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Wunderlich Syndrome – Minimally Invasive Treatment

Piyoosh Priyadarshee[1], Baskar A[*2], G. Murugan[3], Venkatraman Indiran[4], Ashwinkumar A.[5]

[1] MBBS, Post-graduate (MD) student, Department of Radiodiagnosis, Sree Balaji Medical College & Hospital, 7 Works Road, Chromepet, Chennai, Tamilnadu (600044), India

Email ID: piyoosh528@gmail.com

ORCID: 0009-0006-3352-4873

[*2]MBBS, MD Radio-diagnosis, DNB, MICR, Assistant Professor, Department of Radiodiagnosis, Sree Balaji Medical College & Hospital, 7 Works Road, Chromepet, Chennai, Tamilnadu (600044), India

[3]MBBS, DMRD, MD Radio-diagnosis, Head of the Department, Department of Radiodiagnosis, Sree Balaji Medical College & Hospital, 7 Works Road, Chromepet, Chennai, Tamilnadu (600044), India

E-Mail: dr.gmurugan@yahoo.com

[4]MBBS, MD Radio-diagnosis, DNB, Professor, Department of Radiodiagnosis, Sree Balaji Medical College & Hospital, 7 Works Road, Chromepet, Chennai, Tamilnadu (600044), India

Email ID: ivraman31@gmail.com

[5]MBBS, MD Radio-diagnosis, DNB, MICR, Associate Professor, Department of Radiodiagnosis, Sree Balaji Medical College & Hospital, 7 Works Road, Chromepet, Chennai, Tamilnadu (600044), India

Email ID: ashgilli@gmail.com

*** Corresponding Author**

Baskar A

Email ID: baskarmdrd@gmail.com

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ABSTRACT

Wunderlich Syndrome (WS), first described in 1856, is a rare, potentially life-threatening condition caused by spontaneous retroperitoneal hemorrhage, often from renal angiomyolipoma (AML). Symptoms include sudden flank pain, a palpable renal mass, and hypotension, making early diagnosis challenging. WS is most commonly caused by benign renal AMLs and malignant renal cell carcinoma. Treatment depends on the underlying cause, hemorrhage severity, and patient's hemodynamic status, ranging from conservative care to more invasive options like embolization and surgery.

This case report discusses a 60-year-old woman who presented with acute right flank pain, pallor, and dizziness. Imaging revealed a large AML in the right kidney with significant hemorrhage. The patient underwent successful emergency renal artery embolization (RAE), which stabilized her condition and stopped the bleeding. Post-procedure imaging confirmed the efficacy of RAE, emphasizing its role in managing WS, offering renal preservation with fewer complications compared to nephrectomy.

Keywords: *Wunderlich Syndrome, Renal, Angiomyolipoma, Renal Artery Embolization*

1. INTRODUCTION

Wunderlich syndrome (WS), first described in 1856 by Carl Wunderlich, also known as spontaneous retroperitoneal hemorrhage, is a rare but potentially life-threatening condition characterized by sudden, spontaneous bleeding into the subcapsular and perirenal spaces [1].

Although the condition is uncommon, it can lead to significant morbidity due to the difficulty in early diagnosis. It often occurs without any obvious abdominal trauma. The typical presentation includes sudden, severe flank pain (seen in ~ 53% of the cases), palpable renal mass (~47%) and hypotension with signs of hypovolemic shock due to massive blood loss (Lenk's Triad) [2]. Other less common symptoms may include nausea, vomiting, anemia and renal failure [3]. A key challenge in the diagnosis is differentiating it from other causes of acute abdominal pain, making a high degree of clinical suspicion essential.

The etiology of Wunderlich syndrome is diverse, ranging from benign to malignant causes – with the most common benign cause being renal angiomyolipoma and malignant cause being renal cell carcinoma [4, 5]. These two make up around 60% of the cases of WS [1].

Management of WS depends on the underlying cause, the severity of the hemorrhage, and the patient's clinical status. Conservative treatment, embolization, and surgical interventions may all be considered, with early diagnosis being critical to the patient's outcome.

