



CASUISTIC PAPER

## Wunderlich syndrome – report of a rare case with comments on clinical implications

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### ABSTRACT

**Introduction and aim.** Spontaneous renal hemorrhage, known as Wunderlich syndrome, is a rare clinical condition that occurs without any history of trauma. The most common causes of this syndrome are both benign and malignant renal tumors. The treatment strategy is determined based on the patient's hemodynamic stability.

**Description of the case.** We report a case where a patient was admitted to the emergency department experiencing persistent pain in the right flank for three days. A diagnosis of spontaneous renal hemorrhage, secondary to an angiomyolipoma, was established through CT imaging. In this case study, we detail the diagnostic process and management of a patient who, due to the absence of hemodynamic instability, did not require surgical intervention. Instead, the patient was monitored and managed with conservative treatment.

**Conclusion.** This case highlights the importance of prompt diagnosis, implementation of appropriate treatment, and the relevance of active follow-up in hemodynamically stable patients receiving conservative treatment.

**Keywords.** angiomyolipoma, flank pain, renal hematoma, spontaneous renal hemorrhage, Wunderlich syndrome

### Introduction

Wunderlich syndrome (WS), also known as spontaneous renal hemorrhage is a rare condition in which there is bleeding into the subcapsular and/or perirenal space without the presence of trauma as a causative factor.<sup>1</sup> The most common cause of WS is renal tumors, with both malignant and benign tumors serving as etiological factors, among which angiomyolipoma (AML) represent the most significant alteration.<sup>2</sup> Other causes of this rare condition include vasculitis,

arteriovenous malformations, and aneurysms.<sup>3</sup> WS is a severe condition which may require urgent nephrectomy.

### Aim

We report the case of a 46-year-old female patient who presented to the emergency department with acute abdominal pain mainly localized in the right flank. This work has been reported in line with the CARE (for CASE Reports) criteria.<sup>4</sup> This case enriches the existing data

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on rare kidney abnormalities and consolidates the information published on the topic to date.

### Description of the case

A 46-year-old female patient presented herself to emergency department, reporting a persistent right flank pain that had lasted for three days and radiated to the right lower quadrant. She had no history of hematuria, fever, or trauma. Vomiting occurred only on the first day of the reported pain. The patient denied any medical history. Upon her arrival, the cardiopulmonary examination was within normal limits.

**Table 1.** Relevant initial blood work results\*

Parameter	Value of our patient	Reference range
Leukocytes ( $10^3/\mu\text{L}$ )	8.5	4–10
Hemoglobin (g/dL)	12.4	12–16
Hematocrit (%)	36	38–45
Platelets ( $10^3/\mu\text{L}$ )	293	150–400
hsCRP (mg/dL)	2.64	0–0.5
APTT (s)	32.4	25.4–36.9
Prothrombin time (s)	13.3	9.4–12.5
Creatinine (mg/dL)	0.6	0.55–1.02
eGFR ( $\text{mL}/\text{min}/1.73\text{m}^2$ )	114	>60
Sodium (mmol/L)	137	136–145
Potassium (mmol/L)	4	3.5–5.2

\* hsCRP – high sensitivity C-reactive protein, APTT – activated partial thromboplastin time

**Table 2.** Relevant urinalysis results

Parameter	Value of our patient	Reference range
Dipstick urinalysis		
Color	yellow	–
Clarity	turbid	–
pH	6	5–7
Specific gravity (g/mL)	1.020	1.015–1.025
Glucose	negative	negative
Blood	positive	negative
Protein	positive	negative
Bilirubin	negative	negative
Ketones	negative	negative
Ascorbic acid	positive	negative
Nitrate	negative	negative
Urine microscopy		
White blood cells (cells/hpf)	many per hpf	2–5
Red blood cells (cells/hpf)	singular per hpf	2
Bacteria	many	negative
Squamous epithelial cells (cells/hpf)	few	15–20

The abdomen was soft with tenderness in the right side and pain radiating to the right flank. Costovertebral angle tenderness (Goldflam's sign) on the right side was questionable and on the left side was negative. Initial blood tests revealed elevated levels of high-sensitivity C-reactive protein. The hemoglobin level was found to be within the normal range (Table 1). Urinalysis showed

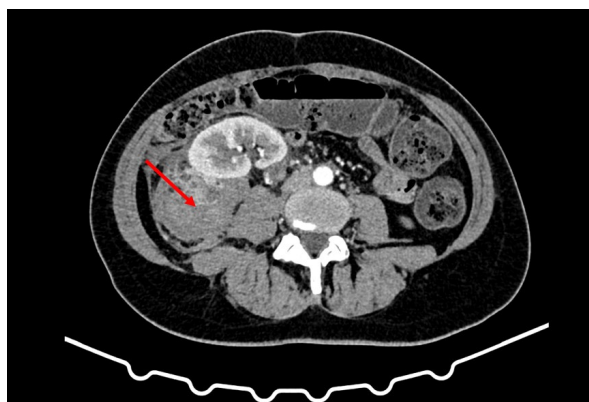
the presence of blood cells, leukocytes and numerous bacteria in the urine (Table 2).

