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Diagnosis of "Wunder" ment! - A Rare Case Report.

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ABSTRACT

Angiomyolipoma is a relatively rare benign mesenchymal renal tumour. It was earlier considered to be a hamartoma, however the clonality of origin is now well established and it is known to belong to a family of lesions characterized by the proliferation of perivascular epithelioid cells (PEComa) . Herein, we report a case of multiple bilateral renal angiomyolipomas presenting with Wunderlich's syndrome in a 53 year old women.

Keywords: Angiomyolipoma, Wunderlich's syndrome, Renal tumor,Histopathology

Introduction

Angiomyolipoma is a relatively rare benign mesenchymal renal tumour [1]. It was earlier considered to be a hamartoma, however the clonality of origin is now well established and it is known to belong to a family of lesions characterized by the proliferation of perivascular epithelioid cells (PEComa) [2]. Majorities are sporadic and a lesser number are seen associated with Tuberous Sclerosis which usually are bilateral and multiple [2]. Those measuring more than 10 cm are called "Giant Angiomyolipomas" and occur much less frequently [3,4]. Most cases are diagnosed incidentally on imaging while only a minority presents symptomatically [1]. The most common symptom when present is flank pain caused due to compression of adjacent structures or as a result of hemorrhage, one such presentation is known as Wunderlich's syndrome [1,5] Herein, we report a case of multiple bilateral renal angiomyolipomas presenting with Wunderlich's syndrome.

Case Details:

A 53 year old woman with no prior personal or familial history consulted with chief complaints of right flank pain which was sudden in onset and severe. There was no history of trauma or hematuria. On examination, tenderness over the right flank was observed and vitals were stable. Abdominal ultrasound showed suspicious masses in bilateral kidneys. Plain and contrast enhanced abdominopelvic computed tomography scan (CT) was performed which revealed bilateral multiple renal angiomyolipomas. The largest lesion in the right kidney measured 11.6 x 9.3 cm (Fig. 1a) and largest in the left kidney measured 2.0x1.7 cm (Fig. 1b). Also noted were multiple perinephric, retroperitoneal as well as intralesional hemorrhages on the right side (Fig. 2a, 2b) Though there was bilateral involvement, as the left kidney had only small lesions and was functional, it was preserved and right nephrectomy was done. Our Pathology department received distorted right nephrectomy specimen measuring 19x14x10 cm. External surface showed lobulations (Fig. 2a). Cut section showed variegated appearance with hemorrhagic and a few solid yellowish and grey-white areas. Peripherally pushed and intermixed normal renal parenchyma was also seen (Fig.2b).

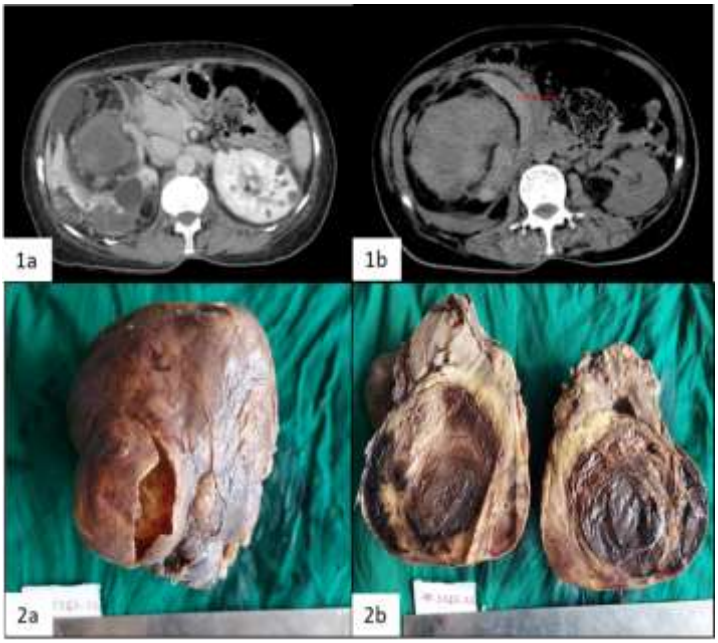

Fig 1a- Abdominopelvic CT showing multiple small AMLs in the left kidney and a giant AML in the right kidney (fat attenuation effect),

1b-Abdominopelvic CT showing perinephric and retroperitoneal hemorrhage,

Fig 2a- Gross image of right nephrectomy specimen, external surface with lobulations

2b- Cut surface showing well demarcated lesion with hemorrhagic areas, solid grey white areas, yellow areas and surrounding normal renal parenchyma.

