Wunderlich's syndrome - Spontaneous rupture of a giant renal angiomyolipoma

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Abstract :

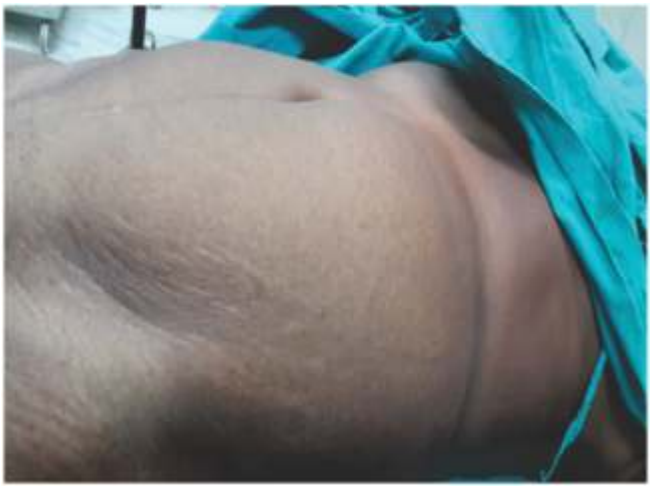
This is a rare case of spontaneous rupture of a giant renal angiomyolipoma (AML), with symptoms of hypovolemic shock (Wunderlich's syndrome), which was managed by total nephrectomy. Enlarged renal AMLs can rupture. This can be sudden and painful with manifestations of hypovolemic shock. The management of AMLs has been correlated with symptoms. Patients with life-threatening retroperitoneal haemorrhage, require exploration as retroperitoneal bleeding can lead to severe complications, increasing morbidity. In case of giant angiomyolipoma with intratumoral haemorrhage, and symptoms of Wunderlich's syndrome, partial or total nephrectomy is a good treatment option.

Keywords : Giant angiomyolipoma (AML) of kidney, Wunderlich's syndrome, Hypovolemic shock, Retroperitoneal hemorrhage, Nephrectomy.

Introduction : Renal angiomyolipoma (AML) is an infrequent tumor that, in most cases, follows a benign course and has clearly defined radiological [1] and histological characteristics [2]. Enlarging AMLs can develop micro and macro-aneurysms that can rupture. This can be sudden and painful, and occasionally life threatening. In 10% of the cases, they manifest as hypovolemic shock, resulting from massive retroperitoneal bleeding, known as Wunderlich's syndrome[3]. The frequency of symptoms and the risk of bleeding due to rupture increases with the size of AML.

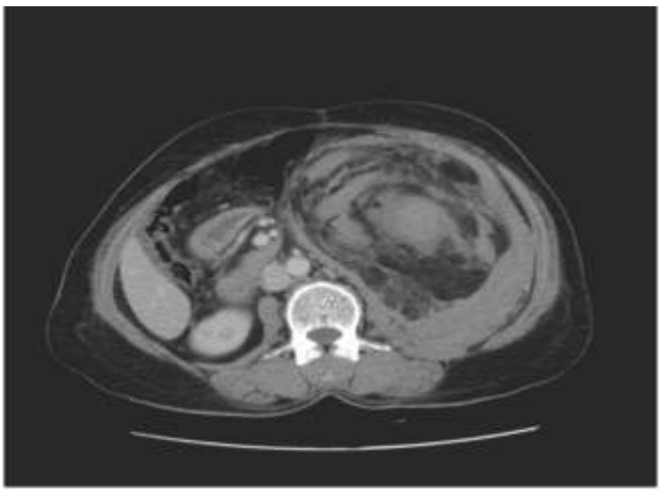
This is a rare case of hypovolemic shock due to rupture of a giant renal AML (Wunderlich's syndrome), which was managed by total nephrectomy.

Presentation of case : A 56 years old female was admitted to hospital with complaint of acute left flank pain of 6 hrs duration, accompanied with progressive painful swelling of the left lateral abdominal region with retention of urine (Fig. 1).



(Fig. 1 Swelling of the left lateral abdominal area.)

She was a known case of of ischemic heart disease. On clinical examination, a hard painful swelling was noted along her left abdomen region. She was having pallor, dehydration, tachycardia & hypotension. Her blood pressure was 88/60 mm of Hg, pulse 120 per min. Patient was catheterized immediately. Hematuria was absent. An emergent ultrasonic examination revealed perinephric fat stranding & disruption of normal architecture of lower pole of the left kidney. Her Hb was 6.5 gm/dl. Her anti-coagulant profile was deranged with electrolyte imbalance. Patient was resuscitated with IV fluids & blood components. Lump decreased on size & patient got relief after conservative management. Computer tomography scanning revealed well defined heterogenous fat containing lesion seen at lower pole of left kidney with hyperdense areas seen within s/o bleed which was most likely to be suggestive of angiomyolipoma with ? rupture with perinephric hematoma & fat stranding (Fig. 2).



