CASE REPORT

Selective renal artery embolization for the management of Wunderlich syndrome in a horseshoe kidney

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Abstract

The spontaneous, non-traumatic rupture of the kidney is called Wunderlich’s syndrome and represents an emergency, possibly life-threatening condition. Tumors of renal parenchyma, especially benign angiomyolipomas are the most common cause. On the other side, horseshoe kidney is the most frequent congenital abnormality of the kidneys, characterized by the fusion of the two organs by a functioning parenchymal or fibrous band. In this paper, a rare case of spontaneous rupture of an angiomyolipomas in a horseshoe kidney is presented, which threatened the life the patient. The condition was managed successfully via angiography and selective embolization of the bleeding site.

Introduction

The spontaneous, non-traumatic rupture of the kidney, also known as Wunderlich’s syndrome, is an infrequent urologic emergency which may become life-threatening. Tumors and especially angiomyolipomas (AML) are reported as the major cause of bleeding. The latter may occur incidentally or accompany disorders like tubular sclerosis and lymphangioleiomyomatosis. Nevertheless, the syndrome is not associated with horseshoe kidney (HSK) which represents the most common congenital renal abnormality. We herein present a rare case of Wunderlich’s syndrome in a young man with...
history of horseshoe kidney. The patient was treated with selective renal arterial embolization.

**Case presentation**

A 39-year old man presented to our department reporting acute onset of diffuse, abdominal pain. The patient denied history of trauma or recent other illnesses. He reported a known history of horseshoe kidney diagnosed 1 year ago due to urinary lithiasis, for which he underwent percutaneous nephrolithotomy. In addition, the patient reported to have a small AML of 2 cm maximal diameter in his kidney. Physical examination demonstrated a painful abdomen and lumbar region, while a mass was palpable abdominally. The vital signs of the patients were normal, reflecting no hemodynamical instability. Hematocrit was 36% and urea and creatinine levels were within normal limits. A contrast enhanced computed tomography was performed which demonstrated a bleeding site in the right section of the horseshoe kidney (**figure 1a**), which resulted in a large retroperitoneal hematoma (**figure 1b**). The patient was hemodynamically stable; thereby, the conservative management was decided, with close monitoring of the vital signs and hematocrit.

During the next hours, the patient gradually demonstrated signs of hypovolemic shock, while hematocrit reached nadir of 22%, despite intravenous infusion of colloid solutions and transfusion with 5 units of concentrated red blood cells. Thus, the performance of renal angiography and if possible, selective embolization of the bleeding vessels was decided. During the angiography session, bleeding vessel on the right median to lower pole of the horseshoe kidney was observed. The vessel was occluded with coils and the bleeding was successfully ceased, while an infarcted area was left behind (**figure 2**). The patient was further transfused with 2 more units of concentrated red blood cells. The hematocrit was stable after the transfusions. Nevertheless, the intraabdominal collection of fluid resulted in bilateral thoracic collections with the right side collection having significantly higher volume. The patient respiratory capacity gradually deteriorated and the drainage of the right thoracic collection was decided on the 5th day after the embolization. The respiratory status was not improved and the drainage of the left collection was also decided (8th day) (**figure 3**). The patient respiratory capacity was improved and he was eventually discharged after removing the thoracic drainages and a total hospitalization of 17 days (**figure 4**). At day of discharge, the hematocrit was 36%. Urea and creatinine levels were 24 mg/dl and 0.8 mg/dl, respectively.

