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Wunderlich syndrome as the presenting future of polyarteritisnodosa

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ABSTRACT

Wunderlichsyndrome (WS) is an uncommun condition characterised by acute onset of spontaneous, non traumatic renal hemorrage into the subscapular and perirenal spaces. WS is clinically characterised by thelenk's triad: acute lombo-abdominal pain, palpable mass and general deterioration with hypovolemic shock. The most commun causes of WS are angiomyolipoma and renal cell carcinoma. Vasculitis is a rare cause of WS. We describe a 50-year-old woman with spontaneous renal hemorrhage, ascribed to polyarteritisnodosa (PAN) after serologic testing, computed tomography, and angiographic studies.

Key words: polyarteritisnodosa; vasculitis; Wunderlich syndrome.

INTRODUCTION

WS is a rare condition classically defined as spontaneous renal bleeding of non traumatic origin, confined to the subscapular and perirenal space[1].the clinical manifestations can be varied and non specific [2]. It is of great importance to recognize this disease entity early enough to permit medical or surgical hemostasis [3]. A wide spectrum of neoplastic and non neoplastic renalpathologies may result in WS [2, 4]. Renal neoplasms are the most common cause for WS, with angiomyolipoma being the most common benign neoplasm, whereas renal cell carcinoma is the most common malignant neoplasm [5].Vascular causes of Wünderlich syndrome are infrequent.In this report, we describe a patient with PAN and renal hemorrhage and review the use of computed tomography, angiographyand immunosuppressive therapy in the management of the hemorrhage and the active PAN.

MATERIALS AND METHODS

CASE RAPPORT: A 50-year-old woman was admitted to our department in June 2013, complaining of weakness and right lower radiating to the back. She had been in good health until April 2013, when she complained of anorexia and generalized myalgia, arthralgia of small joints. Though there was moderate weight loss, she denied nausea, vomiting, or black stools and had no history of trauma, bleeding diathesis, or anticoagulant therapy. There was no history of oral ulcers,

or photosensitivity. There was also no history of oliguria, hematuria, cough, or shortness of breath. On admission, physical examination revealed moderate pallor and a blood pressure of 140/90 mmHg. There was a livedoreticularis in both legs without lymphadenopathy. There was no hepatosplenomegaly but a palpable mass in the right paraumbilical region that was firm, mildly tender, nonmobile with vague margins. Other systems were normal. Laboratory evaluation revealed hemoglobin 8,2 g/dL, white blood cell count 10,000 cell/mm3 with normal differential count and platelets 189,000/mm3, with elevated indirect bilirubin and serum lactate dehydrogenase.Prothrombin and partial thromboplastin time were mildly elevated. Fibrinogen and haptoglobinlevels were normal. Direct Coombs test was negative. Peripheral smear did not show schistocytes or spherocytes. Fibrinogen and haptoglobinlevels were normal. C3and C4 complement levels were normal.

The creatinine (Cr) was 0.7 mg/dl and C-reactive protein (CRP) 21.6 mg/dl. Urinary protein excretion was about 1.5 g/day, 2+ hematuria, suggesting active glomerulonephritis.

Hepatitis B Surface antigen, and Serum titers of antinuclear antibody (ANA), anti DNA, anti-Sm, anti-RNP, anti-SS-A/Ro, anti-SS-B/LA, MPO-ANCA, and PR3-ANCA were all negative.

The abdominal ultrasonography and computed tomography (CT)showed a large hematoma encroaching on the right kidney, with compression of the renal parenchyma and multiple hypodense lesions in spleen[Figure 1]. The rest of the viscera were unremarkable, and there was no abnormal fluid accumulation within the Morrison pouch, splenorenal space, or Douglas pouch. CT-angiography also revealed bilateral, multiple aneurysms sacs in both kidneys [Figure 2, 3]. The aorta, main renal arteries, and mesenteric arteries were normal without obvious stricture or aneurysmal dilatation. Based on the clinical picture and multiple aneurysms on arteriogram, a diagnosis of polyarteritisnodosa (PAN) was made inaccordance with the American College of Rheumatology (ACR) 1990 criteria. She was given three doses of intravenous methyl prednisolone (1 gm each) followed by intravenous cyclophosphamide (750 mg/m2).Although there was no further bleeding but she developed a severe pneumonia and died of respiratory failure 3 weeks after the episode of renal hemorrhage.