(Fig. 2 CT Scan)

So we planned for left nephrectomy. A roof top incision was taken. No free blood was found in the peritoneal cavity, no disruptions of the retroperitoneum was there and a non-pulsatile large mass occupied all of the left retroperitoneal area extending upto left dome of diaphragm & spleen was found on exploration. The left posterior parietal peritoneum was transected and the abdominal aorta was checked blindly up to the level of the renal arteries. Renal artery was ligated first to achieve hemostasis. Then we ligated renal vein. The giant haemorrhagic mass was found originating from the lower pole of the left kidney, was mobilized and removed en-block with the left kidney & a part of left ureter after extensive tissue dissection (Fig. 3 & 4). The retroperitoneal area was washed, a haemostasis was done. Drain was placed in the left paracolic region and the abdominal wall was closed.

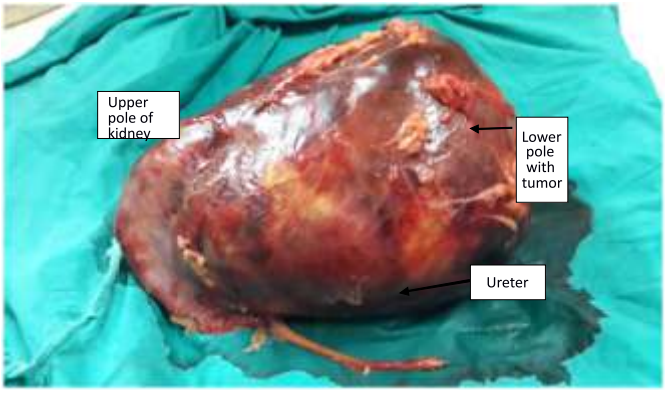


Fig. 3 Nonpulsatile mass

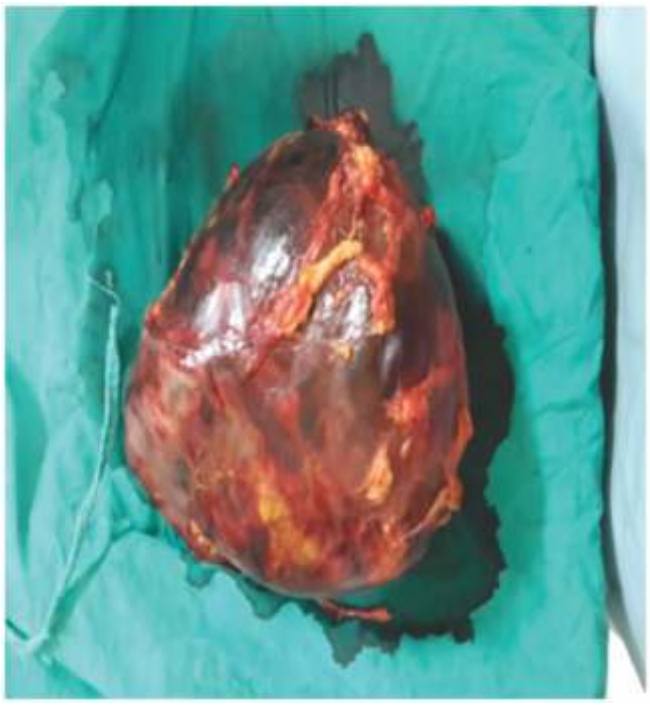


Fig. 4 Mass

(Fig. 3 & 4 : A non-pulsatile large irregular mass occupied all of the left retroperitoneal area extending upto left dome of diaphragm & spleen. The surgical specimen: Giant haemorrhagic mass (24×16×10 cms) originated from the lower pole of the left kidney, en-block with the left kidney.)

Her recovery was uneventful. The patient was discharged from the hospital on the 13th post-operative day after suture removal.

Routine biochemistry and blood analysis at 1 month post-operative follow-up were normal.

Discussion : The AMLs are uncommon tumors. They were first described in 1951 by Morgan et al.[4]. The overall prevalence of AMLs, in population, is 0.44% with a pronounced female predominance (2:1)[5]. In most patients it presents later in life, during the fifth or sixth decade. In our present case the patient was 56 years old female.

