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Understanding Wunderlich Syndrome: Causes, Diagnosis, and Challenges

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Abstract: ***Wunderlich syndrome (WS), which was named after Carl Wunderlich, is a rare clinical syndrome characterized by an acute onset of spontaneous renal hemorrhage into the subcapsular, perirenal, and/or pararenal spaces, without a history of antecedent trauma. Patients may present with a multitude of symptoms ranging from nonspecific flank or abdominal pain to serious manifestations such as hypovolemic shock. The classic symptom complex of flank pain, a flank mass, and hypovolemic shock referred to as the Lenk triad is seen in a small subset of patients. Rare causes of WS include renal infections, cystic diseases, calculi, kidney failure, and coagulation disorders. Cross-sectional imaging modalities, particularly multiphasic CT or MRI, are integral to the detection, localization, and characterization of the underlying causes and facilitate optimal management.***

Keywords: Wunderlich, rare, spontaneous renal hemorrhage, flank mass, hypovolemic shock, Lenk triad

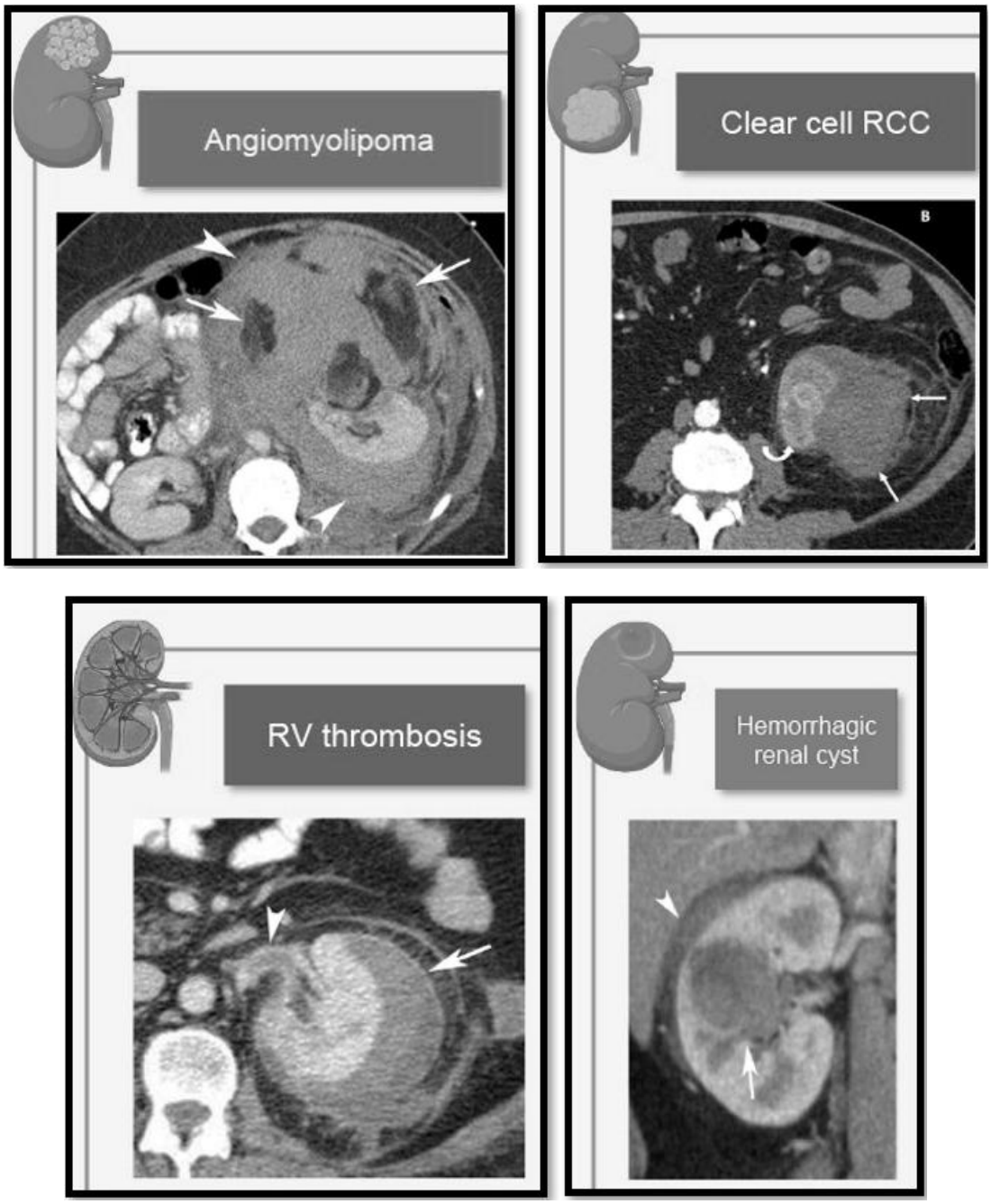
1. Introduction

Wunderlich syndrome is eponymously named after Carl Wunderlich, who first described a patient with spontaneous subcapsular and perinephric bleeding without preceding trauma in 1856. It is also commonly referred to as spontaneous renal hemorrhage in surgical literature. As the name implies, trauma and iatrogenic causes such as recent biopsy or surgical procedure need to be excluded prior to labeling as WS. More than two-thirds of patients present with flank pain as the presenting symptom. In one published series of 13 patients, only 2 patients demonstrated the classic Lenk's triad of pain, mass and hypovolemic shock, while all 13 patients had flank pain.[3] Microscopic hematuria is more common than gross hematuria. Given the nearly universal presenting symptom of acute onset flank pain, the most likely study to be performed in the emergency room would be a noncontrast CT.

Acute hemorrhage is hyperdense to the renal parenchyma on the unenhanced phase, with density ranging from 40 to 70 Hounsfield Units (HU). Clotted blood has a higher density closer to 60 HU, with clots seen closer to the bleeding site, also referred to as the sentinel clot sign. Sentinel clot sign is of greater help in cases of diffuse hemoperitoneum to detect the bleeding organ rather than in retroperitoneal hemorrhage. Density