

Wunderlich syndrome due to Renal angiomyolipoma: A case report.Patel N¹, Pandya N^{2*}, Patel U³¹Assistant Professor, ^{2,3}Resident Doctors, Radiology Department, B J Medical College, Civil Hospital Ahmedabad, Gujarat, India.**Abstract:**

Angiomyolipoma is the most common benign renal neoplasm. With increase in size chances of complications increases. Angiomyolipoma greater than 4 cm in size require close follow up & intervention to minimize risk of complication. Wunderlich syndrome or spontaneous renal haemorrhage is a life threatening complication which usually presents as an acute emergency condition. We present a case of sub-acute presentation, Angiomyolipoma with spontaneous haemorrhage & we would also like to demonstrate role of imaging in pre-operative accurate diagnosis & prevention of unnecessary emergency laparotomy.

Keywords: Angiomyolipoma, Tuberous sclerosis, Wunderlich syndrome.

Introduction

Wunderlich syndrome or spontaneous retroperitoneal haemorrhage is a rare non traumatic life threatening haemorrhage. Neoplastic etiology being the most common cause of these kinds of haemorrhages, Angiomyolipoma being the most common benign neoplasm & renal cell carcinoma representing most common malignant neoplasm. Classic angiomyolipoma is benign mesenchymal tumor seen mainly in kidney; consist of adipose tissue, mature smooth muscle & large thick walled capillaries in varying proportion. A second type is containing large amount of perivascular epithelioid

cells making it susceptible to bleed. Incidence among general population ranges from 0.07% to 0.3%. Approximately 80% of them are sporadic in nature while 20% are associated with tuberous sclerosis^[1,2]. Other rare syndromic association includes Neurofibromatosis type I, Von Hippel Lindau & ADPKD syndromes. Sporadic lesions are large, unilateral, single lesion occurs in age of 40 to 60 years with female predominance (1:4). Several studies have demonstrated that frequency of the symptoms & risk of bleeding increases with the tumour size. Approximately 68–75 % angiomyolipoma less than 4 cm in size remain asymptomatic while 82-95% lesions with more than 4 cm in size are symptomatic. Those presenting with symptoms usually present with classic symptoms of abdominal or flank pain,

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Image 1: USG showing fat echogenicity lesion & Peri-lesional hematoma.

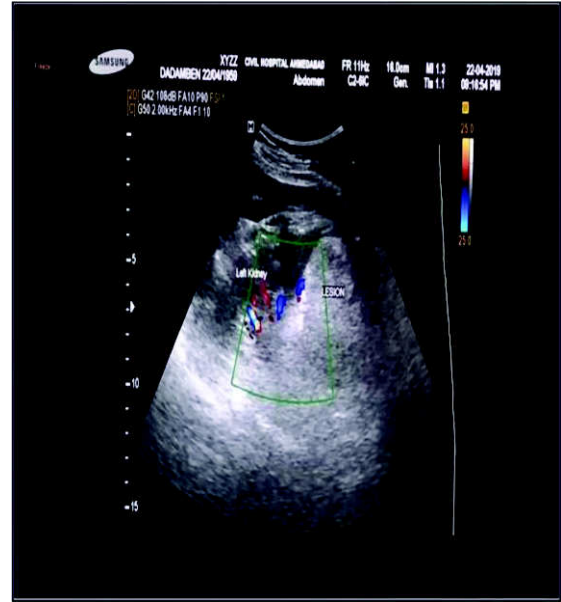


Image 2: USG showing lesion in lower pole of left kidney supplied by branches of left renal artery.

