pararenal retroperitoneal angiomyolipoma is an uncommon disease entity and present as incidentalomas upon imaging for other condition. Surgical resection is always advocated to differentiate suspected pararenal retroperitoneal angiomyolipomas from other retroperitoneal lesions like liposarcomas, lipoma, leiomyosarcomas and leiomyomas. These tumors have prominent vascular supply and may present symptomatically with abdominal pain and hemorrhagic shock. Their management generally lead to nephrectomy, so early surgical intervention is needed prevent the complication and to preserve the kidney.

2. Case Report

A 33 years old female presented with left sided abdominal pain since four months. On examination abdomen was soft and tender. Other systemic examinations and vital parameters were within normal limits. Patient had no history of hypertension or diabetes mellitus. There was no significant past history. The patient was subjected to routine laboratory and radiological investigations. All the hematological and biochemical investigations were found to be within normal limits. The computed tomography of abdomen showed a large mass showing fat attenuation arising from lower pole of the left kidney (Figure 1). Provisional diagnosis of renal angiomyolipoma was made. Patient was re-evaluated for evidence of tuberous sclerosis, but there were no signs and symptoms favoring it. Patient was planned for partial nephrectomy of left kidney. During surgical procedure mass was found to be pararenal retroperitoneal location without involving the left kidney. Surgical removal of mass was done without resecting the left kidney. Excised mass was sent for histopathological examination. The following observations were made on gross and microscopic examinations.

Gross Examination

Gross specimen included one soft tissue piece, capsulated and measuring 7.5x5.5x5.0 centimeters. External surface was smooth and yellowish and cut surface was yellow and fleshy.

Histopathological Examination

Histopathological examination of excised tissue revealed predominantly adipose tissue with multiple thick walled vascular channels lined by flattened endothelial cells. Perivascular epithelioid cells were proliferating and emanating from blood vessel wall and extending into the surrounding adipose tissue. There was no renal tissue seen in the excised tissue. The histopathological diagnosis of pararenal angiomyolipoma was made (Figure2).

The biopsy material was further subjected to immunohistochemistry where it demonstrated strong positivity for HMB-45 and smooth muscle actin. These findings were consistent with histopathological diagnosis of pararenal angiomyolipoma.

3. Discussion

Angiomyolipomas are rare benign renal neoplasm composed of smooth muscles, thick walled blood vessels and mature adipose tissue. Angiomyolipomas more commonly occur in female and they account for one percent of renal lesions.[1] Angiomyolipomas usually arise in kidney but rarely they can occur at extrarenal sites. Liver is the most common extrarenal site. Other extrarenal sites are the uterus, retro-peritoneum, head, vagina, penis, abdominal wall, spermatic cord and colon. Retroperitoneal angiomyolipomas are exceedingly rare. Like renal angiomyolipomas extrarenal angiomyolipomas are more prone to bleed because these tumors have prominent vasculature and thick wall blood vessels which are deficient of elastic fibrillae. Generally these patients present with abdominal pain, bleeding, shock or symptoms due to tumor compression to adjacent organ. On CT scan of abdomen angiomyolipomas show fat attenuation

which makes the diagnosis rather easy. On CT angiography, presence of aneurysm like vascular lesions outside the kidney with normal arterial branching inside the kidney suggest the possibility of extrarenal angiomyolipoma or renal angiomyolipoma with exophytic growth.[2]

The perivascular epithelioid cells of angiomyolipoma show immunoreactivity for epithelial membrane antigen, keratin, vimentin, desmin, smooth muscle actin and HMB-45. HMB-45 a monoclonal antibody against a melanoma-associated antigen used to differentiate angiomyolipomas from other retroperitoneal tumor like liposarcomas, lipoma, leiomyosarcomas and leiomyomas as angiomyolipoma show strong positive immunoreactivity for it. Renal angiomyolipoma has strong association with tuberous sclerosis as 30-40% patients of renal angiomyolipoma have features of tuberous sclerosis and 80% of patients with tuberous sclerosis will develop renal angiomyolipoma. So brain CT scan is recommended for patients with renal angiomyolipomas to rule out the possibility of tuberous sclerosis. The brain CT scan of these patients typically demonstrates characteristic periventricular subependymal nodules with calcifications.[3] In contrast extrarenal angiomyolipoma has no association with tuberous sclerosis.[4]

Surgical removal is the treatment of choice for large and symptomatic extrarenal angiomyolipomas. Most of the patients of retroperitoneal extrarenal angiomyolipomas present with hemorrhage and shock and their management generally lead to nephrectomy, so early surgical intervention is needed to prevent massive bleeding and, most importantly, to preserve the kidney. Selective arterial embolization has been used effectively to control bleeding in hemodynamically unstable patients. This results in tumor regression and subsequently allowing for elective removal of tumor .[3]

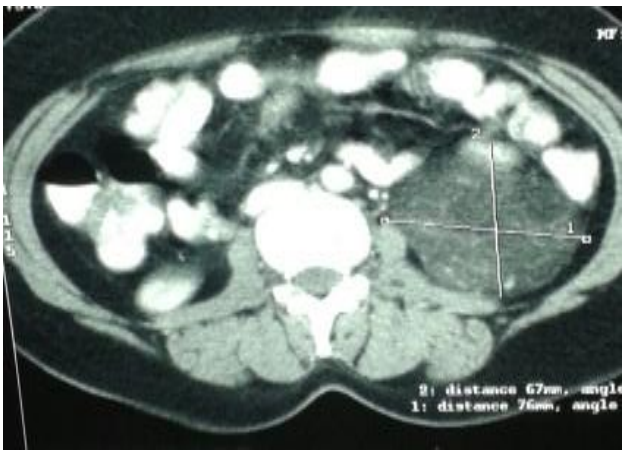

Figure 1

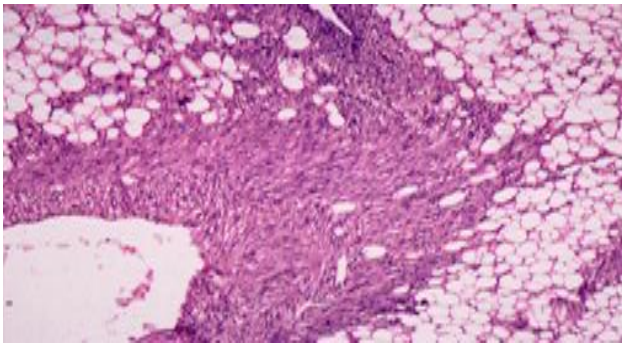

Figure 2

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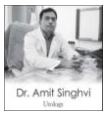

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