

Wunderlich Syndrome: A Rare Clinical Syndrome

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ABSTRACT

Introduction: Wunderlich's syndrome is a rare clinical condition characterized by acute onset of spontaneous, non-traumatic renal haemorrhage into the subcapsular and perirenal space. They have characteristic features on ultrasound and CT scanning which may enable their diagnosis pre-operatively.

Case report: A case of renal angiomyolipoma with perirenal hematoma diagnosed for which partial nephrectomy following conservative route of treatment when the diagnosis is made pre-operatively using radiological investigations.

Conclusion: Management of renal angiomyolipomas should thus be individualised. Symptomatic and large tumours with subcapsular hematoma or haemodynamic instability require surgical intervention. Smaller, asymptomatic tumours may be observed, but require close monitoring by serial CT scanning. Wunderlich's Syndrome is a surgical emergency which should be immediately diagnosed and treated, with minimally invasive embolization or surgical exploration with hematoma evacuation and partial or complete nephrectomy as performed in our case.

Keywords: Angiomyolipoma (AML), Computed Tomography (CT), Partial Nephrectomy.

INTRODUCTION

Wunderlich's syndrome is a rare clinical condition characterized by acute onset of spontaneous, non-traumatic renal haemorrhage into the subcapsular and perirenal space. It is classically characterized by the triad of acute flank pain, flank mass and hypovolemic shock.¹ Renal mass is the most common cause of Wunderlich's Syndrome with angiomyolipoma being the most common benign neoplasm.² Renal angiomyolipomas are uncommon benign tumours which clinically mimic renal cell carcinoma. They have characteristic features on ultrasound and CT scanning which may enable their diagnosis pre-operatively.³ We review our experience of a case of renal angiomyolipoma with perirenal hematoma. We advocate partial nephrectomy following conservative route of treatment when the diagnosis is made pre-operatively. Partial/complete nephrectomy should be performed if there is suspicion of malignancy or a complication like massive hemorrhage.

CASE REPORT

A 45 year old female patient presented with history of left loin pain since 3 days which was found to be radiating to the back, with no other significant history. She was conscious, well oriented and afebrile on evaluation. Her blood pressure was found to be 140/74, Pulse Rate was: 120/min. Abdominal palpation revealed tenderness in the left loin region but no rigidity or guarding was elicited.

Investigations: A CT- KUB was done which revealed an ill-defined fat density lesion measuring 3.9 x 3.9 cm with internal well enhancing foci measuring 1.2cm x 1.0 cm. Disruption of the superior margin of this fat lesion was noted. Diffuse hyper-dense collection with 2.5 cm width was noted surrounding the left kidney with a collection measuring 4.2 cm x 4.8 cm noted inferiorly.(1a,b)

CT angiogram showed left lower pole AngioMyoLipoma being fed by the left lower segmental artery which also formed a pseudoaneurysm within the lesion. These findings alongwith perinephric hematoma were suggestive of Wunderlich Syndrome.(1c)

Partial nephrectomy was performed which revealed a mass arising from the lower pole of left kidney, with surrounding hematoma around the entire left kidney. A 4cmx3 cm mass was resected and hematoma was drained. It was found to be histologically consistent with AML.

DISCUSSION

Spontaneous renal hematoma was first described by Wunderlich in 1856.⁴ Wunderlich's Syndrome is a life threatening medical emergency, defined as spontaneous non-traumatic bleeding confined to perinephric region. Renal neoplasms are the most common cause of Wunderlich's Syndrome with renal AML being the most common benign cause.² Incidence of renal AML as a cause varies between 35-40%.³ Other causes include vasculitis, renal artery aneurysm, arteriovenous malformation and fistula, venous thrombosis, cystic renal diseases, renal infections, nephritis and coagulation disorders.²