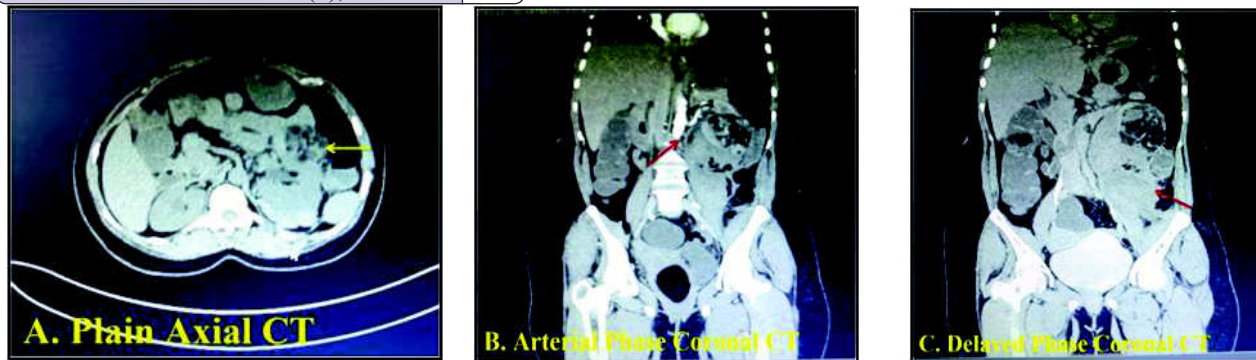


Image 3: CT scan (A) showing fat density lesion in lower pole of left kidney. (B) showing branches arising from left renal artery supplying the lesion. (C) Intra-lesion & retro-peritoneal hematoma along left psoas

palpable mass & gross hematuria (Lank's triad)^[3,4,5]. Other less common symptoms fever, nausea, vomiting, anaemia, hypotension, renal failure. The three hemorrhagic presentation are 1) Non traumatic spontaneous haemorrhage - Wunderlich Syndrome 2) Post traumatic rupture 3) Pregnancy induced changes in hormonal level causing bleed. We present the case of Wunderlich syndrome^[6,7].

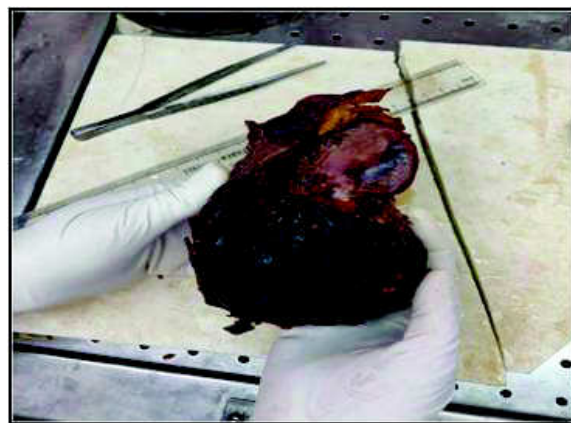
Case report

A 60 year old hypertensive female presented to our emergency sonography room with acute surgical reference of left sided abdominal pain & palpable lump for last 10 days. She had no complained of fever, vomiting or hematuria. Her pulse rate was 86 per minute & BP was 138/92.

Ultrasound was performed, on which a fat echogenicity lesion in the lower pole of left kidney with hypoechoic fluid collection in left perinephric space extending along left upper psoas was found. Rest of the kidney demonstrated normal vascularity. Rest of the abdomen ultrasound was unremarkable. Diagnosis of angiomyolipoma with retroperitoneal bleed was made.

To demonstrate arterial supply & any active bleeding point if present, triple phase CT abdomen pelvis was advised. The CBC, RFT & LFT were within normal limits. Triple phase CT abdomen pelvis was done, on which approximately 8.9x9 cm sized fat density lesion containing approximately 4x4.5 cm sized intralesional hematoma was found with approximately 4.5x6x11 cm sized perinephric hematoma.

The lesion appeared to be supplied by upper segmental branch of anterior branch of left renal artery. There was no active bleeding spot. There was small angiomyolipoma in right kidney as well. Rest of the scan was unremarkable. Diagnosis of



Wunderlich syndrome was made. The patient underwent planned nephrectomy in our hospital considering the size of lesion & risk of rebleeding.

Image 4: Gross specimen of left kidney showing lower pole hematoma

During surgery considerable sized angiomyolipoma with large perinephric hematoma was found. Later histopathology confirmed the diagnosis of angiomyolipoma.

