Case Report

Renal angiomyolipoma: A radiopathological diagnosis

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ABSTRACT

Angiomyolipoma (AML) is a benign renal neoplasm derived from perivascular epithelioid cells and composed of fat, and vascular and smooth muscle element besides spindle cells. It has an incidence of 0.3 - 3% and has increased due to advances in imaging modalities. The overall female to male ratio is approximately 4:1 which is suggesting hormonal component to tumor growth. It can occur sporadically or may be associated with tuberous sclerosis complex. We present a case of renal angiomyolipoma in a 28 year old woman who presented with symptoms of pain off and on and the diagnosis was based on the computed tomography and was confirmed on histopathology.

Key words: Angiomyolipoma, kidney tumor, renal angiomyolipoma

INTRODUCTION

Angiomyolipoma (AML) is a benign renal neoplasm derived from perivascular epitheloid cells and composed of fat, and vascular and smooth muscle element besides spindle cells.[1] It has an incidence of 0.3 - 3% and has increased due to advances in imaging modalities.[2] The overall female to male ratio is approximately 4:1 which is suggesting hormonal component to tumor growth.[3] It can occur sporadically or may be associated with tuberous sclerosis complex. Isolated AML which occurs sporadically, is often solitary and accounts for 80% of the tumors. AML associated with tuberous sclerosis accounts for 20% of the tumors, these are larger, often bilateral and multiple. Interestingly 80% of the cases involve the right kidney.[4]

CASE REPORT

A 28-year-old woman presented to the department of Surgery with chief complaints of pain in right lumbar area with radiation to right iliac fossa since 8 months. She went to a local medical practitioner who gave her analgesics and antacids and advised ultrasonography. Patient kept on taking the medications and was relieved of the pain. After a period of 6 months, she again developed pain in right lumbar area which was severe this time. She got ultrasonography done which revealed a mass of size 7.37 × 6.3 cm in relation to anterior cortex of right kidney. Echogenic areas of fat were seen in the mass. The mass also showed increased colour uptake. The left kidney was normal in size and echotexture. Bilateral pelvicalyceal systems were normal. A differential diagnosis of angiomyolipoma/ hypernephroma was kept.

Since it did not clear the picture, a Computed Tomography (CT) scan was advised which showed a well defined heterogeneously enhancing mass lesion measuring 10 × 7.5 × 7 cm with internal areas of hypodensities with negative HU values suggestive of fat arising from inferomedial aspect of right kidney causing compression of pelvicalyceal system. A final impression of angiomyolipoma was thus made [Figure 1].

The right sided partial nephrectomy was done which included the tumor with some renal tissue adherent to it. Proper hemostasis was achieved and the abdomen closed. Postoperative period was uneventful and she was discharged with stitch removal done on the 7th day. The partial nephrectomy specimen received in the department of pathology on gross examination revealed a tumor mass

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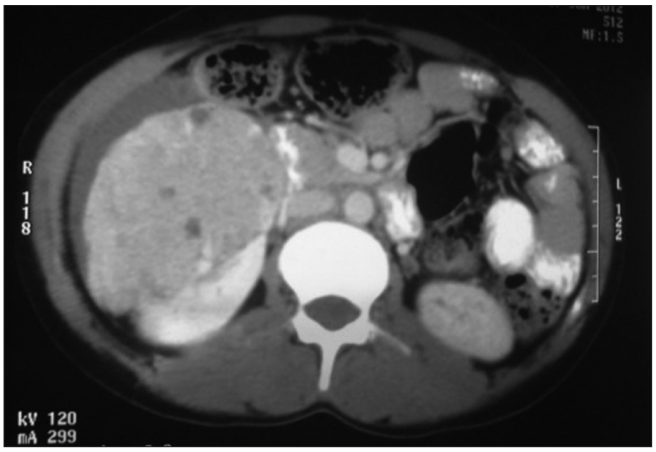