An abdominal and pelvic computed tomography (CT) scan, urological and gynecological examinations were performed to evaluate the cause of the pain.

The CT image was performed in spiral acquisition with 1.5 mm layers in native phase and after contrast enhancement (multiphase). Both kidneys with the presence of numerous heterogeneous fat lesions about 4 cm in size on the right side and 6 cm on the left side – lesions indicative of an angiomyolipoma (AML) (Fig. 1, Fig. 2 and Fig. 3).



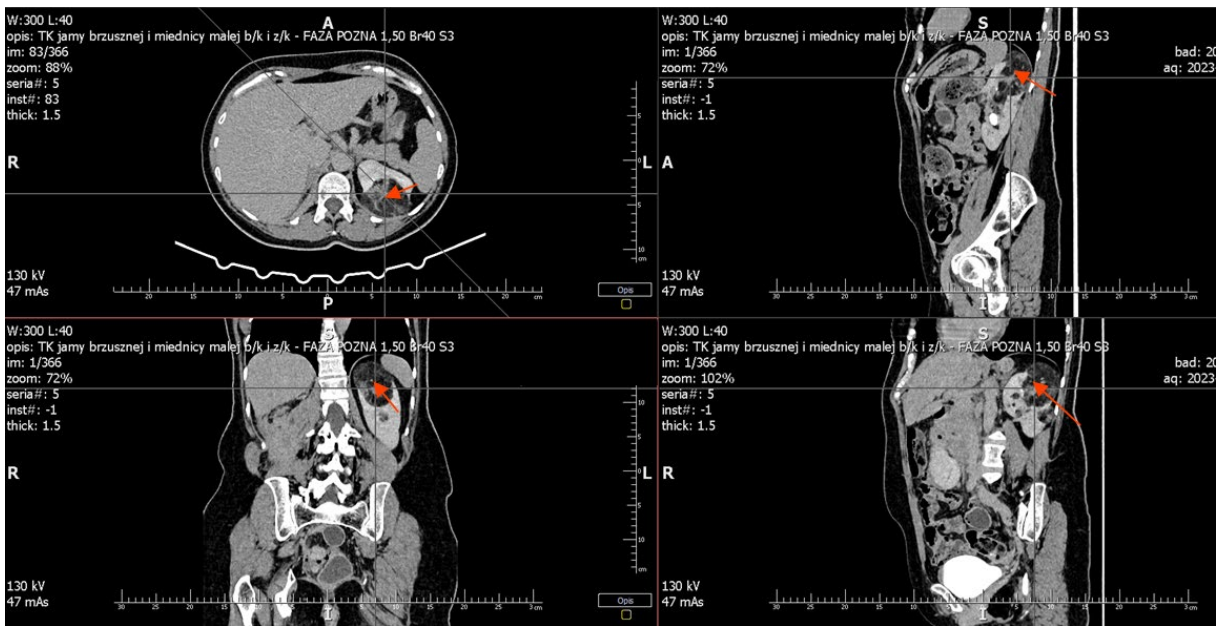
**Fig. 1.** The right kidney with numerous heterogeneous fat lesions up to 4 cm in size - AML-like lesions, right kidney visibly displaced forward by hematoma and AML-like lesions