of blood is strongly dependent on the age of the blood, with lower attenuation values if imaged after 48 hours of the bleeding event. Occasionally, a portal venous phase contrast-enhanced CT might be the first study if the patient presented with abdominal rather than flank pain; in such cases, hemorrhage would be hypodense to isodense relative to the enhanced renal parenchyma. In older cases prior to 2005, ultrasound (US) was more commonly performed than CT for evaluation of flank pain. Perinephric and retroperitoneal hemorrhage can have a varied appearance on USG, ranging from uniformly hyperechoic hematomas to mixed density collections.

The clinical significance of promptly recognizing perinephric hemorrhage and its management is two-fold. In the immediate setting, patients with shock and massive hemorrhage have to be stabilized with conservative measures and angiographic or surgical intervention as needed. The more sinister long-term implication is the need to identify the underlying cause as there is a strong association with neoplasm and thus appropriate recommendation for follow up must be conveyed to the emergency room physician. We shall review the common causes, describe the imaging features and provide an update on the surgical management.

Etiologic Factors of Wunderlich Syndrome:



Etiologic Factors of WS

Renal neoplasms • Angiomyolipoma • RCC • Metastases • Renal sarcomas • Urothelial carcinoma • Oncocytoma
Renal vascular diseases • Polyarteritis nodosa • RAAs and pseudoaneurysms • Renal arteriovenous malformations • Renal arteriovenous fistulas • Renal vein thrombosis
Renal infections • Acute pyelonephritis • Emphysematous pyelonephritis • Renal abscesses
Nephritis and kidney failure
Hematologic and coagulation abnormalities • Hemophilia • Anticoagulant drug induced
Renal cystic disorders • Acquired cystic kidney disease • Simple or hemorrhagic renal cyst • Autosomal dominant polycystic kidney disease
Renal calculus
Miscellaneous • Recreational drug use • Idiopathic WS

2. Case Report

A 51 year old male presented to the emergency room with history of left flank pain since 3 days associated with fever

and chills. H/o burning micturition with hesitancy and frequency, hematuria.

On examination: CNS study was unremarkable.
PA: left renal angle tenderness with guarding and rigidity.
RS: was unremarkable
CVS: patient presented with clinical signs of shock
Patient was advised USG KUB with SOS CT KUB.
Patient however underwent CT KUB without any USG screening.