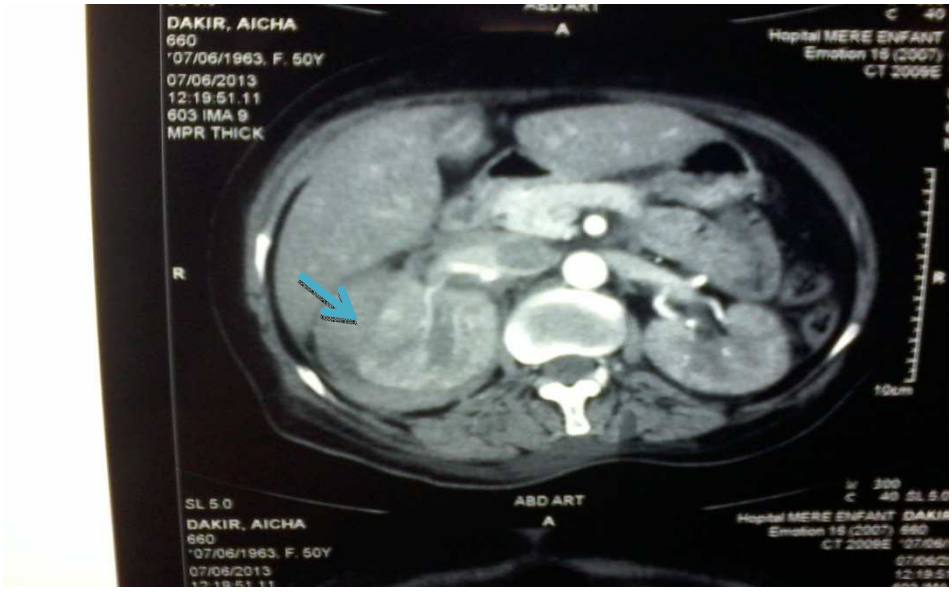

Figure 1: CT scan showing a large right perinephric hematoma pushing the kidney anteriorly.

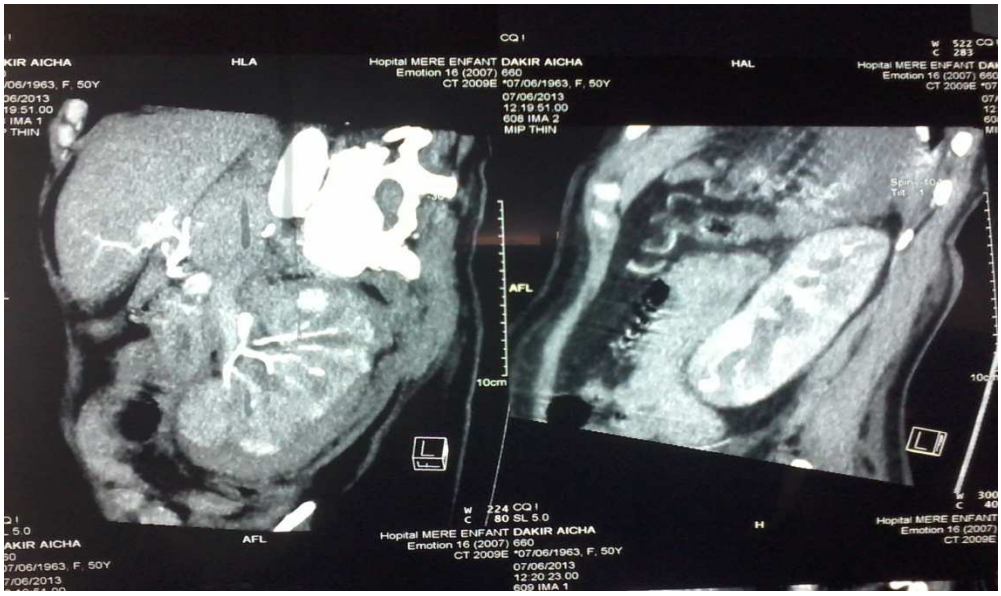

Figure 2:CT angiogram indicating multiple microaneurysms and a large aneurysm (arrow) in the left kidney.

