

Sudden Onset Right Flank Pain: A Case Report of Wunderlich's Syndrome in Emergency Department Related to Ruptured Renal Angiomyolipoma

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Abstract

We reported a case of Wunderlich's syndrome in a patient with massive retroperitoneal haemorrhage who was admitted to the Emergency Department with acute onset right flank pain that began in the previous 2 days. During the stay in ED, unstable hemodynamics were occurred due to haemorrhagic shock and nephrectomy was planned. In the postoperative period, the patient had good convalescent and no complications were detected. After 10 days of follow-up, the patient was hemodynamically stable, blood laboratory results were normal and the patient was discharged. In addition, no hemorrhage, no metastasis and no recurrence were observed at the end of the 6-months follow-up.

Keywords: Hemodynamics; Syndrome; Diagnosis; Disorder

Introduction

Acute flank-abdominal pain are one of the frequent disorder for patients in Emergency Medicine Departments (ED) and physicians should reduce mortality and morbidity with careful observation and immediate intervention in these cases. The etiologies of acute surgical or fatal abdominal emergencies except for the Wunderlich's syndrome, such as acute cholecystitis, acute appendicitis, intestinal perforation, mesenteric ischemia, aortic dissection, ruptured aortic aneurysm, are also known by physicians.

Wunderlich's syndrome is described as spontaneous non-traumatic renal bleeding confined to the subcapsular and/or perinephric area [1]. The syndrome is clinically characterized by Lenk's triad, which includes flank or abdominal pain, palpable mass, and hypovolemic shock due to spontaneous retroperitoneal haemorrhage. Various etiologies are identified for the syndrome, such as benign and malignant renal neoplasms, vascular diseases, nephritis, infections, hematological disorders, and anatomical lesions. In recent studies, Renal Angiomyolipomas (R-AMLs) are the most common cause of the Wunderlich's syndrome among the other etiologies [1]. R-AMLs are rare tumors and more than 50% of patients with R-AMLs are related with Tuberous Sclerosis (TS) [2]. In recent studies, the prevalence of R-AMLs are varied between 0.28% and 0.60% [3,4]. As a mesenchymal tumor, R-AMLs are composed of smooth muscle, blood vessels, and adipose tissue [3]. In recent decades, the incidental diagnosis of R-AMLs are increased due to the frequent use of cross-sectional imaging in asymptomatic patients [4]. In addition, the most common symptomatic presentation is life-threatening spontaneous retroperitoneal haemorrhage in patients with R-AMLs [4].

In our report, we have presented a rare case of a patient, who was admitted to the ED with flank pain and after four hours follow-up in ED, hypovolemic shock was occurred due to spontaneous retroperitoneal haemorrhage. Finally, the patient was diagnosed as Wunderlich's syndrome related to ruptured R-AMLs.

Case Presentation

In December 2017, a 67-year-old female patient, without any previous medical history, was admitted to the ED with acute onset right flank pain that began in the previous 2 days. There was no history of weight loss and loss of appetite. On admission, the patient was hemodynamically stable with normal sinus rhythm (67/minute)

and normotensive (110/60 mmHg), respiratory rate was 16 breaths per min, body temperature was measured 36.8°C and GCS 15. Physical examination was unremarkable except for tenderness of the right flank region. In addition, physical examination revealed symmetrical pulses in the extremities and no signs of pulsatile abdominal mass with palpation of abdomen. Blood laboratory results were normal except for WBC; 15.8 × 10³/ml, haemoglobin 11 mg/dl, CRP 51 mg/L and urine dipstick test revealed haematuria. In ED, the patient underwent an ultrasound examination by Emergency Physician and an echogenic solid mass was identified around the right kidney but not clearly visualized. Contrast-enhanced Computerized Tomography was thereupon performed and revealed a right renal tumor which was 7 × 9 cm in diameter with comprising areas of retroperitoneal haemorrhage with a ruptured subcapsular haematoma (Figures 1 and 2). We immediately sought consultation from a surgeon and a urologists, after CT scanning. During the stay in ED, unstable hemodynamics were occurred due to haemorrhagic shock and at the same time, the patient's haemoglobin was diminished to 8.4 mg/dl. Thus, nephrectomy was planned and the patient was transfused simultaneously. Inoperative diagnosis was based on actively bleeding retroperitoneally of ruptured R-AML by Urologists and pathological findings were confirmed diagnosis of R-AML. In the postoperative period, the patient had good convalescent and no complications were detected. After 10 days of follow-up, the patient was discharged. In addition, no hemorrhage, no metastasis and no recurrence were observed at the end of the 6-months follow-up.

