CASE REPORT

Wunderlich syndrome secondary to ureteropelvic junction obstruction

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ABSTRACT

Wunderlich syndrome (WS) is defined as a rare spontaneous renal hemorrhage. It mostly occurs with concomitant diseases without trauma. It usually presents with the Lenk triad and is diagnosed in emergency departments with the effective use of advanced imaging modalities such as ultrasonography, computerized tomography, or magnetic resonance imaging scanning. In the management of WS, conservative treatment, interventional radiology, or surgical procedures are decided according to the patient's condition and treated appropriately. Conservative follow-up and treatment should be considered in patients whose diagnosis is stable. If diagnosed late, the progression can be life-threatening. As an interesting case of WS, a 19-year-old patient was presented with hydronephrosis due to ureteropelvic junction obstruction. Spontaneous renal hemorrhage without a history of trauma is presented. The patient, who presented to the emergency department with the sudden onset of flank pain, vomiting, and macroscopic hematuria was imaged by computed tomography. The patient could be followed and treated conservatively for the first 3 days, and on the 4th day, his general condition deteriorated, and he underwent selective angioembolization and then laparoscopic nephrectomy. WS is a serious, life-threatening emergency, even in young patients with benign conditions. Early diagnosis is mandatory. Delays in diagnosis and non-energetic approaches can lead to life-threatening situations. In hemodynamically unstable non-malignant cases, the decision for immediate treatment, such as angioembolization and surgery, should be taken without hesitation.

Keywords: Laparoscopic simple nephrectomy; selective angioembolization; spontaneous renal hemorrhage; wunderlich syndrome.

INTRODUCTION

Wunderlich syndrome (WS) is an acute, spontaneous, urological emergency characterized by nontraumatic subcapsular and perirenal hemorrhage. Idiopathic WS is a scarce condition. WS was first described in 1856 by Carl Reinhold August Wunderlich from Germany.[1] The most common benign and malignant causes are renal angiomyolipoma and renal cell carcinoma.[2] Other causes are stone-related obstruction, renal artery rupture, arteriovenous malformations, polyarteritis nodosa, renal cysts, and infection. Retroperitoneal bleeding causes life-threatening blood loss and hemodynamic instability.[3] Patients generally apply to hospitals with acute flank pain, palpable flank mass, and hypovolemia, known as the components of Lenk's triad.[4] Ultrasound, multidetector computed tomography, and/or magnetic resonance imaging play a significant role in the diagnosis.[5] Conservative follow-up should be considered in patients with stable conditions. In unstable patients, nephrectomy should be regarded as if transcatheter angioembolization fails.

CASE REPORT

A 19-year-old male was applied to the emergency department with complaints of sudden and persistent right flank pain, vomiting, and macroscopic hematuria. There was no history or evidence of physical trauma. Physical examination revealed pale skin, cold sweating, and tenderness at the right

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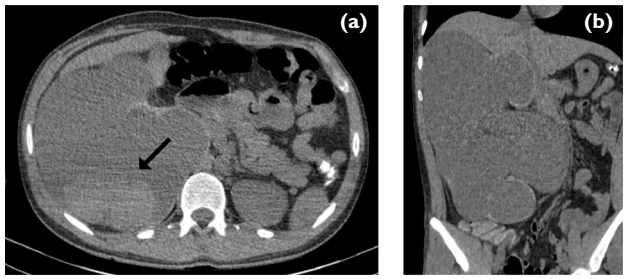

Figure 1. (a) Axail and **(b)** Coronal images of hydronephrotic kidney with hemorrhage (arrow)

costovertebral angle. Blood pressure and heart rates were 90/60 mm/Hg and 120/min, respectively. Body temperature was 36°C. There was no previous surgery or systemic diseases in the past medical history. Blood count indicated deep anemia secondary to acute blood loss. Hemoglobin was 7.2 g/dL, hematocrit 23,5%, WBC count was 9700 cells/μL, platelets were 164000, prothrombin time, INR, and aPTT were normal. Renal and liver function tests were completely normal. The patient had HBsAg positivity.

Emergency abdominal ultrasound and noncontrasted computerized tomography (CT) revealed a huge-sized right kidney with severe hydronephrosis (Fig. 1). CT images were suggestive of ureteropelvic junction obstruction as the cause of hydronephrosis. At this stage, because of the patient's unstable condition, urography and renal scintigraphy were not considered necessary.

Subsequently, the patient was transferred to the urology ward, and three units of erythrocyte suspension were transfused. After the patient's vital signs were stabilized, a percutaneous nephrostomy tube was placed into the right kidney for pain management and monitoring bleeding. During the 2nd and 3rd days in the ward, the patient had manageable pain and completely normal vital signs. The urine from the nephrostomy tube was dark brown but not actively hemorrhagic. On the 4th day, the situation changed rapidly. Low blood pressure (65/35 mmHg), tachycardia (over 120/min), and other signs of hypovolemia were observed. Aggressive liquid replacement and blood transfusions were made. The patient was immediately transferred to the Interventional Radiology room for angioembolization. The patient was not stable after renal artery coil placement, and an urgent nephrectomy was planned. Laparoscopic transperitoneal nephrectomy was performed. Immediately after vascular (artery and vein) ligation, the patient rapidly stabilized (Fig. 2).