Most of these tumors appear in the kidney, however these can be found in other locations, such as the spleen, liver, uterus and fallopian tubes [6],[7].

Most small AMLs are asymptomatic and found incidentally upon radiological studies [7].

Renal AML is a benign tumor known to occur sporadically and in association with genetic syndromes like tuberous sclerosis (TS) and lymphangioleiomyomatosis. Patients with lymphangioleiomyomatosis may have multiple renal and hepatic AMLs, multiple pulmonary cysts, enlarged abdominal lymph nodes and lymphangiomyomas [7], [8]. Renal AMLs arise from the mesenchymal elements of the kidney and they are composed of varying proportions of mature adipose tissue, smooth muscle and abnormally thick-walled blood vessels [9]. For this reason they are also known as renal hamartomas.

AMLs are found in approximately 45–80% of patients with TS and tend to occur in adolescents and young adults. Tuberous sclerosis is a familiarly inherited disorder comprising adenoma sebaceum, mental retardation and epilepsy [10]. Bilateral renal AML in TS is a rare entity and the female/male prevalence is nearly equal [11]. In our case, fortunately, these findings were absent.

The classical clinical presentation of AML is a palpable tender mass, flank pain and gross haematuria, known as Lenk's triad [12]. In our case hematuria was absent. Less frequent associated symptoms include nausea or vomiting, fever, anaemia and blood pressure alteration.

The two major morbidities associated with renal AML are retroperitoneal haemorrhage and impingement of

AML on the kidneys and other vital organs [7]. Enlarged AMLs can develop micro and macro-aneurysms that can rupture. This can be sudden and painful and occasionally life threatening. In 10% of the cases, they manifest as hypovolemic shock resulting from massive retroperitoneal bleeding, known as Wunderlich's syndrome [3],[12]. The frequency of symptoms and the risk of bleeding (rupture) increases with the size of an AML [13]. In our case report, the tumor size was 24×16×10 cms. So possibility of rupture was suspected.

Rupture can occur spontaneously in patients on anticoagulants, or with trauma to the kidney. Rupture has also been reported during pregnancy or in the post-partum period [7]. Our patient did not have this kind of history. So we fortunately got better outcome.

AML is the only benign renal tumor that is confidently diagnosed on cross-sectional imaging. There are two imaging characteristics that are highly suggestive of an angiomyolipoma. On ultrasound, they are echogenic. On CT scanning, they have the density of fat, which is less than that of water[14]. The presence of fat (confirmed on non-enhanced thin-cut computed tomography) by a value of −20 Hounsfield units or less seen within a renal lesion on imaging is considered the diagnostic hallmark. MRI can be used to identify the fatty tissue. However because the presence of bleeding in any renal tumor can mimic the typical pattern of angiomyolipoma, MRI should not be considered the diagnostic method of choice. AML can vary in size from a few millimeters to larger than 20 cm. It is unusual to see an AML over 10 cm, therefore many studies have demonstrated that any AML measuring over that number is considered as “giant”. CT findings usually help to differentiate AMLs from other tumors, such as perinephric liposarcomas [1].

Arterial angiography can reveal neovascularity, similar to that of renal cancer and therefore it is not helpful in differential diagnosis.

The management of AMLs has been correlated with symptoms. Most patients with small tumors (<4 cm) that tend to be asymptomatic are managed conservatively, under periodic ultrasonography follow up [13].

Tumors larger than 8 cm as in our case, generally tend to be symptomatic and they are at a greater risk of spontaneous or traumatic rupture resulting in haemorrhagic complications. These patients are therefore treated by angiography and selective arterial embolisation as a first line [14]. Patients with life-threatening haemorrhage, require exploration and in the majority of the cases require total nephrectomy [15].

We considered nephrectomy since our patient was having acute drop of haematocrit levels, increased size of retroperitoneal haematoma and hypovolemic shock (Wunderlich's syndrome).

Histopathologically, AML consists of mature adipocytes, thick-walled blood vessels, and epithelioid stromal cells in various proportions[9]. Usually it displays as a pattern of typical fat and perivascular epithelioid cells arranged around a blood vessel. In our case, histopathological findings were consistent with rupture & heamorrhage in renal angiomyolipoma.

Conclusion : Patients with a life-threatening angiomyolipoma haemorrhage, require intervention, as retroperitoneal bleeding can lead to severe complications, increasing morbidity. In case of giant angiomyolipoma with intratumoral haemorrhage, and symptoms of Wunderlich's syndrome, partial or total nephrectomy is a good treatment option in order to save the patient's life.

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