Discussion

Carl Wunderlich in 1856 first, described spontaneous perinephric subcapsular hematoma without known underlying etiology. Pathologies like renal neoplasm - benign or malignant, renal vascular disease like vasculitis, AVM, arteriosclerosis, renal arterial aneurysm rupture, infectious disease like nephritis can cause non traumatic perirenal retroperitoneal haemorrhage, among which neoplastic etiology - benign neoplasm angiomyolipoma in sporadic form is most common.^[1,2]

Now they come under the category of PEComas - perivascular epitheloid cell tumors. In syndromic association with tuberous sclerosis they

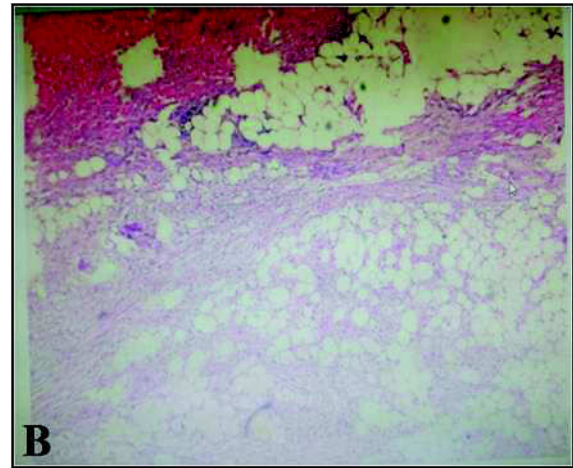
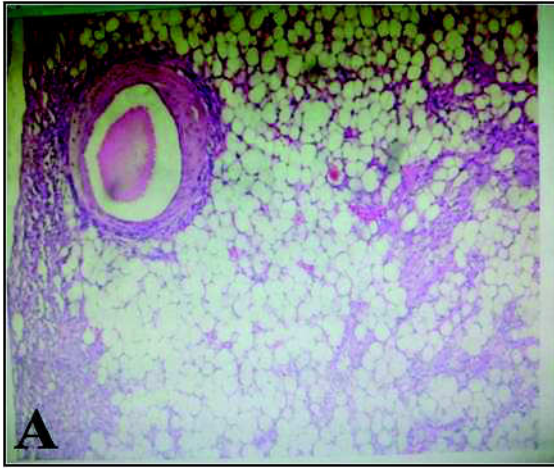


Image 5: Microscopic slide (H & E Stain - 40 X) **A.** showing arteriole & fat cells. **B.** showing fat cells & muscle cells

often present as bilateral, multiple small lesions.

Risk of bleeding depends on the size. Several studies have demonstrated that up to 76% lesions less than 4 cm in size remain asymptomatic while up to 92% lesions more than 8 cm size cause symptoms, classic being Lank's triad.^[3,4]

CT scan is the modality of choice to characterize the nature of lesion & its complication as it clearly depicts intralesional fat density & perirenal hematoma when there is retroperitoneal bleed. The asymptomatic patients can be monitored on follow up scan on annual bases (<4 cm) or semester bases (>4 cm).

The larger lesion can be classified into 3 categories according to vascularity.

- 1) Minimal vascular lesion containing few small stretched vessels.
- 2) Moderately vascular lesions containing medium sized dilated tortuous vessels with few or none smaller (<4 mm) aneurysms
- 3) Markedly vascular lesions containing numerous large dilated tortuous vessels with few or none large aneurysm (> 5 mm).^[4,5]

The risk of bleeding increases during pregnancy mainly due to increase in blood volume, renal blood flow & hormonal changes. The larger lesions with minimal vascularity rarely undergo spontaneous rupture. Lesions more than 8 cm are likely to be symptomatic.^[5,6]

They can be managed with minimal invasive procedure like selective trans-arterial embolization at first line. Nephrectomy, partial or radical can be done in hemodynamically unstable patients, those with failed embolization or suspected

malignancy.^[8,9,10]

Conclusion

A case of Wunderlich syndrome with massive retroperitoneal bleed is reported. Wunderlich syndrome being the most fearful complication of angiomyolipoma which can turn fatal if not correctly diagnosed & managed. CT scan being the modality of choice, can appropriately diagnose Wunderlich syndrome & its underlying pathology & also helps in planning selective trans-renal arterial embolization & in avoiding unnecessary laparotomy.

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