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Case Report

A case of Giant Renal Angiomyolipoma in Tuberous Sclerosis Complex

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ABSTRACT

Angiomyolipomas also referred to as renal hamartoma is a benign lesion. They are either sporadic or associated with syndromes like tuberous sclerosis. Here we present a case of 58years old male patient, who presented with left sided abdominal pain for 1 month. On examination over face Adenoma Sebaceum was present. Ultrasound abdomen and contrast enhanced computed tomography showed bilateral renal angiomyolipoma with aneurysm over left side. On plain computed tomography of head subependymal calcifications were present. Left Nephrectomy was done and histopathology revealed Angiomyolipoma.

Keywords: Adenoma sebaceum, Giant renal angiomyolipoma, Nephrectomy, Tuberous sclerosis

INTRODUCTION

Renal AML is a benign neoplasm arising from mesenchymal elements and was first reported in 1951.[1] AML (also referred as hamartoma) is a benign lesion consisting of varying amounts of mature adipose tissue, smooth muscle, and blood vessels.[2] They are considered to be originating from neural crest cells perivascular epithelioid cells. The inheritance pattern of renal AML is autosomal dominant. They are either sporadic or associated with syndromes like Tuberous sclerosis 20%, Sporadic Lymphangioleiomyomatosis.

It was previously reported that renal AML may grow by 4 cm each year in its maximum dimension.[3,4] When the diameter of renal AML reaches >10 cm, the tumor is referred to as 'giant'. Reports of giant renal AML in the literature are uncommon, with the largest renal AML (39 × 25 × 9 cm) reported in 2013 by Taneja et al.[5,6] The great majority of angiomyolipoma's arises in the kidney. Extrarenal angiomyolipoma's are extremely rare and reported in the liver, nasal cavity, vagina, spermatic cord, skin, mediastinum and GI tract.

CASE REPORT

In November 2017, a 58 years old male patient was admitted in Department of Urology in ASRAM (Eluru) with complaints of pain abdomen over left side for 1 month and one episode of bilious vomiting 10 days back. No significant family history.

On examination there were multiple hyperpigmented papules which were bilaterally symmetrical present over cheeks and nasal bridge suggestive of Adenoma Sebaceum (Figure 1). On Per abdomen examination there was a vague intra-abdominal mass approximately measuring 12 × 10 cms in left lumbar region. Mass was bimanually palpable and moving with respiration. On percussion dull note present. Routine blood investigations were within normal limits. On ultrasound abdomen there is ill-defined hyperechoic lesion measuring 12.7 × 7 cm

with internal vascularity arising from left kidney and 2.4 × 2 cms from right kidney.

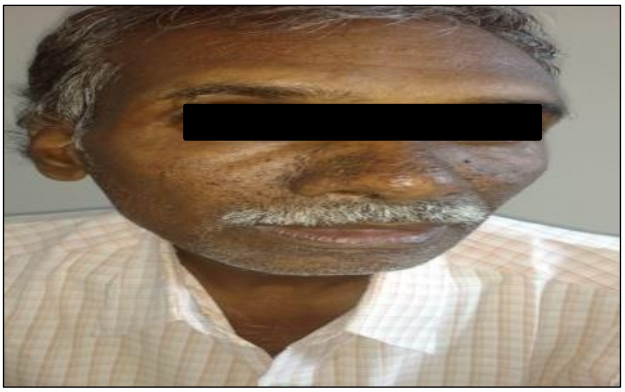

Figure 1: Adenoma sebaceum over cheeks.

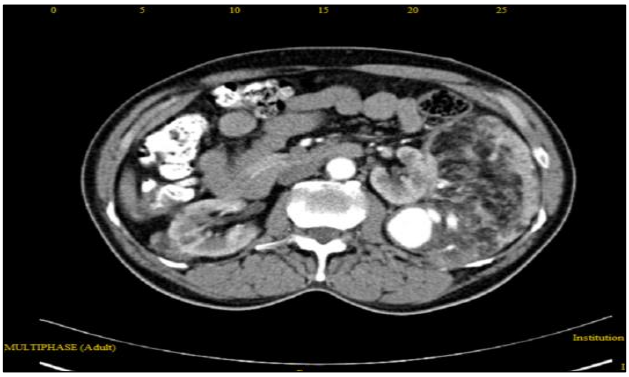

Figure 2: CECT abdomen.

