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Exophytic Renal Angiomyolipoma Unmasked: A Case of Wunderlich's Syndrome

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Abstract: ***Wunderlich syndrome (WS), characterized by spontaneous bleeding into the kidney's surrounding spaces, is a rare but potentially life - threatening condition. This report presents a case of WS secondary to an exophytic renal angiomyolipoma. A 35 - year - old female presented with progressive left flank pain, leukocytosis, and normal hemoglobin levels. Contrast - enhanced Computed Tomography revealed a large fat - containing tumor with active contrast extravasation and hematoma formation over the right perirenal space. Emergency total nephrectomy was performed, confirming the diagnosis of renal angiomyolipoma. WS represents a diagnostic challenge necessitating prompt identification and intervention to mitigate its potentially severe complications. Multidisciplinary collaboration and advanced imaging techniques are crucial for optimal management and patient outcomes in WS cases.***

Keywords: Angiomyolipoma, renal hemorrhage, spontaneous

Abbreviations: WS - Wunderlich syndrome, CT - Computed Tomography.

1. Introduction

Wunderlich syndrome, also referred to as WS and named after Carl Wunderlich, is a rare medical condition characterized by sudden bleeding in the kidney's surrounding spaces, occurring spontaneously without any prior injury. Symptoms can vary from vague flank or abdominal discomfort to severe hypovolemic shock. While a few patients may display the classic Lenk triad of flank pain, a palpable mass, and shock, this is observed in a small fraction of cases. Roughly 60%–65% of WS instances are linked to renal neoplasms like angiomyolipomas and clear cell renal cell carcinomas, which are susceptible to bleeding and rupture.

Angiomyolipomas, benign growths consisting of fat, twisted blood vessels, and smooth muscle, are the primary culprit. These growths often arise sporadically, with around 80% of cases occurring without any apparent cause and the remaining 20% linked to tuberous sclerosis complex. [1, 2] Sporadic angiomyolipomas usually manifest as single nodules in middle - aged women, whereas those associated with tuberous sclerosis complex tend to be bilateral, multiple, and larger in size. [2] Symptoms of angiomyolipomas typically remain silent until the tumor expands significantly or ruptures, triggering Wunderlich syndrome. This spontaneous bleeding in the kidney is an uncommon but potentially life - threatening complication, particularly in angiomyolipomas larger than 4 cm. Typical clinical signs include flank pain, a palpable mass, visible blood in urine, and hypovolemic shock.

Multiphasic CT and MRI scans are vital tools for promptly diagnosing and managing Wunderlich syndrome, as they can accurately identify fat - containing kidney masses and associated bleeding. Additionally, the presence of aneurysms within the lesion on CT scans may heighten the risk of tumor rupture.

2. Case History and Imaging

A 35 - year - old female presented to the emergency department with progressive left flank pain for 7 days. She denied recent traumatic events. Initial vital signs were stable with normal blood pressure. Physical examination revealed left flank tenderness. Laboratory data showed reduced haemoglobin level of 8.4 g/dL. The routine urine analysis showed hematuria. After clinical evaluation, an abdominal contrast enhanced computer tomography (CT) was advised.

CECT showed a large solid exphophytic lesion with predominant fat component arising exophytically from the entire lateral aspect of left kidney with interspersed and surrounding hyperdense areas, active contrast extravasation and hematoma formation over the left perirenal space. A diagnosis of renal angiomyolipoma with spontaneous bleeding was made. She was taken urgently to the operation theatre, where a total nephrectomy was performed. Surgery dictation reported a left kidney with 14cm x 13cm x 9cm dimensions and a perirenal hematoma with 950 mL of blood drained, and multiple neoformed vessels in the renal parenchyma with macroscopic features of an angiomyolipoma. An estimated trans - operative bleeding of 1400 mL was reported. Pathology reported angiomyolipoma.