2. CASE REPORT

A 60-year-old female patient presented to the emergency department with acute onset right flank pain. There was a palpable right flank mass. The patient also complained of sudden onset of fatigue associated with dizziness on standing. Her pulse rate was 100/minute and BP was 120/80mmHg. Her serum creatinine was 0.9 mg/dL and urea was 32 mg/dL.

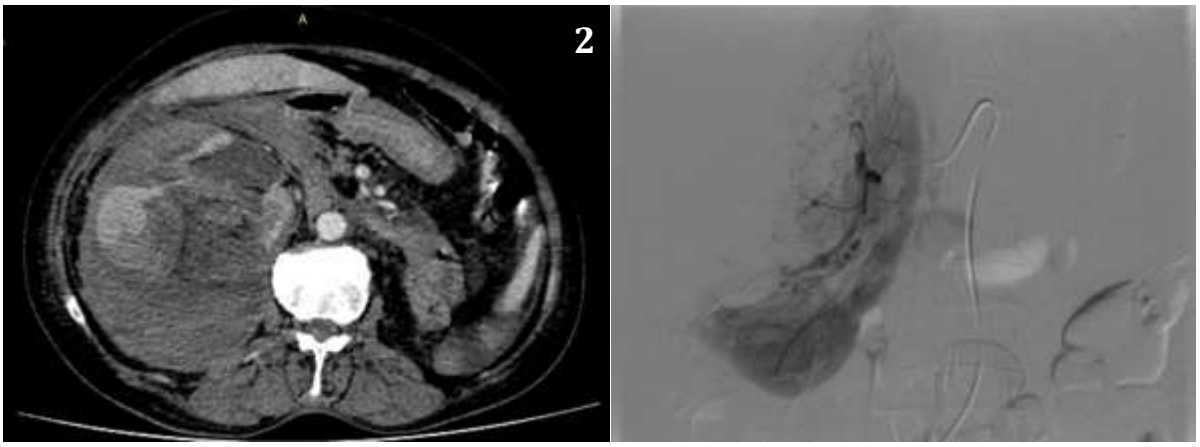

Fig.1: Axial view of contrast enhanced computerized tomography (CECT) image of the abdomen, showing perirenal haemorrhage and active contrast extravasation from upper pole segmental arterial branch indicating active bleeding.

Fig.2: Anteroposterior view of digital subtraction angiogram (DSA) of the right renal artery, showing mildly tortuous and branching arterial vasculature with numerous tiny aneurysms and non-opacification in the upper pole of the kidney.

CT scan showed a large heterodense lesion with internal fat densities arising from the upper and interpolar regions of the right kidney, resulting in loss of normal renal contour. The lesion displaced the pancreas and the second part of the duodenum anteromedially. On contrast enhanced study, there was a significant perirenal haemorrhage, with active contrast extravasation likely originating from a segmental arterial branch of the upper pole, indicating ongoing bleeding.

The patient was diagnosed with right renal angiomyolipoma with perilesional hematoma and was transfused with packed red blood cells. Emergency embolization was performed to reduce blood loss and stabilize the patient.

