Case Report

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Renal Angiomyolipoma: A case report

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Abstract:

Angiomyolipoma is rare benign tumor which is composed of an intimate admixture of blood vessels, smooth muscle cells and fat hence the name. They occur at many sites, more commonly in kidney. They are seen in 25-50% of patients with tuberous sclerosis. The unilateral presentation is uncommon. Angiomyolipoma presenting as enlarged lump may mimic renal cell carcinoma, yet malignancy has to be ruled out. Excision is recommended for definite histological diagnosis in symptomatic patients and to prevent risk of haemorrhage and malignancy.

We describe a 69 years old female with history of vague pain, right flank for 2 years with bimanually palpable painless mass in right flank. She has no history of tuberous sclerosis, haematuria and dysuria. Computerised tomography demonstrated a hypodense lesion measuring 95×54 mm in postero- lateral aspect of right kidney with linear hyperdense area and multiple rounded hypodense area in renal parenchyma. At right radical nephrectomy adrenal gland was completely normal. Histology shows perinephric angiomyolipoma with multiple microscopic angiomyolipoma in the renal parenchyma with peculiar growth pattern completely encompassing entire kidney ,capsule well preserved.

Key words: Angiomyolipoma, Unilateral, Kidney, PEComa, Perivascular epitheliod cell tumor

Introduction:

Angiomyolipoma (AML) is an uncommon, benign, mesenchymal tumour (2-6.4% of all kidney tumours) which arises from either the renal pelvis or the sinus, which is composed of an intimate admixture of blood vessels, smooth muscle cells and fat, AML commonly occurs in women in age group of 40-70yrs [1]. It may occur sporadically as isolated lesions (80-90%) or in association with autosomal dominant disorder tuberous sclerosis (TSC) (25-50%) [2]. Perivascular epithelioid cell tumours (PEComas) includes both classic angiomyolipoma (AML) and its potentially malignant variant epithelioid angiomyolipoma (EAML). PEComas are associated with tuberous sclerosis complex [3]. EAML are often confused with renal cell carcinoma (RCC) given similar symptoms and lack of macroscopic fat on radiographic imaging [4]. Sporadic AML are usually unilateral and those associated with TSC are usually bilateral and can occur in any age and in either sex. Usually it is asymptomatic and is often found incidentally in USG or CT. Sometimes it may present with retroperitoneal haemorrhage [5].

Case summary:

A 69 years old female presented with pain right flank of her abdomen for 2 years, dull aching intermittent with no aggravating or relieving factors. There is no history of haematuria or weight loss. Unremarkable past history of epilepsy or mental retardation and no family history of malignancy.

On physical examination, there was no cutaneous or other stigma of tuberous sclerosis. A large (>10cm) mass was palpable in her right upper quadrant, margins ill defined, non tender, mobile, ballotable . Laboratory results demonstrated normal haemoglobin 12.6 mg% and creatinine 1mg%, as well as negative urine analysis. X ray chest shows raised right dome of diaphragm. Ultrasound of abdomen reports SOL in Right kidney with multiple cholelithiasis without cholecystis. Both kidneys were commented normal in size, shape and countour. CT scan of abdomen with intravenous contrast identified a 95×54 mm hypodense mass in postero-lateral area of right kidney, in continuity with defect in right kidney

showing linear hyperdense area with multiple rounded hypodense area in renal parenchyma.

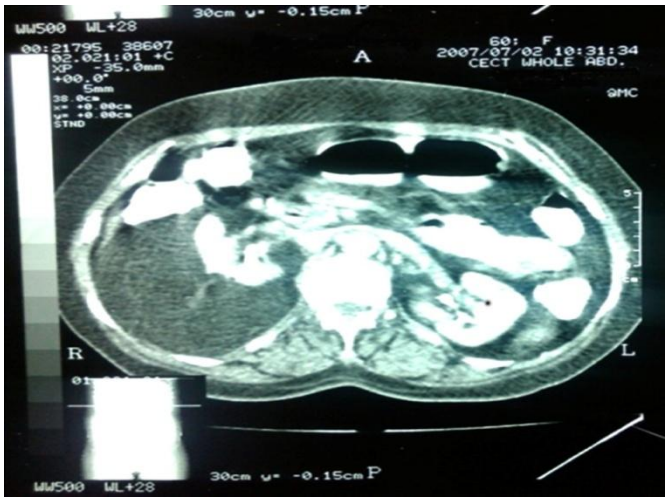

Figure 1: CT scan showing hypodense mass in right kidney.

In view of above findings and pain, malignancy has to be ruled out. She was scheduled for wide excision under GA with consent and possibility for right radical nephrectomy and adrenalectomy. Intraoperatively mass was seen grossly encasing right kidney. Right nephrectomy was done, right adrenal glands was normal and not removed. She made an uneventful recovery and was discharged on POD 8.

In Histopathological examination, macroscopically the mass was solid measured 18×10×9cm with weight of 750grams. Kidney seen completely surrounded by fat with maximum thickness of 10cm. Renal pelvis normal. Cut surface of kidney shows multiple tiny yellowish area largest measuring 0.4×0.4cm.