Image gallery:

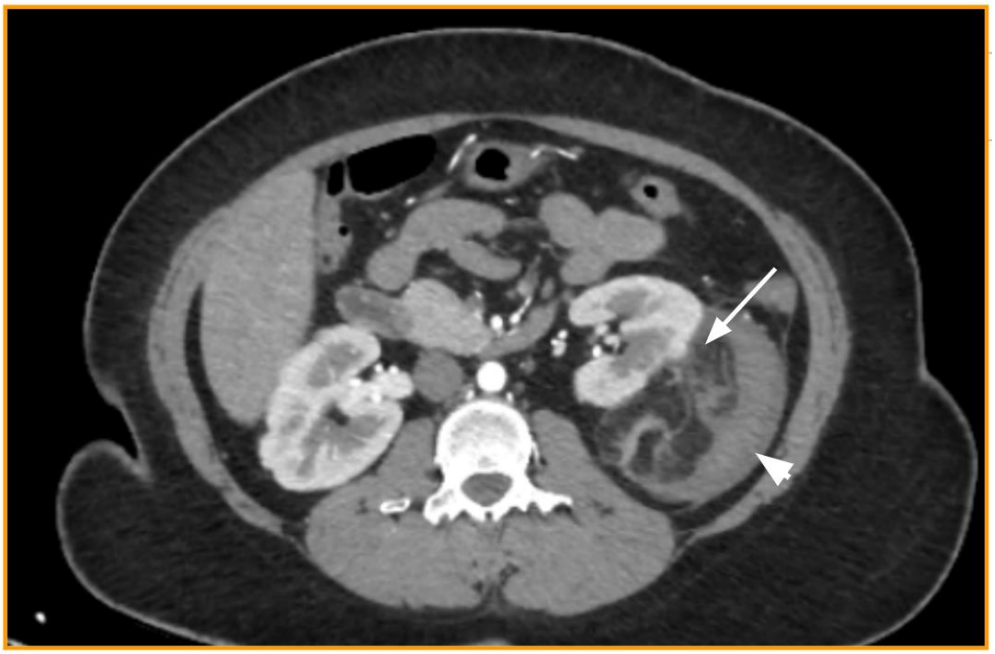

Figure 1: Axial CECT section at the level of kidneys showing an exophytic renal angiomyolipoma (white arrow) with peripheral areas of hemarrhage (arrow head) arising from left kidney.

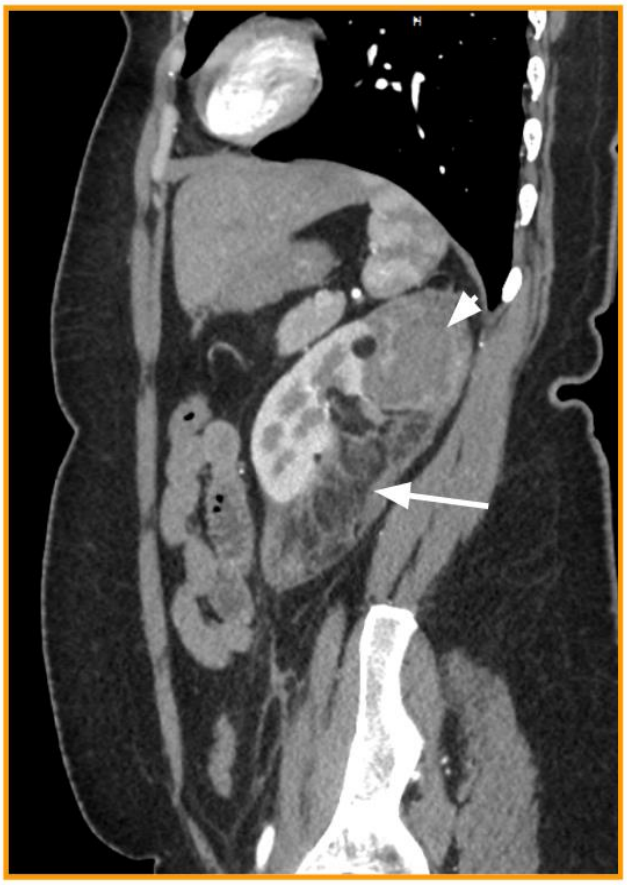

Figure 2: Sagittal reformated image showing exophtyic location of the renal angiomyolipoma (white arrow) with hemorrhagic areas (arrow head).