AML is a mesenchymal neoplasm of the kidney, composed of variable proportion of mature adipose tissue, smooth muscle and abnormal vessels.⁵ Classic AML may occur sporadically (50-70% of the cases) or in association with tuberous sclerosis (30-50% of the cases).⁶ Ultrasound is valuable for rapid identification of the condition but CT has higher sensitivity and specificity than ultrasound for confirmation of renal mass. As in our case, CECT clearly delineated fat lesion, hematoma, soft tissue and vascular component. MRI might have added benefit over CT scan for

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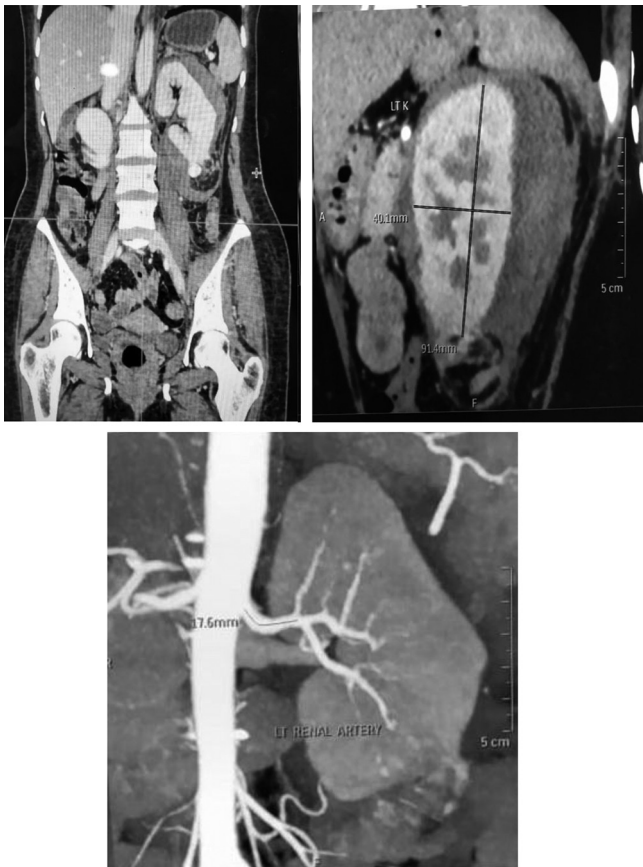


Figure-1: (a) CECT KUB coronal film showing an ill-defined fat density lesion with internal well enhancing foci (pseudoaneurysm); (b) sagittal film showing perirenal hematoma of thickness 2.5 cm; (c) CT angiogram showing left lower pole Angiomyolipoma (AML) being fed by the left lower segmental artery which also formed a pseudoaneurysm within the lesion

evaluating smaller tumors.⁷

Incidence of intratumoral haemorrhage and tumor rupture is known to depend upon tumor size and diameter of the intralesional aneurysm. Larger size (>4 cm) and diameter of intralesional aneurysm (>5mm) correlate directly with tumor related haemorrhage in AML.⁸

Renal angiography with embolization to occlude flow to the pseudoaneurysm is used as the early treatment.⁸ Surgical treatment partial/complete nephrectomy for haemorrhage control or hematoma evacuation in patients with haemodynamic instability or uncontrolled sepsis.⁹ In the present case of Wunderlich's Syndrome, partial nephrectomy with clot evacuation was done in view of hemodynamic instability.

CONCLUSION

The management of renal angiomyolipoma has changed over the last decade. The possibility of making correct radiological diagnosis has encouraged a conservative approach. Management of renal angiomyolipomas should thus be individualised. Symptomatic and large tumours with subcapsular hematoma or haemodynamic instability require surgical intervention. Smaller, asymptomatic tumours may be observed, but require close monitoring by serial CT scanning. Wunderlich's Syndrome is a surgical emergency



Figure-2: (a) Intraoperative picture showing a mass arising from the lower pole of left kidney. (b) Intraoperative picture showing hematoma around the entire left kidney; (c) left: resected renal mass after partial nephrectomy, right: drained hematoma with clots

which should be immediately diagnosed and treated, with minimally invasive embolization or surgical exploration with hematoma evacuation and partial or complete nephrectomy as performed in our case.

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