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Case Report

WUNDERLICH SYNDROME: A CASE REPORT

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ABSTRACT
Spontaneous Renal Hemorrhage (SRH) also known as Wunderlich syndrome is a rare entity but an important clinical problem. The most common underlying conditions includes neoplasms, vascular disease, cystic diseases, long term haemodialysis therapy and anticoagulant therapy. In this case report, we discuss a 65-year-old male, known case of chronic kidney disease on haemodialysis who presented with acute right upper quadrant pain and tenderness. The patient was also a known case of bilateral renal cystic disease. On ultrasound performed, the right kidney was bulky and was seen to be replaced by a large heterogeneous mass with presence of an adjacent retroperitoneal hematoma. These findings were confirmed on computed tomography(CT). The patient was treated conservatively with resolution of symptoms. A follow up CT performed after 3 weeks revealed partial resolution of the renal hematoma and complete resolution of the retroperitoneal hematoma. SRH is a rare but potentially life threatening condition which requires a high index of suspicion and has to be promptly diagnosed and treated in emergency.

Keywords: *Spontaneous Renal Hemorrhage, Renal Hematoma, Wunderlich Syndrome*

INTRODUCTION
SRH is a diagnostic dilemma and a rare condition in clinical practice. The presentation of clinical signs and symptoms depend on the degree and duration of bleeding and may therefore vary significantly. Neoplasms account for the majority of cases, of which angiomyolipoma and renal cell carcinoma are the most common benign and malignant causes respectively (Belville *et al.,* 1989).

CASE
A 65-year-old male presented in the emergency department with acute onset of right upper quadrant pain. There was no history of trauma, fever or haematuria. The patient was a known case of chronic kidney disease (Stage-V) on maintenance haemodialysis, hypertensive on anti-hypertensive medication, and ischemic heart disease on antiplatelet medication and was last dialyzed 12 days back with heparin dialysis (6000 IU). On physical examination, vitals were stable and a per abdomen examination revealed tenderness in the right flank region, with no organomegaly. Laboratory examination revealed haemoglobin- 7.8 g/dl, blood urea – 66mg/dl and serum creatinine was 6.56 mg/dl. Coagulation profile, total blood counts and urine analysis were within normal limits. X-ray Abdomen did not detect any abnormality. On abdominal ultrasound performed, the right kidney was bulky (11 x 8.3 cm) and replaced by a large heterogeneous mass, predominantly hypoechoic with surrounding inflammatory changes and perinephric fluid. Ultrasound also showed presence of a retroperitoneal hematoma with fluid in the adjacent retroperitoneum. Colour doppler ultrasound did not reveal any vascularity within the mass. The left kidney was shrunken in size (6 x 4 cm) and showed presence of renal cortical cysts. Rest of the abdominal organs were within normal limits. A Non-Contrast Computed Tomography (NCCT) performed subsequently revealed the right kidney was bulky (measuring 11.1 x 8.3 cm) and replaced by a large hyperdense mass having average Hounsfield unit of +60 to +70 HU, suggestive of a subcapsular/intraparenchymal renal hematoma. Few calcific foci as well as cysts were noted in the right kidney. Surrounding fluid and perinephric fat stranding was seen, extending along the anterior and posterior para-renal spaces on the right side. There was presence of an adjacent retroperitoneal hematoma measuring approximately 2.1 x 3.6 x 7.3 cm (AP x TR x SI) in size. The left kidney measured 6.1 x 4.1

cm in size and showed cystic changes and dystrophic parenchymal calcifications. The ureters and the urinary bladder were normal.

The patient was admitted and treated conservatively with bedrest, saline dialysis, antibiotics (Meropenem and Trezam/Aztreonam) and antifibrinolyitic agents (Trenexamic acid). There was complete resolution of symptoms in one week after which he was discharged. A follow up CT performed after 3 weeks revealed partial resolution of the renal hematoma and complete resolution of the retroperitoneal hematoma.