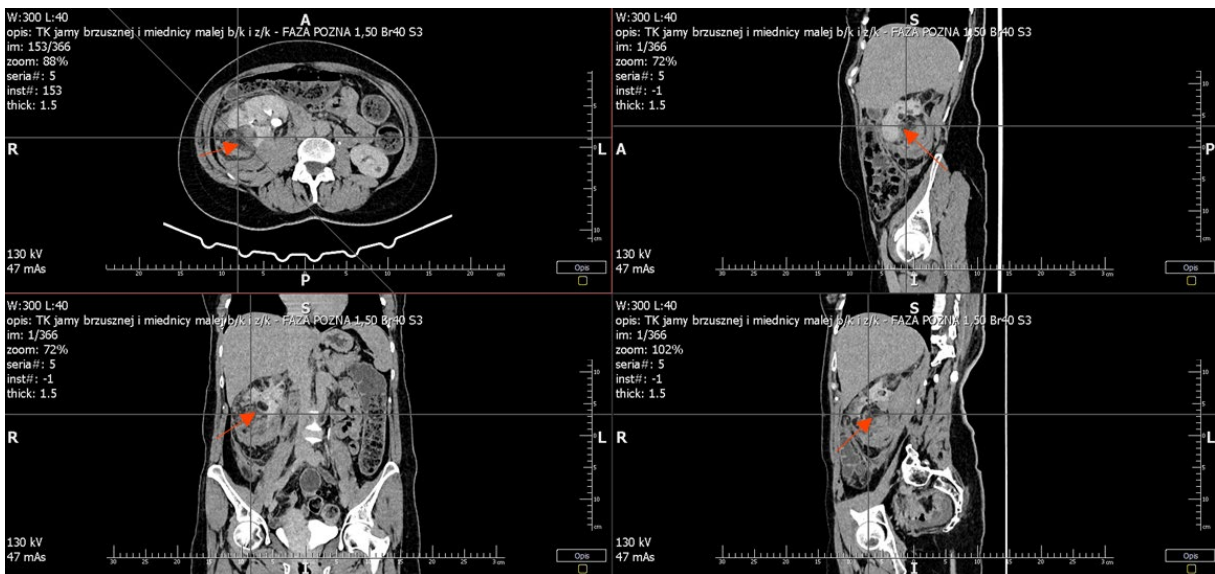
**Fig. 2.** Perinephric hematoma associated with one of the AMLs of this kidney

In the perinephric fatty space on the right side, mainly on the dorsal side, numerous hyper dense bands 2.5x8.3 cm wide, with a density of about 60 HU, not enhancing after contrast – a perinephric hematoma associated with one of the AMLs of this kidney was diagnosed (Fig. 4).

Right kidney markedly displaced anteriorly by hematoma and AML-like lesions. The kidneys secrete



**Fig. 3.** The left kidney with numerous heterogeneous fat lesions up to 6 cm in size – AML-like lesions



**Fig. 4.** In the perirenal fat space on the right side mainly on the dorsal side, numerous hyperdense bands up to 2.5x8.3 cm wide, with a density of about 60 UH, not enhancing after contrast – a perinephric hematoma associated with one of the AMLs of this kidney

urine bilaterally, without any signs of stasis or urine leakage. No calcified deposits were observed in the kidneys. Adrenal glands, ureters, urinary bladder were without pathology. Visible fluid in the sinus of Douglas on the right side was about 19 mm wide. No pathological changes were found in other abdominal organs, the imaged lymph nodes were not enlarged.

A diagnosis of Wunderlich syndrome associated with spontaneous rupture of an AML was made. The patient refused hospitalization. Due to the patient's refusal of further hospital treatment, she was discharged from the emergency department in accordance with the

doctor's recommendation. She was prescribed a strict bed rest, an antibiotic containing sulfamethoxazole + trimethoprim for a genitourinary infection. In addition, a follow-up visit to the general practitioner and the urology clinic was recommended. In the follow-up CT scan conducted 4 weeks after diagnosis, a regression of the hematoma was observed, the patient's clinical condition was assessed as good, and the patient reported no abdominal complaints. At the next follow-up visit, 8 weeks from diagnosis, further regression of the hematoma was observed in the control ultrasound of the abdominal cavity.