On CECT Abdomen (Figure 2) there were multiple well defined heterogenous lesions with fatty, soft tissue components and feeding vessels in both kidneys left measuring 12.2 × 7.9 cm×11.8cms and right measuring 1.7×1.7×1.2cms with aneurysm in left kidney largest measuring 4×2.9 cm.I.e; bilateral renal angiomyolipoma. Plain CT Head Figure 3.

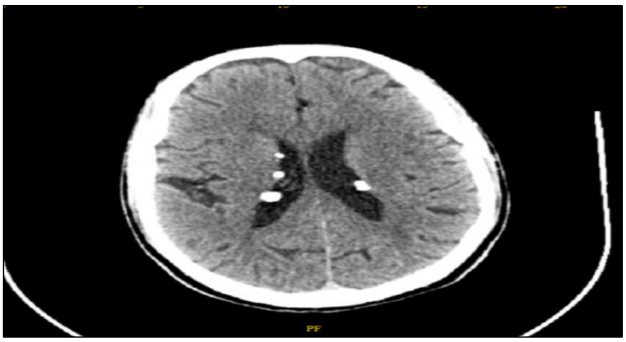

Figure 3: On CT Head calcified subependymal nodules were present.

The diagnosis of Left giant renal angiomyolipoma and right renal angiomyolipoma in tuberous sclerosis complex was made. Left Nephrectomy was done. Specimen sent for histopatholgy revealed Angiomyolipoma. The patient was kept on tab. Everolimus 10 mg once daily (AML in Right kidney).

DISCUSSION

80-90% of Renal Angiomyolipoma are sporadic. They are usually unilateral, smaller in size, incidentally found and aneurysms are rare.

Tuberous sclerosis (TS) or Bourneville-Pringle disease is a relatively rare autosomal dominant disorder with variable penetrance affecting approximately 1 in 10,000 population.[7,8] Classically the disease is described as a clinical trial of adenoma sebaceous, mental retardation and seizures. The pathophysiology can include glial tumours of brain, adenoma sebaceous of skin, rhabdomyoma of heart and hamartomata's tumours of thyroid, retina, liver, pancreas, lung, kidney, adrenals and ovaries.[7,8]

Three types of renal involvement have been described in TS:

- Renal AML (40-80%)
- Cystic disease (occasionally)
- Renal cell carcinoma, with the most common renal lesion being AML[9].

AML in TS are potential for hemorrhage or mass effect. They don't cause renal failure. Usually bilateral, larger in size. Arteries here lack internal elastic membrane hence forms aneurysms leading to rupture and hemorrhage (Wunderlich`s syndrome).

AML is the only benign renal tumor that is confidently diagnosed on cross-sectional imaging. The presence of fat (confirmed on non-enhanced thin-cut computed tomography by a value of 20 [HU] or less seen within a renal lesion on imaging is considered the diagnostic hallmark.In general, asymptomatic AMLs with a diameter of 4 cm or smaller can be followed up. For larger and symptomatic tumours, surgical intervention should be considered.[10] Nephron sparing approach such as angio-embolization or partial nephrectomy is preferable, especially in TS with bilateral, and multiple AML.[11]

Histopathological, AML consists of mature adipocytes, thick-walled blood vessels, and epithelioid stromal cells in various proportions. Usually it displays as a pattern of typical fat and perivascular epithelioid cells arranged around a blood vessel.[12] Positive immunoreactivity for HMB-45, a monoclonal antibody raised against a melanoma-associated antigen, is characteristic for AML and can be used to differentiate this tumor from sarcoma and other tumors.[13]

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