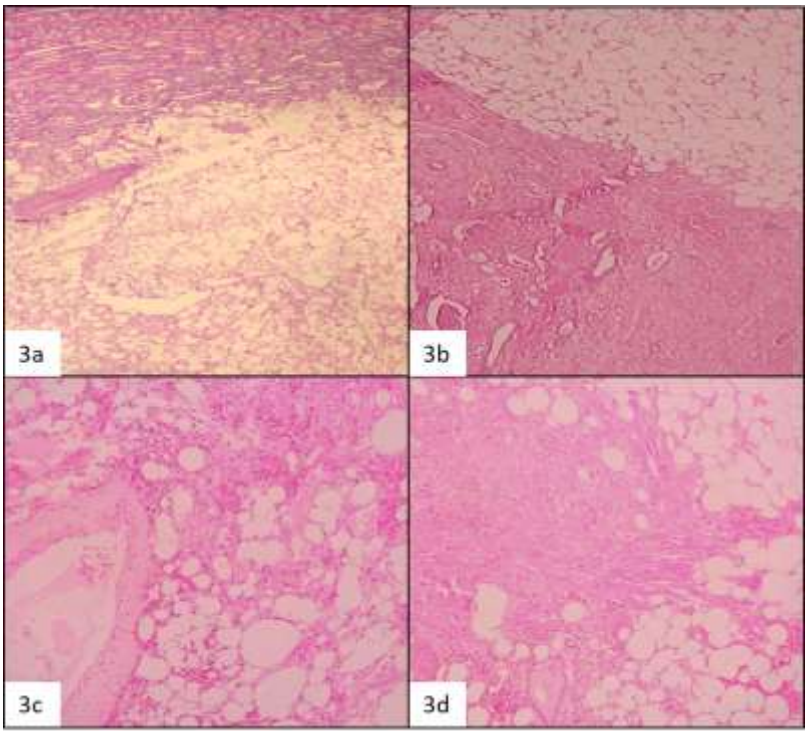

Fig 3a and 3b- Histopathology sections showing lesion predominantly composed of adipose component with surrounding normal renal parenchyma (H and E,4x),

3c-Histopathology section showing various components of the angiomyolipoma- blood vessels, adipose tissue component and hemorrhage; 3d-Histopathology section Tumour showing adipose tissue with smooth muscle component (H and E, 10x)

On microscopic evaluation, it showed multiple lesions with interspersed renal parenchyma (Fig. 3a, 3b), the largest lesion showed the classic triphasic features such as mature adipose tissue which was predominant intermixed with thick walled poorly organized vessels and haphazardly arranged smooth muscle(Fig 3c, 3d) The smaller lesions in the periphery were predominantly composed of adipocytic component. Also noted were areas of hemorrhage.

Diagnosis of renal Angiomyolipoma was given on histopathology. Since the patient presented with acute flank pain together with renal tumors on CT, the case was labeled as multiple bilateral renal angiomyolipomas with Wunderlich's syndrome

Discussion

Renal angiomyolipoma is a rare benign neoplasm with an incidence rate of 0.3-3% with a strong female predilection (Male: Female ratio – 4:1) [5,6,7]. In 80% of the cases, it is sporadic and in these patients, the tumours are unilateral, solitary and slow growing. The remaining 20% of the cases develop in those with genetic disorders, most commonly tuberous sclerosis and the tumours in them usually present at a younger age, are multiple, bilateral and grow faster. Clinically, most sporadic cases are asymptomatic and diagnosed incidentally. The possibility of symptoms increase with the size above 4 cm [2]. The high fat content facilitates the diagnosis by ultrasound or CT [1]. Tuberous sclerosis associated angiomyolipomas, in contrast, are diagnosed at earlier age. Rarely, patients may present with hemorrhagic complications like Wunderlich's syndrome, as in the current case, or post traumatic bleeding owing to the high vascular content of the tumour [8].

Wunderlich's syndrome is a rare but potentially life threatening complication of renal angiomyolipoma that usually occurs in tumours that measure >4 cm in the greatest dimension. Clinically it may present with flank pain and tenderness, palpable mass, gross hematuria and/or hypovolemic shock [8]. Grossly they vary in size from 1 cm to 20 cm or more in diameter. Though is no definite consensus regarding the size cut off for 'giant' AMLs, there are few authors who suggest a size of more than 10cm in greatest dimension qualifies as giant AML. In continuation of the context of size, it is imperative to know that the tumours above 4 cm in size have a higher risk of bleeding complications [3,4] The gross colour varies in conjunction with predominant component and the presence of hemorrhage, most commonly they are yellowish owing to the adipose tissue content. The case in discussion similarly had yellowish areas admixed with hemorrhagic areas. They are unencapsulated but well demarcated and may be locally infiltrative [1,2]

Histologically, it is classically composed of lipomatous, angiomatous and smooth muscle components in varying proportions. Diagnosis is usually straightforward as the microscopic features of this tumour are usually characteristic unless there is a rare instance of epithelioid change in the smooth muscle component which may mimic an epithelial tumour. In cases with a marked predominance of the adipocytic component, extensive sampling may be necessary to differentiate it from a lipoma [1,2,9]. There have been instance where local lymph nodes and/or the renal vein showed fragments of the tumour but this should not be considered as a metastasis [2].

The treatment for AMLs is determined by factors such as size, multiple or giant tumors, preserving the intervening normal renal parenchyma and symptoms due to compression of adjacent structures and hemorrhagic complications. Most cases of giant angiomyolipomas or those with severe symptoms or hemorrhagic complications are treated with nephrectomy as in our case. In other patients, active surveillance may be an option [9].

Although the presentation in this case being multiple, bilateral and giant angiomyolipomas points towards tuberous sclerosis association, the patient did not have history of epilepsy and did not have Ashleaf spots, Shagreens patches or facial nodules. However she was not evaluated for brain or heart tumours characteristic for tuberous sclerosis.

In summary, we present a rare case of giant and multiple bilateral renal angiomyolipomas presenting with Wunderlich's syndrome and was not associated with classic tuberous sclerosis features. These patients with multiple and/or giant lesions are usually symptomatic owing to the size of the tumour and diagnosed quite easily with a CT scan due to the fat attenuation effect. However, histopathological examination remains the gold standard for diagnosis with identification of multiple components. The presence of multiple lesions always warrants an examination and/or investigation for associated tuberous sclerosis related lesions such as facial angiofibromas, cardiac rhabdomyomas, lymphangioleiomyomas, subependymal nodules and cortical tubers. Though these tumours may require nephrectomy but have a good prognosis.

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