## Discussion

Spontaneous, nontraumatic renal bleeding limited to subcapsular and/or perirenal space, WS was described for the first time in 1856 by Carl Reinhold August Wunderlich.<sup>1</sup> Known causes of WS range from neoplasms such as AML, renal cell carcinoma, sarcomas, lymphomas or pheochromocytomas to non-neoplasm like acquired cystic kidney disease, simple and/or hemorrhagic renal cysts, infections (acute and chronic pyelonephritis, renal abscess, emphysematous pyelonephritis, nephritis), ureteropelvic junction obstruction (Kliknij tutaj, aby wprowadzić tekst. vascular diseases (vasculitis, renal artery arteriosclerosis, renal artery aneurysm rupture, polyarteritis nodosa and renal vein thrombosis), undiagnosed or new occurring (like microangiopathic hemolytic anemia) hematological disorders and anatomical lesions.<sup>5-10</sup> A case of WS caused by pancreatic pseudocyst has also been described.<sup>6</sup> The presence of multiple unilateral or bilateral AMLs has been linked to tuberous sclerosis (TS), making TS a potential risk factor for Wunderlich syndrome. Therefore, TS should be considered when diagnosing a patient with this condition. Among patients with suspected WS, factors preceding trauma, anticoagulant treatment, hemorrhagic diathesis, arteritis, tuberous sclerosis and chronic hemodialysis should be excluded.<sup>12</sup> In addition, a rare case of Wunderlich Syndrome was described, which was diagnosed by prenatal ultrasound and manifested bilateral hydronephrosis and fetal bladder dysfunction.<sup>13</sup> However, the most common cause of Wunderlich syndrome is rupture of tumor, with AML accounting for the majority.<sup>5-7</sup> The second leading cause of WS syndrome is diseases of vascular etiology, such as polyarteritis nodosa (PAN), with smaller contributions from aneurysms, arteriovenous malformations, renal vein thrombosis and myocardial infarction accounting for about 20–30%.<sup>2,14</sup> Among patients without previously diagnosed tuberous sclerosis, PAN should be considered if bilateral or recurrent WS is present.<sup>2</sup> Sometimes the cause of the syndrome cannot be determined.<sup>5,15</sup>

Clinical presentation of patients with WS varies depending on the extent, duration and cause of bleeding. The most commonly reported symptom is unilateral flank pain that may radiate medially or downwards,<sup>5,15</sup> which may be accompanied by hematuria, vomiting, weakness, fever, renal failure, anemia. Hemorrhagic shock may occur in 11–26.5% of patients. The classic presentation (Lenk's triad) includes flank or abdominal pain, palpable tender mass and hemorrhagic shock but only 20% of patients present those three together.<sup>5,16</sup> Preliminary diagnosis of WS can be based on ultrasound, however a method with 92–100% sensitivity is contrast-enhanced CT, furthermore, performed during the time of hemorrhage enables us to identify all WS caused by AML.<sup>5,10,15</sup> Combining CT with MRI enables detection of approximately 80% WS causes additionally providing 100% sen-

sitivity. In other cases, selective renal angiography may be useful in determining the source of bleeding.<sup>7</sup>

Management of patients with spontaneous, non-traumatic renal bleeding will be determined by the cause and hemodynamic stability of patients. Currently, patients are eligible for initial treatment by three methods i.e. conservative/medical management involving only hemodynamic stabilization using fluids and blood products with monitoring of the patient's clinical condition; transarterial embolization, which is the most common treatment of choice and surgery, which is generally based on performing a total nephrectomy, but can also involve exploration/wash out, ureteric stenting or partial nephrectomy.<sup>17</sup> In hemodynamically stable patients, observation and conservative treatment can be used along with regular follow-up visits.<sup>18</sup> Patients who presented with hemorrhagic shock in the clinical picture are more likely to require surgical management, but even in patients in severe hypovolemic shock, improvement can be achieved with conservative/medical management alone.<sup>8,9,15,17</sup> Initial surgical treatment is also used in patients with suspected malignant lesions.<sup>9,17</sup> Embolization, on the other hand, is the treatment of choice more often when patients cannot be stabilized with conservative management or instead of it, and when the causes are AML or vascular lesions.<sup>10,16,17</sup>

However, there are no rigidly established management protocols, and the final choice of management will depend on the patient's condition, the technical capabilities of the staff and their experience.

## Conclusion

Wunderlich syndrome is a rare but important condition to take into consideration when diagnosing a patient with sudden flank pain and hematuria. If not recognized quickly enough, it can lead to hemodynamic instability which is a life-threatening condition. Consequently, in patients with spontaneous non-traumatic renal hemorrhage, it is important to perform a contrast-enhanced CT scan without delay and implement proper conservative or surgical treatment, depending on the patient's hemodynamic stability. Our case highlights the importance of timely diagnosis and active follow-up in hemodynamically stable patients treated conservatively.

## Declarations

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### Author contributions

Conceptualization, K.K and K.B.; Methodology, K.K. and A.K.; Software, S.R.; Resources, S.R. and K.B.; Data Curation, S.R. and A.R.; Writing – Original Draft Preparation, K.K. and A.K.; Writing – Review & Editing, S.R. and A.R. and K.K.; Visualization, K.B.; Supervision, K.B.

**Conflicts of interest**

The authors declare that there are no conflicts of interest regarding the publication of this article.

**Data availability**

Not applicable.

**Ethics approval**

Written informed consent for publication was obtained from the patient. We complied with the policy of the journal on ethical consent.

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