Case 13685

Giant, sporadic, multicentric, locally aggressive, renal angiomyolipoma, complicated by emphysematous pyelonephritis, pseudoaneurysm and Wunderlich syndrome

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Section: Uroradiology & genital male imaging
Area of Interest: Abdomen Kidney Oncology Vascular
Procedure: Diagnostic procedure
Procedure: Contrast agent-intravenous
Procedure: Contrast agent-oral
Technique: Ultrasound
Technique: CT
Special Focus: Neoplasia Haemorrhage Aneurysms
Case Type: Clinical Cases
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Patient: 35 years, female

Clinical History:

A 35-year-old female patient presented with complaints of breathlessness, tachycardia and acute onset pain in the abdomen for 4 hours preceded by fever and burning micturation for 2 days. A diffuse abdominal lump was palpable. Her haemoglobin was 8.4g/dl, TLC-17600/ml and urine culture was positive for E. coli with elevated CRP.

Imaging Findings:

Ultrasound and CECT of abdomen were done, which revealed a large 22(CC)x 15(TR)x 17(AP) cm mass lesion arising from the upper pole of the left kidney (Fig.1, 2). The lesion was heterogeneous on ultrasound and showed internal necrotic/haemorrhagic areas with internal hyperechoic areas with fat density on CT. On high resolution ultrasound, the lesion showed soft tissue components resembling muscle tissue (Fig. 3). The rest of the left kidney showed multiple well-defined hyperechoic lesions of variable sizes with fat density on CT, suggestive of multiple angiomyolipomas (Fig. 4). Mild perinephric collection and free fluid was noted in the abdomen (Fig. 2, 5). On CECT abdomen, multiple vascular channels, enhancing soft tissue, fat density and multiple air foci were noted in the lesion with peri-lesional fat stranding (Fig. 5, 6). An area of contrast pooling was noted, suggestive of pseudo-aneurysm formation (Fig. 7). The lesion showed loss of fat planes with descending colon. The right kidney was normal. MRI of the brain was normal.

Discussion:

Based on the imaging and laboratory findings, the provisional diagnosis was giant, multicentric, left renal angiomyolipoma (AML) with intralesional haemorrhage, extracapsular rupture and haemoperitoneum, with intralesional pseudoaneurysm and emphysematous nephritis. The patient was treated conservatively with medical
management by fluid resuscitation and systemic antibiotics and responded to medical management with spontaneous resolution of internal bleeding. The nephritis resolved by systemic antibiotics. Biopsy of the renal lesion was done, which suggested epithelioid variety of angiomyolipoma positive for HMB-45 stain characteristic of angiomyolipoma. Tumour cells were also noted in biopsy specimen from perirenal adipose tissue suggestive of locally aggressive tumour. Nephrectomy was advised as definitive management but the patient refused surgery.

AML is a rare, complex mesenchymal tumour, characterised by proliferation of blood vessels, smooth muscle and adipose tissue elements, occurring in tuberous sclerosis or sporadically. It is considered a benign neoplasm, even though it may exhibit atypical histological features, involve regional lymph nodes and be multifocal. [1–4] AML occur most commonly in the kidney. Even in this location, they are rare, comprising 0.3% of all renal neoplasms. [1] The presence of fat density and aneurysm like vascular markings of renal AML are characteristic findings that allow its diagnosis by ultrasonography and CT. [5] Recently, an epithelioid variant of AML (EAML) has been described, and is thought to be more aggressive. Extrarenal AMLs (ERAML), especially epithelioid types, may not be diagnosed on imaging, as they often lack fat densities. Immunoreactivity to HMB-45 stain is a useful tool to differentiate ERLMs from other tumours.

Clinical features of AML vary depending on the presence or absence of tuberous sclerosis. With the increasing use of imaging modalities, AMLs are often detected incidentally as hyperechoic lesions on ultrasound or well-defined fat density lesions on CT. As the size of the lesion increases, it becomes more heterogeneous and can mimic other renal tumours like renal cell carcinoma and renal oncocytoma. AMLs with a size of more than 4 cm are at an increased risk of rupture and haemorrhage either spontaneously or with minimal trauma and can be fatal due to haemorrhagic shock, hence prophylactic treatment merits consideration in these patients [6]. Wunderlich syndrome (spontaneous non-traumatic renal haemorrhage) is clinically characterized by acute flank pain, flank mass and hypovolaemic shock. If the patient is responsive to fluid resuscitation and the haemorrhage is self-limiting, the patient can be managed conservatively. Angiographic subselective particle embolization has superseded surgical techniques in most cases [7].

**Differential Diagnosis List:** Aggressive, multicentric angiomyolipoma with emphysematous pyelonephritis, psuedoaneurysm and Wunderlich syndrome, Renal cell carcinoma, Liposarcoma

**Final Diagnosis:** Aggressive, multicentric angiomyolipoma with emphysematous pyelonephritis, psuedoaneurysm and Wunderlich syndrome

**References:**

Description: Ultrasound panoramic view of left renal lesion showing a 22 x 11 cm mixed echogenic mass lesion with internal hyperechoic fat and areas of necrosis/collection. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: Ultrasound image shows a large mass lesion arising from upper and mid polar region of left kidney. Mild perinephric collection is noted. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: High resolution ultrasound (9MHz) of lesion shows myomatous component in the lesion resembling the superficial rectus muscle. The lesion shows internal hyperechoic area suggestive of fat. Area of haemorrhage/necrosis is noted. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: High resolution ultrasound (9Mhz) of lower pole of left kidney shows multiple geographic hyperechoic lesions in lower pole suggestive of multiple angiomyolipomas. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: Large mass lesion is seen arising from upper and mid polar region of left kidney. The lesion shows enhancing soft tissue component, hypodense fat component and internal air foci. Mild hyperdense free fluid is noted. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: Large mass lesion with enhancing soft tissue and fat component is noted arising from left kidney. The lesion shows internal air foci. Hyperdense free fluid is seen in Morrison's pouch. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: CECT sagittal section through the lesion shows pooling of intravenous contrast within the lesion suggestive of pseudo-aneurysm formation. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: CECT sagittal section through the lesion shows pooling of intravenous contrast within the lesion suggestive of pseudo-aneurysm formation. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.
Description: Corresponding axial ultrasound image shows the pseudo-aneurysm as a collection with internal echoes. Origin: Department of Radiology, P.D.U. Medical College, Rajkot.