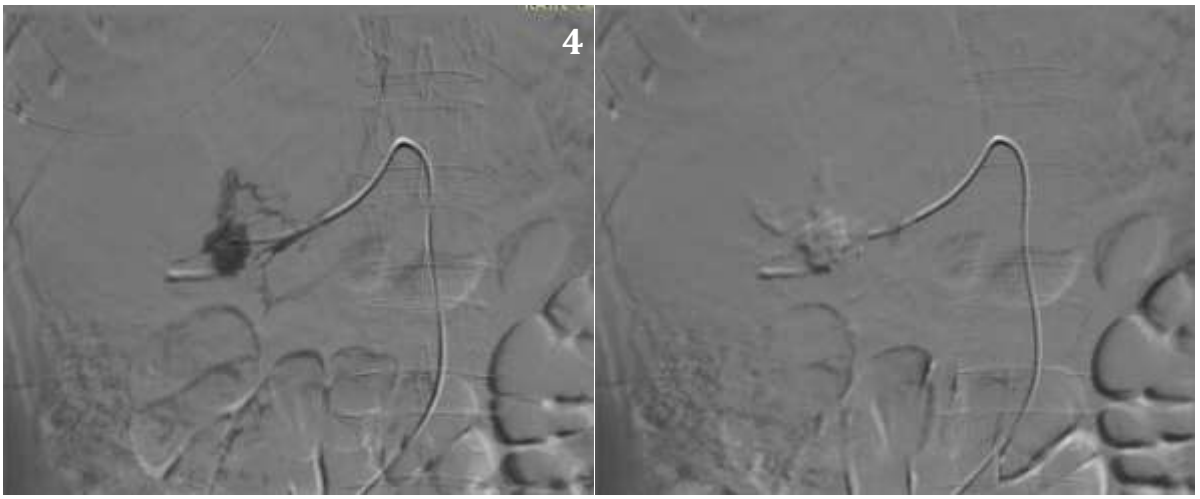

Fig.3: DSA image of right renal artery before embolization, showing leakage of contrast.
Fig.4: Post-embolization DSA image of right renal artery showing successful cessation of bleeding.

3. DISCUSSION

WS typically results from spontaneous bleeding into the retroperitoneal space, often due to renal AML, as in this case. AML is a benign mixed mesenchymal tumour which consists of blood vessels, smooth muscle and mature adipose tissue in different proportions [3]. Renal AMLs are classified into fat-rich (HU -10 or less on CT), fat-invisible and fat-poor [6].

Given its potential for rapid deterioration, the management of WS is focused on immediate hemorrhage control, patient stabilization, and renal preservation. Treatment decisions are guided by the severity of the hemorrhage, the patient's hemodynamic stability and the underlying etiology.

In patients presenting with active bleeding, as observed in this case, renal artery embolization (RAE) is the preferred first-line intervention [7]. This minimally invasive procedure allows for targeted embolization of the arteries supplying the hemorrhagic renal mass, thus controlling the bleeding without the need for open surgery. The advantage of RAE lies in its ability to rapidly stabilize the patient, arrest active hemorrhage, and preserve renal function. The pre- and post-embolization images in this case demonstrate successful cessation of bleeding. Studies show that RAE not only provides immediate hemostasis but also carries a lower complication rate compared to nephrectomy, with renal preservation being an additional benefit [7]. It is particularly effective in patients with hemodynamically unstable conditions, as it reduces the need for transfusions and other intensive measures.

Nephrectomy is considered in cases where embolization fails or when there is a significant risk of continued or recurrent hemorrhage, as in cases with large or ruptured AMLs. However, nephrectomy is associated with higher morbidity, including the loss of renal function, and is reserved for more severe or refractory cases.

Most of the small AMLs are asymptomatic and incidental finding on radiological studies performed for unrelated conditions [8]. In asymptomatic cases or those with smaller tumors (<4 cm), conservative management and regular monitoring are often sufficient. However, tumors greater than 4 cms in diameter have a higher risk of rupture and aneurysm formation [3, 9, 10], require more aggressive surveillance, and interventions like embolization or surgical resection may be considered even in the absence of active symptoms [7].

Overall, Wunderlich syndrome represents a urological emergency that necessitates a multi-faceted approach. Selective renal artery embolization stands out as the cornerstone of treatment for active bleeding, particularly in AML-related cases [11], while surgery is reserved for cases where embolization is not successful or when malignancy is suspected. The successful management of this patient through embolization emphasizes the critical role of timely, targeted treatment in improving patient outcomes and reducing the risk of long-term complications.

4. CONCLUSION

Wunderlich syndrome (WS) remains a rare yet critical condition due to its potential for rapid deterioration. As illustrated in the case report, WS often presents with nonspecific symptoms like flank pain, pallor, and signs of hypovolemic shock, complicating early identification. The etiology is most frequently benign, with renal angiomyolipomas (AMLs). The main treatment for active bleeding is renal artery embolization (RAE), which provides effective hemorrhage control and preserves renal function. Nephrectomy remains a last resort for refractory or severe cases; RAE offers a less invasive alternative with fewer complications.

5. ABBREVIATIONS

WS	Wunderlich Syndrome
AML	Angiomyolipoma
CT	Computerized Tomography
CECT	Contrast Enhanced Computerized Tomography
DSA	Digital Subtraction Angiogram
RAE	Renal Artery Embolization

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Informed consent:

Informed consent was obtained from the participant included in the study. No personal identifying information is included in this article.

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