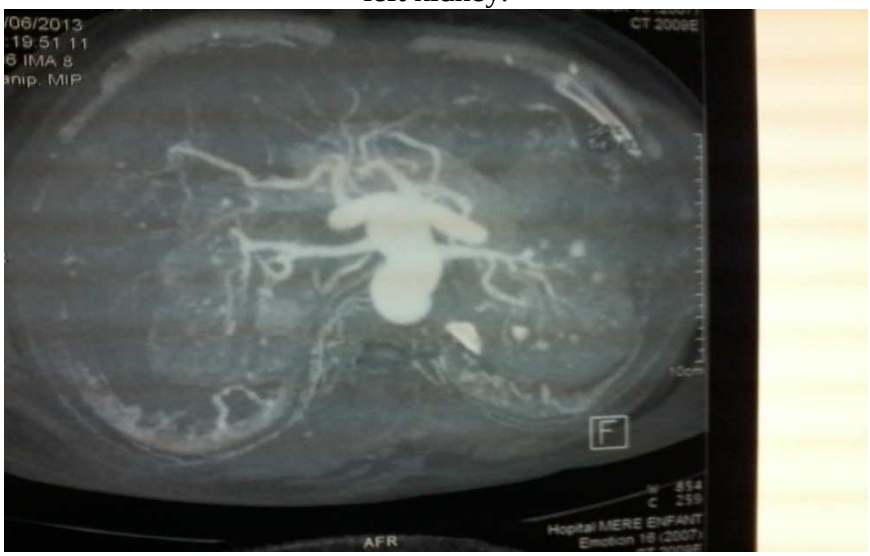

Figure 3: CT angiogram showing multiplemicroaneurysms in both kidneys with a large aneurysm (arrow) in the right kidney.

RESULTS AND DISCUSSION

Carl Wunderlich is credited with the first clinical descriptionof WS in 1856 as spontaneousrenal hemorrhage confined to the subcapsular and perinephricspace in patients with no history of trauma [6]. Patients may present with the classic Lenk triad, including acute flank or abdominal pain, palpable flank mass, and hypovolemia, but symptoms also can be obscure and subtle, depending on thevolume of blood loss and rapidity of onset [2]. Zhanget al [7], in a meta-analysis of 165 cases, found that80% of patients presented with acute flank or abdominalpain, but only 10% manifested hemodynamicinstability.Benign and malignant neoplasms accounted for the majority of cases (62.2%), with the most common being angiomyolipoma and renal cell carcinoma [7]. Other causes

of WS include vascular causes, cystic renal diseases, renal calculi, nephritis and coagulation disorders [2,8].In 1908 Schmidt described the first case of WS caused by rupture ofmicroaneurysms in a patient with PAN [9].Our patient presented with the typical features of Lenk triad except for the hypovolemia.

According to the ACR and Chapel-Hill Consensus Conference Classifications, PAN describes the pathologic process of necrotizing inflammation of medium or small arteries without involvement of arterioles or venules [10,11].Most cases of PAN occur in the fourth or fifth decade, although it can occur at any age and there is male preponderance. An association with hepatitis B virus infection is welldocumented in 7–36% of cases [11, 12].Diagnosis of PAN should be confirmed with biopsy of the involved organs: the skin and peripheral nerves are preferred as they are less invasive. Kidney biopsy may be complicated by excessive bleeding due to the presence of aneurysms. In the absence of biopsy, diagnosis can be made with angiogram in association with other clinical features in accordance with ACR 1990 criteria to diagnose PAN (3 or more yield 82% sensitivity and 87% specificity for diagnosis) [11].

Our patient was diagnosed as PAN in view of weight loss, general weekness, livedoreticularis and multiple, bilateral renal microaneurysms on angiography. Involvement of the splenic arteries is a rare occurrence in PAN. In our patient there were multiple hypodense lesions in spleen in CT scan. Most probably these were splenic infarcts secondary to involvement of splenic arteries.

A ruptured aneurysm resulting in a huge perirenal hematoma usually necessitates an emergency operation.Sometimes urgent nephrectomy is life-saving[12]. But few cases can be treated by a conservative drainage of the hematoma or by selective arterial embolization[13]. A concomitant immunosuppressive therapy may be warranted in active disease. Conventional treatment with corticosteroids and cyclophosphamide has shown a better outcome compared to steroids alone. Five-year survival in untreated and treated PAN is 13% [12] and 80% [14], respectively. Antiviral agents and plasma exchanges are indicated in the treatment of Hepatitis B virus-associated PAN [15].

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