**Discussion**

Wunderlich’s syndrome is defined as a spontaneous renal bleeding, confined in subcapsular, perinephric and retroperitoneal spaces1. The clinical manifestation of the condition has been described as Lenk’s triad consisting of acute lumbar and abdominal pain,
palpable abdominal mass and hemodynamic deterioration which may result in hypovolemic shock. Hematuria may also be present but does not necessarily reflect the severity of the condition. Angiomyolipomas, renal cell or urothelial carcinomas are reported as the main origin of bleeding, while uncontrolled anticoagulation therapy, infection or vascular pathologic lesions like panarteritis nodosa or vasculitis have been reported less common causes. AMLs are reported as the most common cause of the syndrome; these mesenchymal tumors are benign and consist of 3 components; abnormal vessels, special spindle cells and mature adipocytes. The diagnosis of the condition is established with computerized tomography imaging, which demonstrates perirenal or retroperitoneal hematoma. Nonetheless, the CT cannot always determine the causative lesion and only the presence of fat density areas may justify the diagnosis of AML. The therapeutic approach is based on the performance of renal angiography with embolization which results in control of the bleeding in the majority of the patients and the excision of the kidney is avoided.

Horseshoe kidney is the most common congenital
renal abnormality and its incidence is estimated at one in 400-800 births. The most common pathologic consequences are emerged from obstruction which may result in calculi formation (up to 60%) and infection. Tumorigenesis is presented with higher incidence than the normal population. Renal cell carcinomas have been observed as the majority of tumors affecting HSK and spontaneous rupture of such tumors has been reported. However, the presence of a AML in HSK has been limited reported in the literature.

Considering the above evidence, the currently presented case is uncommon and has been described very limited in the literature. The management of the current case was based on the support of the vital signs, contrast enhanced tomography and selective angiographic embolization. The latter algorithm would have also followed in the case of a prolonged bleeding of the kidney. Nevertheless, vascular abnormalities of HSKs may be present in up to 80% of the cases and pose a challenge for the diagnosis and subsequent angiographic management. These abnormalities may also include supernumerary main arteries accompanying the fusion and may originate from the aorta, the mesenteric or the iliac arteries. In the current case, the contrast enhanced CT before the definitive treatment did not provide diagnosis of the bleeding site and the angiographic investigation was decided based on clinical criteria. In fact, the presence of the AML and its possible implication to the bleeding may have been impossible to diagnose if the history of the patient was not available to our department. The combination of the modalities eventually resulted in successful resolution of the bleeding and a surgical exploration was avoided. The thoracic collections were related to the large retroperitoneal bleeding and were successfully managed. Nevertheless, these collections could develop life threatening conditions and the treating physician should always treat them with extra care.

As a conclusion, Wunderlich’s syndrome is a life-threatening condition which demands prompt intervention. When renal bleeding is complicated with HSK, the urologist has to take into consideration the vascular variations that accompany the kidney. The combination of contrast enhanced computer tomography and angiography offers beneficial information for a successful embolization. In our case, such an approach led to definitive resolution of bleeding, eliminating the need of surgical intervention. 

Conflicts of interest
The authors declared no conflicts of interest.

Abbreviations
AML = Angiomyolipoma
HSK = Horseshoe kidney
CT = Computer Tomography
Η αυτόματη, μη τραυματική ρήξη του νεφρού, ή αλλιώς σύνδρομο Wunderlich, αποτελεί μια οξεία, δυνητικά απειλητική για τη ζωή κατάσταση. Οι όγκοι του παρεγχύματος, και ειδικά τα καλοήθη αγγειομυολιπώματα είναι η συνήθεστερη αιτία. Από την άλλη πλευρά, ο πεταλοειδής νεφρός είναι η πιο συχνή συγγενής ανωμαλία των νεφρών, χαρακτηριζόμενος από την ένωση των δύο οργάνων μέσω μιας λειτουργικής παρεγχυματικής ή μιας ινώδους ταινίας. Στην παρούσα εργασία παρουσιάζεται μια οποία περίπτωση αυτόματης ρήξης ενός αγγειομυολιπώματος σε πεταλοειδή νεφρό η οποία απέληφε τη ζωή του ασθενούς. Η κατάσταση αντιμετωπίσθηκε επιτυχώς μέσω αγγειογραφίας και εκλεκτικού εμβολισμού της αιμορραγούσας εστίας.

**Περίληψη**


**References**

**Λέξεις ευρετηριασμού**

σύνδρομο Wunderlich, πεταλοειδής νεφρός, αγγειομυολίπωμα, αγγειογραφία, εμβολισμός