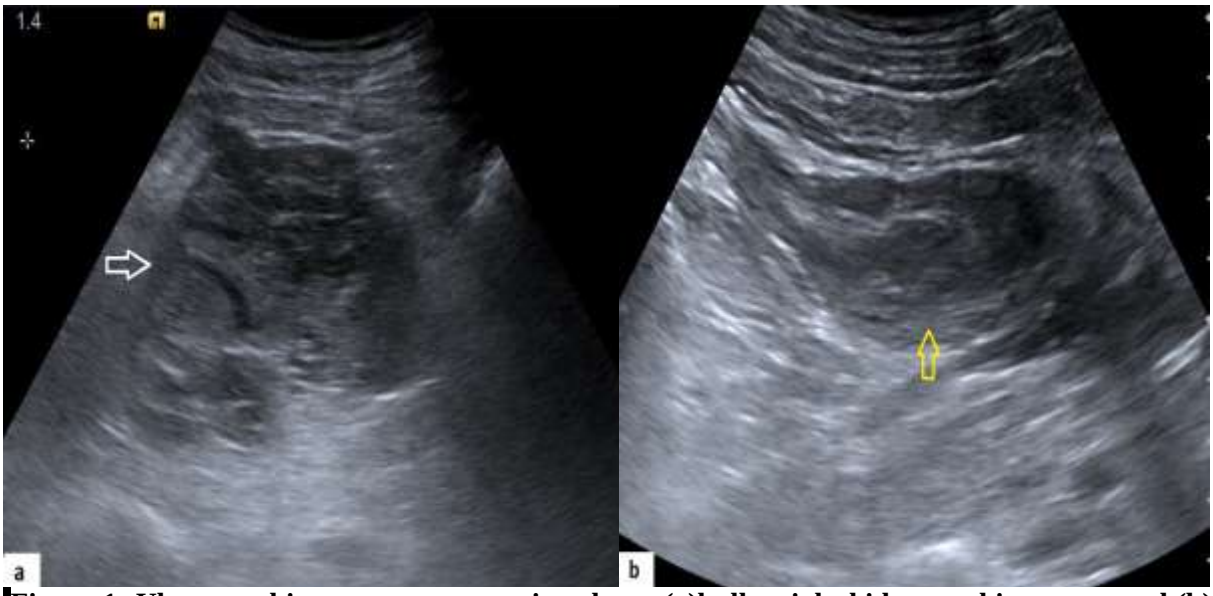

Figure 1: Ultrasound images at presentation shows (a)bulky right kidney –white arrow and (b)the right sided retroperitoneal hematoma- yellow arrow.

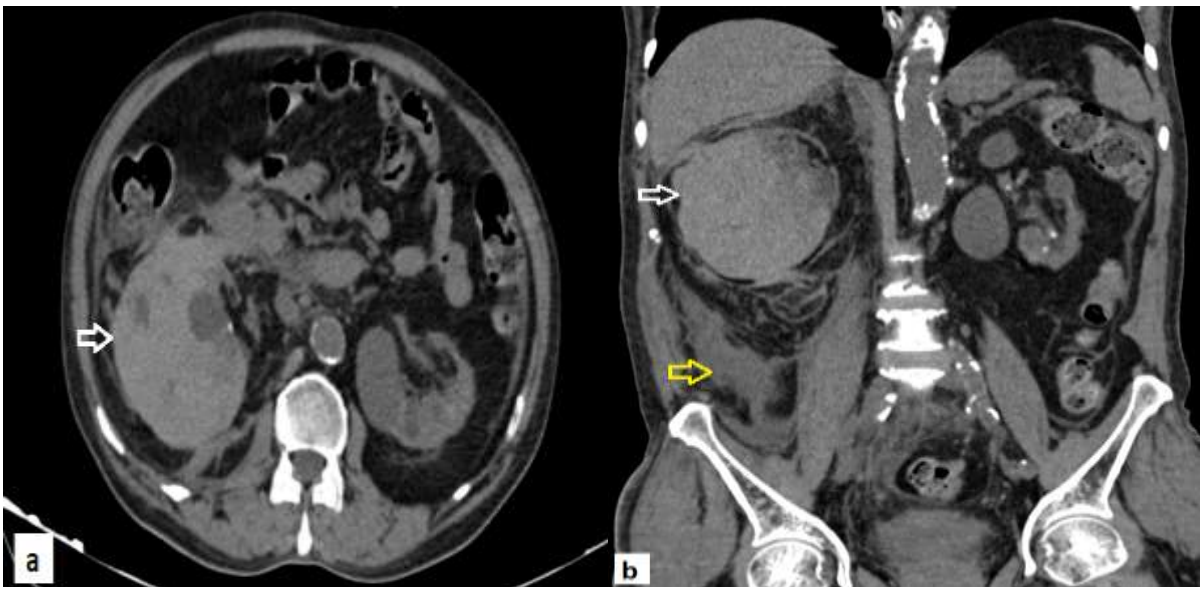

Figure 2: Computed tomography images at presentation shows the right sided subcapsular/ intraparenchymal renal hematoma (white arrows in a and b). Yellow arrow in figure (b) indicates the adjacent right sided retroperitoneal hematoma.

