

A Rare Diagnosis of Flank Pain in an Emergency Department as Wunderlich Syndrome

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Abstract

Wunderlich syndrome (WS) is an uncommon cause of flank pain wherein spontaneous bleeding occurs into the subcapsular and perirenal spaces of the kidney. It is most commonly attributed to renal tumors, especially angiomyolipomas. We report a case of a patient who presented to an emergency department (ED) with flank pain and symptoms of hemodynamic instability; radiologic imaging led to the diagnosis of WS. The patient's condition was managed conservatively first; then, selective embolization was performed. For patients who present to ED with flank pain and negative hematuria, an important diagnosis that can be fatal, if not recognized early, is WS. It is classically presented with acute flank pain, flank mass, and hypovolemic shock, together known as Lenk's triad. It can be fatal if not recognized and managed early, and radiological confirmation is required for accurate diagnosis. Therefore, physicians in ED should keep this syndrome in mind to prevent misdiagnosis (for e.g., as renal colic pain); and thereby the mortality.

Keywords: Wunderlich, flank pain, renal hemorrhage

Introduction

Wunderlich syndrome (WS) is a very uncommon (frequency not known) cause of flank pain wherein spontaneous bleeding occurs into the subcapsular and perirenal spaces of the kidney (1). It is most commonly attributed to renal tumors, especially angiomyolipomas (AMLs) (1). If not recognized and treated aggressively, it can be fatal (2).

We report a case of a patient who presented to an emergency department (ED) with flank pain and symptoms of hemodynamic instability and was soon diagnosed with WS.

Case Presentation

A 73-year-old female presented with sudden right sided flank pain. She had no complaints of fever or hematuria, and there was no history of trauma or previous urolithiasis. She had hypertension and diabetes and was on medications for the same. On physical examination, she was pale and had tenderness over the right flank.

She had a blood pressure of 97/58 mmHg, pulse rate of 93 beats/min, respiratory rate of 20 breaths/min, and body temperature of 36°C. A complete blood count was performed, and the results revealed hemoglobin of 10.7 g/dL, platelet count of 284000/ μ L, and white blood cell count of 19200/ μ L. Her renal function tests were normal with a creatinine level of 0.84 mg/dL.

Abdomen Ultrasonography (US) of the abdomen revealed a generally hyperechoic lesion with local hypoechoic areas measuring 6.6x9.9 cm in the upper pole of the right kidney (AML). There was associated heterogeneously hypo-anechoic collection measuring 2.2 cm in depth around the middle and lower parts of the right kidney, suggestive of retroperitoneal hemorrhage.

Based on the results of US, an urgent contrast-enhanced computed tomography (CT) scan was performed for further examination (Figure 1). It showed a mass lesion arising from posterior of middle and lower pole of right kidney, measuring 7.9x7.1 cm with few hypodense areas of fat attenuation, confirming it as AML. There was a retroperitoneal

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Figure 1. Contrast-enhanced abdominal CT showed a mass lesion arising from posterior of middle and lower pole of right kidney, measuring 7.9x7.1 cm with few hypodense areas of fat attenuation, confirming it as angiomyolipoma



Figure 2. Contrast-enhanced abdominal computed tomography demonstrated a retroperitoneal hematoma

hematoma 9.5 cm in depth secondary to AML, and it displaced the kidney anteriorly (Figure 2).

The patient was consulted with urologist and was admitted. The patient was managed conservatively under active monitoring for 10 hours, and then, selective embolization was performed. Partial nephrectomy, with the intention of enucleating the AML and removing of the hematoma was decided as the definitive treatment.

Written informed consent was obtained from the patient who participated in this study.

Discussion

Wunderlich syndrome is a rare condition of acute spontaneous renal hemorrhage into the subcapsular and perirenal spaces. This condition

was first described by a German physician called C.R. Wunderlich in 1856 (1). Patients classically presented with acute flank pain, flank mass, and hypovolemic shock, collectively known as Lenk's triad (2). According to a meta-analysis, frequency of these symptoms were 83%, 19%, and 11% for acute onset of flank pain, hematuria, and signs and symptoms of hypovolemic shock, respectively (3).

The most common etiological factor of WS is renal neoplasms (61.5%), with a predominance of AML (29.1%) and renal cell carcinoma (26.1%) (3). AMLs are benign tumors originating from perivascular epithelioid cells and composed of abnormal blood vessels, muscle cells, and adipose tissue (4). Nearly 10% of patients with AML presenting with WS, and the risk of WS is increasing especially if the mass is larger than 4 cm in diameter (1, 4). Moreover, other underlying conditions include arteriovenous malformations, vasculitis (polyarteritis nodosa), cystic renal diseases, infections, nephritis, and renal calculi (2, 5).

Patient with mild symptoms of WS can be misdiagnosed as having renal colic pain; however, 33% of patients with AML bleeding can develop hypovolemic shock. Therefore, imaging is necessary (5, 6). US is usually the initial choice of imaging for rapid evaluation, but sometimes, retroperitoneal hemorrhages may be misdiagnosed as renal tumor or abscess using US; CT is 100% sensitive for diagnosis and much more valuable for demonstrating the underlying etiology of hematoma (2, 5).

Management of WS is based on the hemodynamic status of the patient and the underlying etiology. Because the most common cause is malignancy, in past, urologists recommended expletory laparotomy or nephrectomy in most cases (5). However, beside the improvement of the life-threatening condition, the secondary goal of treatment is trying to preserve the kidney. For asymptomatic AML, minimal invasive techniques, such as selective renal arterial embolization or radiofrequency ablation, can be treatment options (6). These techniques can also be used preoperatively to avoid excess blood loss during surgery. Another treatment option; which was the treatment modality that chosen for our patient, is laparoscopic partial nephrectomy and this technique also preserves the kidney (1).

Conclusion

For patients who present to ED with flank pain and negative hematuria, an important diagnosis that can be fatal, if not recognized early, is WS. It refers to spontaneous renal hemorrhage into the subcapsular and perirenal spaces, and in 1/10th of the patients hypovolemic shock can be observed. It is difficult to diagnose clinically and can be a life-threatening condition if not recognized and managed early; radiological confirmation is required in almost all cases.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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