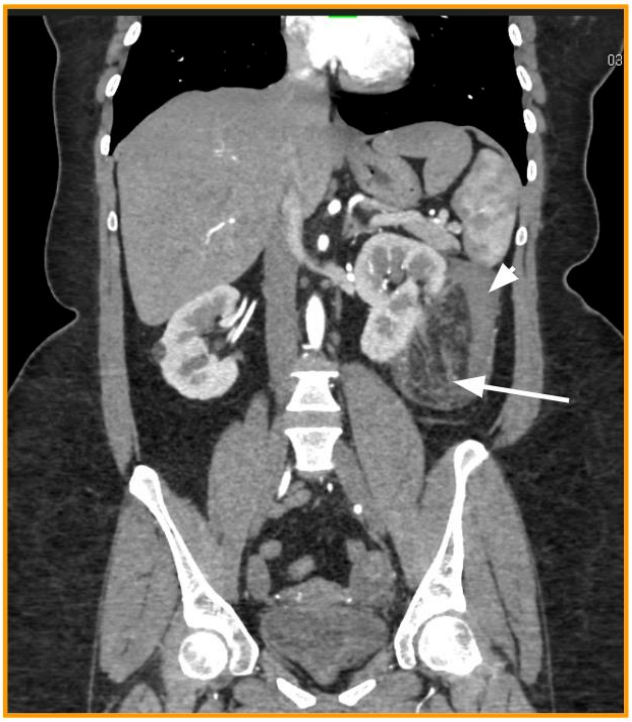

Figure 3: Coronal reformated CECT image showing left renal angiomyolipoma (white arrow) with areas of hemorrhage (arrow head).

3. Discussion

Wunderlich syndrome represents a critical medical condition demanding a collaborative approach for prompt identification and treatment. It denotes spontaneous, non - traumatic hemorrhage occurring within the subcapsular or perirenal spaces [4, 5]. While the classic Lenk triad is evident in only a minority of cases, the clinical scenario of our patient underscores the urgency of early recognition and intervention [4].

In this instance, initial symptoms of flank pain and an abdominal mass rapidly progressed to hypovolemic shock, necessitating urgent laparotomy. This emphasizes the pivotal role of diagnosing WS before hemodynamic instability ensues, facilitated by advanced imaging techniques like CT scans.

CT scans, sensitive to fat - containing renal masses and associated hemorrhage, are pivotal in identifying hallmark features of WS, including intralesional aneurysmal formations heightening the risk of tumor rupture [3].

Moreover, CT scans offer heightened sensitivity in detecting retroperitoneal masses or hemorrhages, boasting a sensitivity range of 92 to 100%. Particularly, angiomyolipomas larger than 4cm pose an elevated risk of spontaneous hemorrhage [6]. Treatment strategies for angiomyolipomas are tailored based on the patient's hemodynamic status, ranging from active surveillance for small, asymptomatic tumors to preoperative angioembolization followed by partial nephrectomy for larger lesions [7] Total nephrectomy is reserved for cases necessitated by renal cell carcinoma or hemodynamic instability. In cases of WS, surgical intervention takes precedence to address potential complications effectively.

The optimal therapeutic approach for WS involves surgical intervention to mitigate the inherent risks associated with this condition. This may encompass either exploratory laparotomy or embolization, contingent upon the tumor size and the prognosis for preserving renal function [6, 7]. Overall, expeditious diagnosis and judicious management are pivotal in optimizing outcomes for patients grappling with WS.

4. Conclusion

In conclusion, Wunderlich syndrome (WS) remains a diagnostic challenge due to its rarity and variable presentation. Prompt recognition of WS is crucial, especially in patients presenting with sudden onset flank pain and hematuria, as it may indicate underlying renal pathology such as angiomyolipomas. Imaging modalities like CT scans play a pivotal role in accurate diagnosis and timely management of WS, allowing for early intervention to mitigate potential life - threatening complications. Treatment strategies, tailored to the patient's hemodynamic status and tumor characteristics, range from active surveillance to surgical intervention, depending on the individual case. Overall, a multidisciplinary approach involving urologists, radiologists, and surgeons is essential for optimizing outcomes in patients with WS.

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