Discussion

Wunderlich's syndrome is a rare clinically presentation of abdominal-flank pain in ED. Early treatment of the syndrome is crucial to reduce mortality and morbidity. Treatments of ruptured R-AMLs

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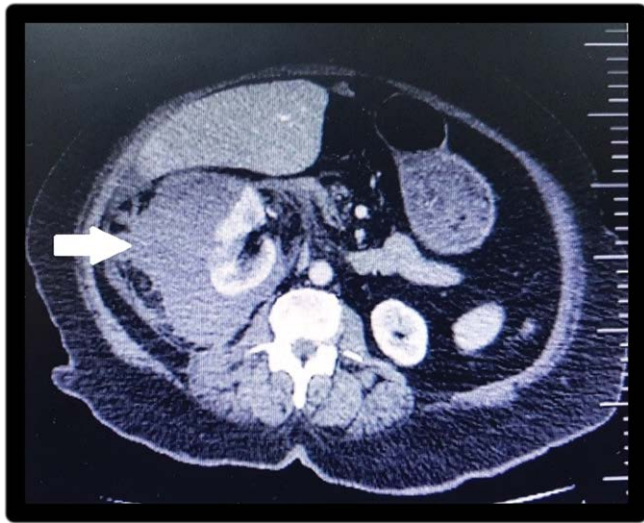


Figure 1: CT scan of the abdomen was confirmed the presence of a perinephric fatty solid lesion, defined as R-AMLs (white arrow).

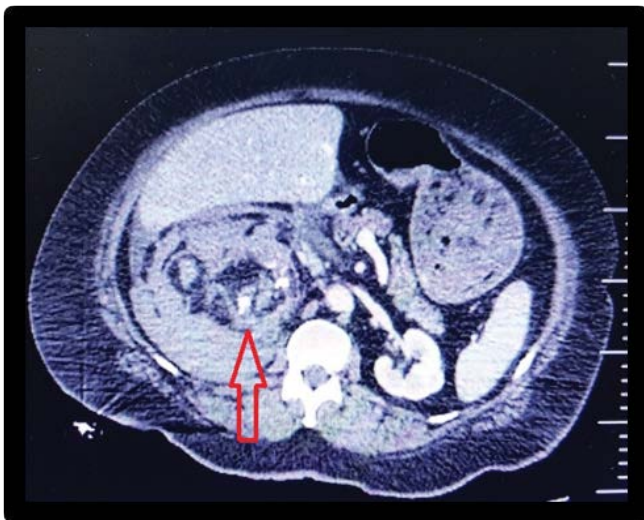


Figure 2: Diagnosis was consistent with angiomyolipoma complicated by haemorrhage (red arrow).

are included selective Renal Artery Embolization (RAE), Nephron-Separating Surgery, Complete Nephrectomy, cryo- and radiofrequency ablation, and treatment with mTOR inhibitors [5]. In a prior study, the use of a larger than 4 cm tumour size was suggested as a criterion for surgery treatments (complete nephrectomy, nephron-sparing surgery) [6]. However, In recent studies, the main indications for intervention are the presence of the symptoms, suspicion of malignancy, diagnosed in female patients at childbearing age, and larger than 4 cm tumors [4]. Selective transarterial RAE is now the first-line treatment option,

especially in the event of acute bleeding or hemodynamic instability [4,7].

In a study, 58 patients treated with nephron-separating surgery for sporadic renal angiomyolipoma, between 1970 and 2004, and the study showed that recurrence rates after surgery are extremely rare [8]. However, in a review; RAE required repeat procedures in 14% of cases after a follow-up of 23 months; these were usually for recurrent symptoms or bleeding [9]. In other study, 59 patients were treated for R-AMLs with RAE or surgery. In the study, surgical management was included 25 (60%) patients underwent radical nephrectomy, 17 (40%) patients underwent partial nephrectomy, 17 (40%) patients were treated by RAE. The study was detected that RAE allows rapid stabilization in patients with acute haemorrhage and provides good renal preservation in patients with multifocal R-AMLs. In the study, RAE was suggested in large masses when partial nephrectomy is not feasible. However, surgery was recommended in cases of diagnostic uncertainty or complex vascular anatomy not amenable to RAE [10].

Conclusion

In our case, haemorrhagic shock was occurred in the patient due to actively bleeding retroperitoneally. We planned partial or total nephrectomy simultaneously and transferred the patient to the operating theater promptly. In operation, Urologists preferred total nephrectomy to make urgent exploration and to reduce fatal risks due to life-threatening haemorrhage. In addition, interventional angiographic radiology clinic was not available at the time, in our medical center. In a conclusion, physicians should be alert on the patients with potentially fatal causes of flank-abdominal pain, such as Wunderlich's syndrome. In ED, the early diagnosis of the fatal and rare etiologies of flank-abdominal pain is crucial to improve outcomes for patients.

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