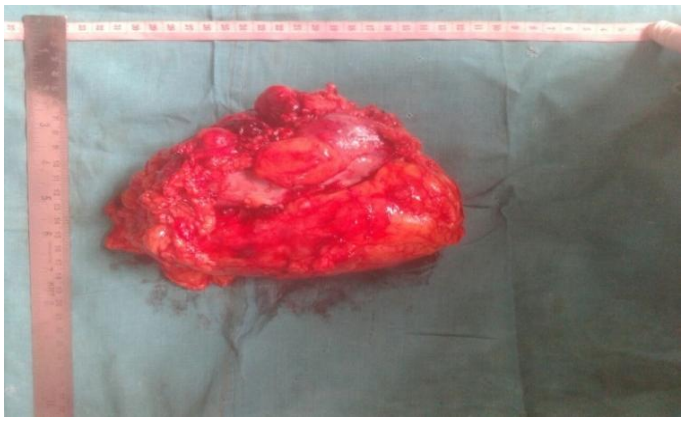

Figure 2: Right nephrectomy specimen –kidney completely surrounded by fat.

Microscopically, peculiar growth pattern identified lobules of mature adipocytes separated by fibro cartilagenous stroma, well circumscribed nodules displaying jumbled up admixture of proliferating benign spindle cells with elongated blunt cigar shaped nuclei, short fasicles and whorled pattern, thickened vessel walls. Kidney architexture well preserved and capsule well preserved.

Final impression was perinephric angiomyolipoma with multiple microscopic angiomyolipoma in the renal parenchyma with peculiar growth pattern encompassing the whole kidney.

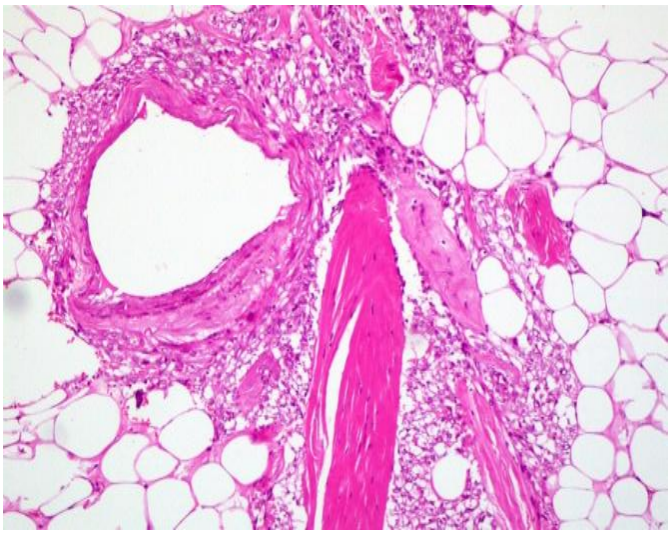

Figure 3: Microphotograph-renal AML tumor showing adipocytes, proliferating spindle cells, thickened vessels(40X).

Discussion:

Angiomyolipoma is a rare benign tumour of kidney. It is found in approximately 45-80% of patients with tuberous sclerosis and are typically bilateral and asymptomatic with F:M predominance of 2:1. In contrast, of the 60-70% patients with AML who do not have tuberous sclerosis present later in life, during 5^{th} and 6^{th} decade and this tumour can be unilateral and tend to be larger than those asssociated with tuberous sclerosis [6].

AML occur not only as a rare tumour which is restricted to kidney, but also as a biologically fascinating and morphologically heterogenous entity. It is also seen in various other sites like skin, appendix, colon, liver, lung and smooth muscles fibres [7]. Differential diagnosis are subtypes of sarcoma including fibrosarcoma, leiomyosarcoma and renal cell carcinoma [8].

Renal epithelioid angiomyolipoma is a recently recognised rare variant which originates from the perivascular epithelioid cell (PEC) and it has aggressive clinical behaviour which includes local

recurrence and metastasis. Hence one should extensively search for epithelioid component. EAML is associated with loss of 16p chromosome and TSC2 [7]. It mimics renal cell carcinoma in imaging studies when it contain less fat, it can be diagnosed accurately with immunostaining with HMB-45, Melanin-A,CD68,CD117 and KI-67 [9].

CTScan, USG, FNAC, Immuno histochemistry play a vital role in diagnosis of renal angiomyolipoma [10].

The main complication of AML is haemorrhage which is related to tumour size, increased vascularity and abnormal thickened blood vessels. Conventional AML has got good prognosis as compared to rare EAML, which is potentially malignant [5].

The patients with intermediate features or should be managed proactively because the likely diagnosis in most cases is renal cell carcinoma. Patients with isolated lesion less than 4cm, can be followed up with a yearly CT Scan or USG to define the growth rate and clinical significance. Patients with asymptomatic or mildly symptomatic lesion greater than 4cm should be followed up with semiannual USG. Patients with lesion greater than 4cm with moderate or severe symptoms (bleeding or pain) should undergo surgical intervention either in form of tumour excision with or without nephrectomy, renal sparing surgery or renal arterial embolization [11].

Initial management of EAML consists of surgical excision. Long term surveillance highly recommended as disease recurrence documented after 5 years of excision [12]. Bleeker et al recently introduced a modified Folpe criteria for the risk stratification of PEComas. Tumours defined as malignant exhibit two or more of the following features: size>5cm, infiltrative growth pattern, high nuclear grade cellularity, mitotic rate>1/50 HPG, necrosis or vascular invasion [13].

For disease recurrence and metastasis, recent studies have demonstrated success with mTOR inhibitors which targets mTOR cell signal pathway involved in RNA translation, which induces TSC2 gene product tuberculin [14,15].

Conclusion:

Angiomyolipoma is a rare tumour of kidney. They can be confused with malignant tumours especially when the presentation is unilateral. CT Scan and USG help in diagnosis. Symptomatic patients require surgical excision.

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