Figure 1: CT scan - A well defined heterogeneously enhancing mass lesion with areas of hypodensities

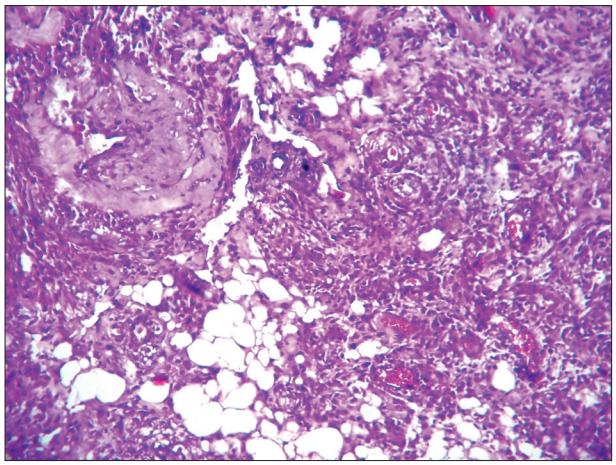

Figure 2: Haphazard arrangement of blood vessels, smooth muscles and adipose tissue with few thick walled blood vessels (H and E, 40×)

measuring 10.5 × 7.5 × 4 cm. Cut surface was variably grey-white to yellow with some areas of congestion.

Microscopic examination revealed admixture of blood vessels, smooth muscles and adipose tissue in haphazard arrangement. Smooth muscles were arranged at places in interlacing fashion. Most of the blood vessels had thick walls. No atypia or necrosis was seen in the sections examined [Figure 2].

DISCUSSION

The Renal AML has an incidence of 0.3 – 3% of all kidney tumors.[2] The tumors may remain silent for many years or are discovered on imaging studies. It may also present with flank or abdominal pain in 50% of patients or hematuria in 20% of patients.[5] Two types are described (a) Isolated AML (b) AML associated with tuberous sclerosis.[6] When associated with tuberous sclerosis 71% are bilateral and 87% are multiple. It has mean age of 41 years with female preponderance. The average size of AML is 9 cm but varies from 3 - 20 cm.[5] AML which are associated with tuberous sclerosis are bilateral, multiple and larger.[6]

The sporadic AML occurs at an average age of 45 yrs and the average size is 9 cms with female preponderance with ratio is 4:1.[7] Right kidney is affected in 80% cases. The tumor is incidental diagnosis on imaging or presented as retroperitoneal haemorrhage. Characteristic absence of elastic tissue in tumor vessels predisposes the patient to aneurysm formation and spontaneous hemorrhage which is the most dangerous complication.[8]

Angiomyolipoma (AML) may be confused with other mesenchymal tumors of the kidney. They are leiomyoma, leiomyosarcoma, liposarcoma and sarcomatoid renal cell carcinoma (RCC). This is particularly problematic in case when predominant smooth muscle component is associated with nuclear atypia and necrosis. The presence of triphasic components like adipose tissue, smooth muscle, and characteristic thick walled blood vessels differentiate it from leiomyoma. Similarly these features with paucity of mitotic figures and necrosis separates AML from leiomyosarcoma. Lipoblast and characteristic chicken wire configuration of vessels are the hallmark of liposarcoma, which were not seen in AML. Sarcomatoid RCC contains biphasic components as interlacing bundles and fascicles of spindle cells with clear cell, chromophobe or papillary carcinomatous elements. Presence of separate huge cyst on gross and microscopic separation between AML (without cystic change) and simple cyst by epithelium and compressed atrophic renal tissue in between both diffentiate it from cystic variant of AML.[9]

Basic therapeutic algorithm for renal angiomyolipoma is follow-up. Tumor size and the presence of symptoms are most decisive to decide best therapy alternative. Patients with AMLs of >4 cm should be warned of the risk of spontaneous tumour rupture with massive bleeding. Conservative renal surgery is possible for most AMLs.[10]

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