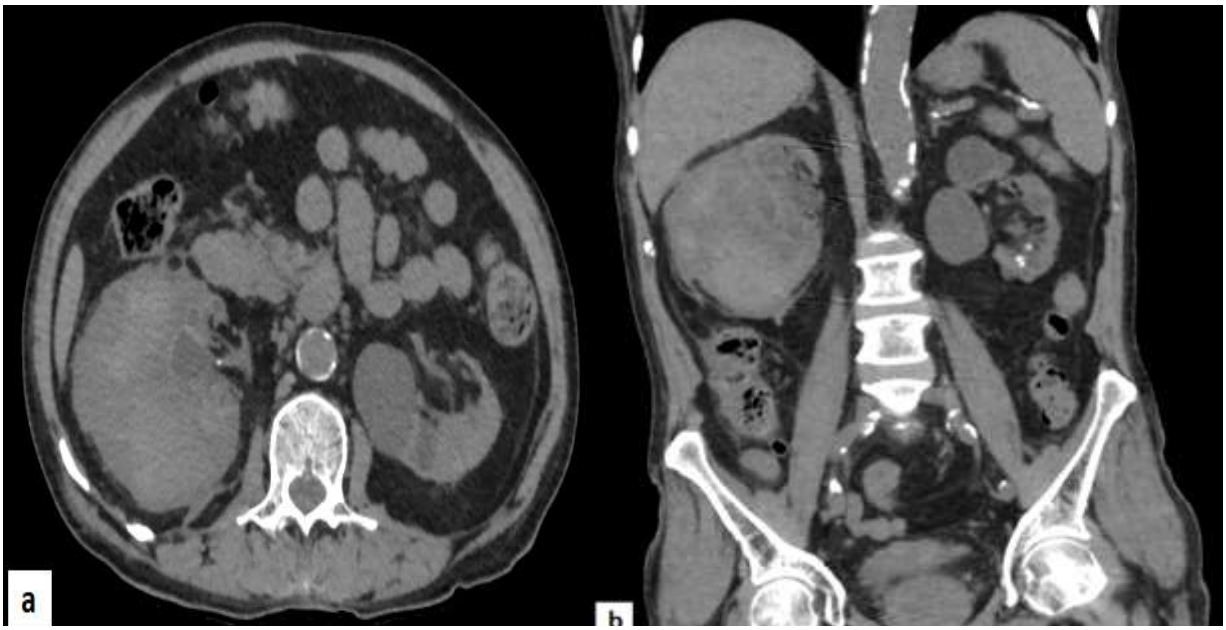

Figure 3: Follow-up computed tomography at 3 weeks revealed partial resolution of the right renal hematoma and complete resolution of the retroperitoneal hematoma.

DISCUSSION

Spontaneous Renal Hemorrhage, also known as Wunderlich syndrome, is a rare but important clinical problem that heralds a variety of renal abnormalities. This syndrome is characterised clinically by a triad of acute flank pain, flank mass and hypovolemic shock referred to as the Lenk's triad (Baishya et al., 2011). Neoplasms account for the majority of cases of SRH, of which angiomyolipoma and renal cell carcinoma are the most common benign and malignant causes respectively. Non-neoplastic causes of spontaneous renal hemorrhage include vascular causes like polyarteritis nodosa, renal artery aneurysms and arteriovenous malformations. Non-vascular causes include renal cysts, anti-coagulation therapy and coagulation disorders (Belville *et al.*,1989).

Anticoagulation with heparin sodium is a routine procedure in patients of chronic kidney disease as in this case. Other agents like aspirin could also be a predisposing factor for Spontaneous Renal Hemorrhage. Several authors have described cystic transformation of kidneys in patients with chronic kidney disease and hemorrhage can be a frequent complication (Pak *et al.*, 1986).

Ultrasonography is the first line imaging modality for identifying the condition while computed tomography is used for confirmation (Tonolini *et al.*, 2015). Computed tomography has higher sensitivity and specificity compared to ultrasound and provides useful information regarding the cause of the hematoma. Identification of fatty attenuation within a mass suggest the diagnosis of angiomyolipoma. MRI can also be used as an alternative to computed tomography while angiography can be used to rule out vascular causes of renal hemorrhage (Kendall *et al.*, 1988).

Conservative approach is adopted for hemodynamically stable patients with SRH. In hemodynamically unstable patients, nephrectomy (partial or total) may be required. In patients with signs of active hemorrhage, angiographic selective embolization may be pursued (Brkovic *et al.*, 1996).

CONCLUSION

SRH is a rare but potentially life threatening condition with a variety of predisposition conditions. It requires prompt diagnosis and treatment with computed tomography playing a crucial role due to its high sensitivity and specificity in diagnosing the condition.

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