CT KUB

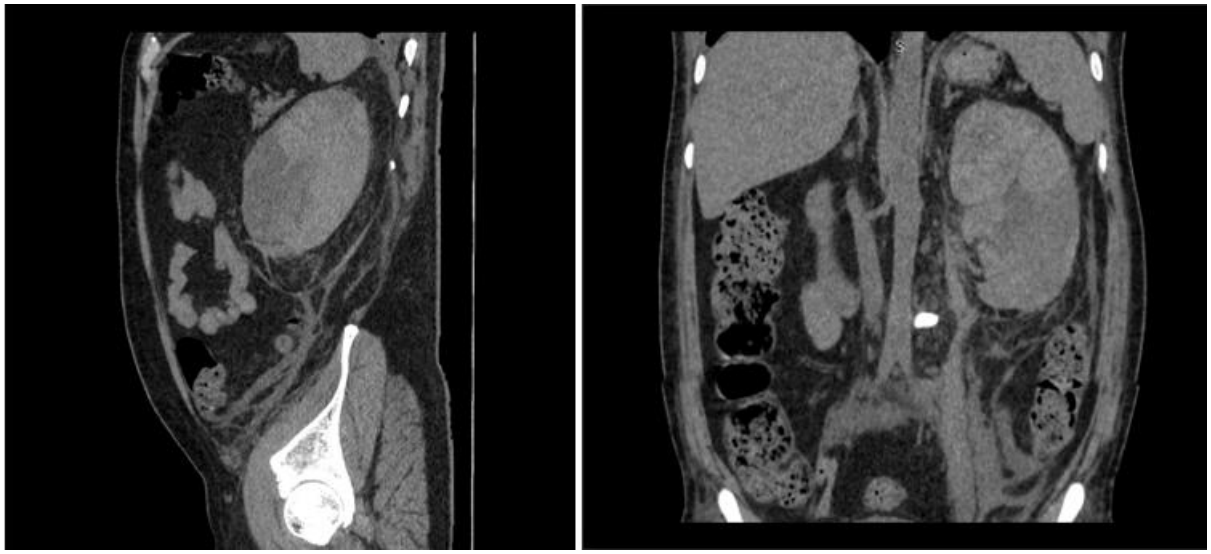


Figure 1: Sagittal section of the left kidney, Coronal section of the left kidney.

Fig 1: Shows the left kidney bulky and heterogeneously attenuating with hyperdensities of near blood attenuation (Average Attenuation + 45 to + 65 HU) seen near replacing the left renal parenchyma with poor appreciation of the renal anatomy, measuring approximately 14.3 x 8.6 cm.

A hyperdense area/collection of near blood attenuation (Average Attenuation + 45 to + 65 HU) is noted in the left pararenal and retroperitoneal space extending from L1 to l5 vertebral bodies, abutting the anterior aspect of the left psoas muscle with loss of intervening fat planes, measuring approximately 21 cm in length and 6.4 cm in maximum width.

CT KUB (CONTINUE)

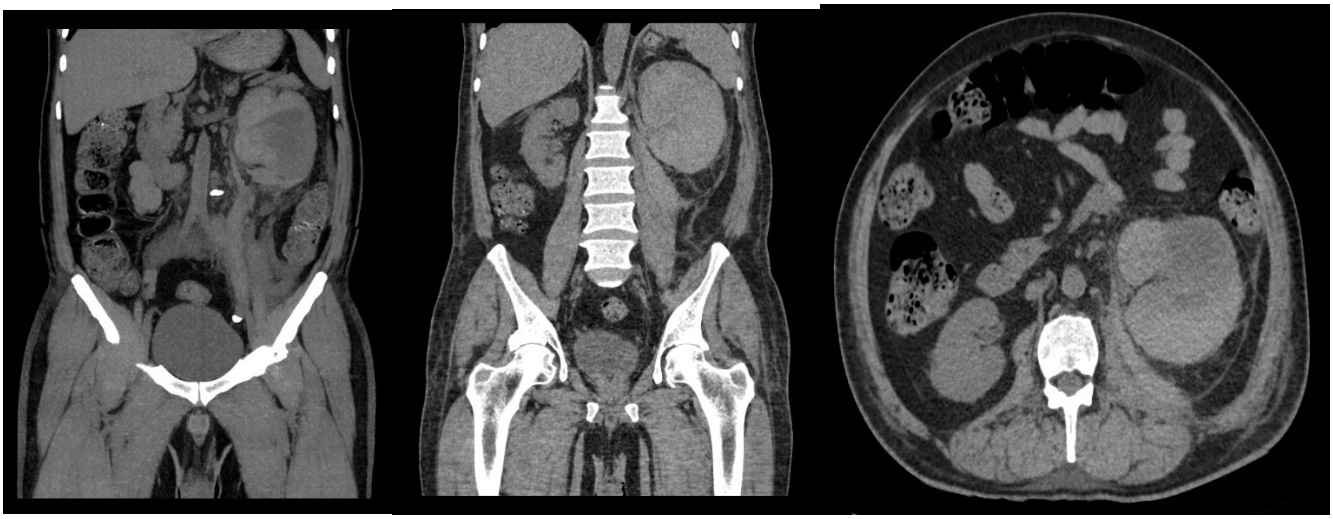


Figure 2: Coronal and Axial sections of KUB

Findings:
Fig 2 Shows A 1.4 x 0.8 cm obstructive calculus (Average Attenuation + 1341 HU) is seen in the left distal ureter approximately 3.1 cm away from the left vesico-ureteric junction causing moderate hydronephrosis and upstream hydroureter, with hyperdense contents also noted in the distal ureter and pelvicalyceal system.

There is moderate to gross left perinephric fat stranding with moderate thickening of the left renal fascia. Left periureteric wall thickening with moderate periureteric fat stranding is noted in proximal and mid ureter. Mild to moderate right perinephric fat stranding with minimal thickening of right renal fascia is also noted.