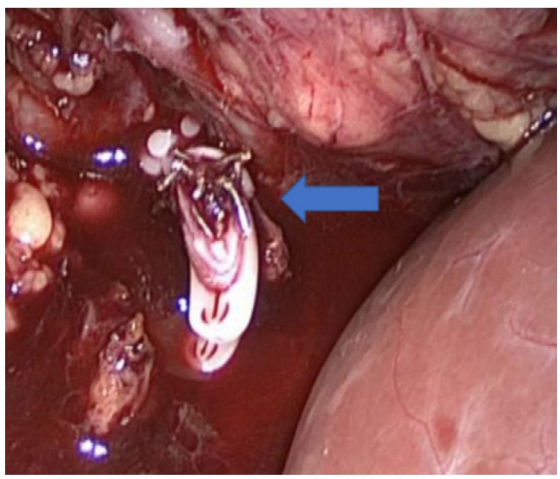

Figure 2. Clipped renal artery with endovascular coil (arrow)

Postoperatively, two more units of erythrocyte suspension were transfused. Kidney function tests were not changed. The urethral catheter and abdominal drain were removed on the 2nd and 4th postoperative days. Finally, he was discharged from the hospital on the 7th day postoperatively.

Histopathology was reported as hydronephrotic changes, parenchymal atrophy, mixt type inflammatory infiltration with fresh bleeding and significant hyperemia, chronic interstitial nephritis, angiomatous changes, and massive interstitial hemorrhage.

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

DISCUSSION

Spontaneous renal bleeding is a rare condition. Although renal tumors are responsible in most cases, this condition may less frequently be caused by mild infections such as hydronephrosis, pyelonephritis, tuberculosis, abscess, nephrolithiasis, nephritis, aneurysm, infarction, and autoimmune diseases.[4] Computed tomography is the most effective imaging method for diagnosis. CT allows us to see intrarenal bleeding as well as perinephric and subcapsular hematomas and, in the majority of cases, helps to find the underlying cause.[4]

Bleeding into the renal collecting system is usually self-limited. Unfortunately, the same cannot be assumed for hydronephrotic kidneys. Even though the bleeding is not excessive, it can turn into a life-threatening situation. For the hydronephrotic kidneys, the area filled with blood is more spacious and less restrictive. The presence of hydronephrosis and infection weakens the kidney structurally, making it more fragile and may cause spontaneous ruptures.

The majority of patients treated conservatively are those with benign conditions. It should be noted that the state of patients who are considered to be stable may be unpredictable and may change dramatically. In hemodynamically unstable patients, treatment options are selective angioembolization or emergency nephrectomy.[6,7] Although there are reported partial excision and repair cases, the surgical procedure usually ends with nephrectomy in most cases.

Conclusion

WS is an entity that can be easily diagnosed due to its typical clinical features together with the help of imaging techniques. Both selective angioembolization and surgical exploration are lifesaving treatments. More research and clinical experience are needed to determine underlying predisposing factors and pathogenesis, especially in patients with benign conditions.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Conflict of Interest: None declared.

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OLGU SUNUMU - *ÖZ*

Üreteropelvik bileşke darlığına sekonder Wunderlich sendromu

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Wunderlich sendromu, nadir görülen bir spontan böbrek kanaması olarak tanımlanır. Çoğunlukla travma olmaksızın zemininde eşlik eden hastalıklarla ortaya çıkar. Genellikle Lenk triadı ile prezente olur ve acil servislerde ultrasonografi, BT veya MRG taraması gibi gelişmiş görüntüleme yöntemlerinin etkin kullanımı ile teşhis edilir. Wunderlich sendromunun tedavisinde konservatif tedavi, girişimsel radyoloji veya cerrahi prosedürler kullanılarak, hastanın durumuna göre karar verilir ve uygun yöntemle tedavi edilir. Tanı da stabil olan hastalarda konservatif takip ve tedavi düşünülmelidir. Geç teşhis edilirse, progrese olması halinde hayat tehdit edici olabilir. İlginç bir Wunderlich Sendromu olgusu olarak, 19 yaşında üreteropelvik bileşke (UPJ) obstrüksiyonuna bağlı hidronefroz hastasında, travma öyküsü olmayan spontan böbrek kanaması sunulmaktadır. Ani başlayan yan ağrısı, kusma ve makroskopik hematüri şikayetleri ile acil servise başvuran hastanın bilgisayarlı tomografisi çekildi. İlk üç gün konservatif olarak takip ve tedavi edilen hasta, 4. günde genel durumu bozulması üzerine hastaya selektif anjiyoembolizasyon ve ardından laparoskopik nefrektomi uygulandı. Wunderlich sendromu, benign hastalıklar zemininde genç hastalarda dahi ciddi, yaşamı tehdit eden bir acil durumdur. Erken teşhis önem arzeder. Tanıdaki gecikmeler ve enerjik olmayan yaklaşımlar yaşamı tehdit eden durumlara yol açabilir. Hemodinamik olarak stabil olmayan benign vakalarda da, anjiyoembolizasyon ve cerrahi gibi acil tedavi kararı tereddüt etmeden alınmalıdır.

Anahtar sözcükler: Laparoskopik basit nefrektomi; selektif anjioembolizasyon; spontan böbrek kanaması; Wunderlich sendromu.

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