Post Operative USG was performed.

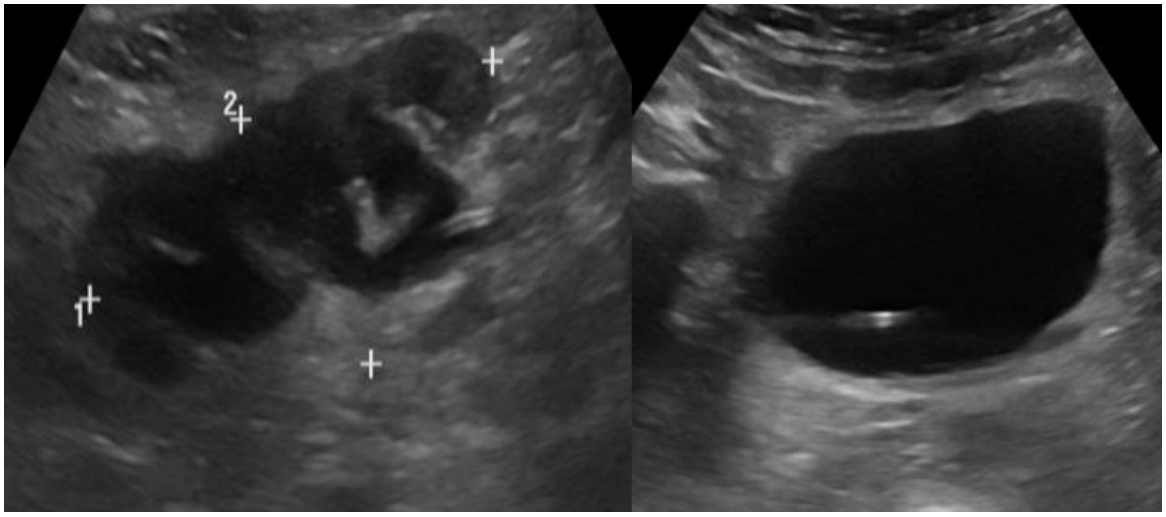


Figure 3: Left kidney and Urinary bladder on USG KUB

Findings

Fig 2 Shows Left kidney measuring approximately 9.5 x 4.7 cm. Proximal end of DJ stent was noted in the mid pole calyces. Mild hydronephrosis with proximal hydroureter was seen. The parenchymal thickens of left kidney at upper pole measured approximately 8.5mm, mid pole 7.2 mm and lower pole 9.1 mm respectively. Post operative changes in form of mildly irregular contour with no evidence of any peri-nephric collection was seen.

Urinary Bladder was well distended and appeared normal in contour and wall thickness. No calculi or mass lesion was noted. Distal end of DJ stent is noted in the mid pole calyces

3. Conclusion

Based on the above-mentioned imaging features and I/v/o blood attenuation possibility of Wunderlich syndrome / peri renal / retroperitoneal hematoma needs consideration. Other possible radiological differentials include obstructive uropathy with hemorrhagic contents within dilated pelvicalyceal systems and ureter with extension in the retroperitoneal space.

4. Discussion

Wunderlich syndrome or spontaneous perinephric hemorrhage is uncommon but radiologists, especially those who work in the emergency section, are likely to encounter it with increasing frequency, given widespread anticoagulation use and greater use of CT in the ER. The most common etiologies are neoplasms, vascular causes and coagulopathies. Once hemorrhage is identified, a multiphase CT is the next step to identify the cause. Active extravasation on imaging indicates need for intervention.

References

[1] Https://pubs.rsna.org/doi/abs/10.1148/rg.230039
[2] https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10904173/
[3] https://www.medicalnewstoday.com/articles/9276
[4] Wang BH, Pureza V, Wang H. A tale of Wünderlich syndrome. J Surg Case Rep 2012;2012(11): rjs015.
[5] Albi G, del Campo L, Tagarro D. Wünderlich's syndrome: causes, diagnosis and radiological management. **Clin Radiol** 2002;57(9):840–845.
[6] Grubb SM, Stuart JI, Harper HM. Sudden onset flank pain: Spontaneous renal rupture. **Am J Emerg Med** 2017;35(11):1787.e1–1787.e3.
[7] Spontaneous perirenal hemorrhage (Wunderlich syndrome): An analysis of 28 cases. Am J Emerg Medial (2019)
[8] Wunderlich's syndrome: Causes, diagnosis and radiological management. Clin Radiol (2002)
[9] Spontaneous perinephric hemorrhage in a 65-year-old man. J Urol (1985)
[10] Spontaneous rupture of the kidney with perirenal hematoma J Urol(1975)