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An Enigma of Wunderlich Syndrome

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Abstract

Wunderlich syndrome (WS) is an extremely rare condition that involves spontaneous non- traumatic haemorrhage within the subcapsular and perirenal spaces of the kidney. Here we present such a case of a 57 year old male patient who presented with sudden onset hypotension and an eventual diagnosis of Wunderlich Syndrome secondary to Polyarteritis Nodosa on the basis of clinical and radiographic evidence was made. Spontaneous perinephric hemorrhage is a rare, but potentially devastating complication of PAN which is what the unfortunate outcome in our patient was. Only about 60 cases have been reported in the literature. Ours is one such enigmatic case from India which shows us that a high index of suspicion is required to diagnose such cases.

Keywords: Wunderlich Syndrome, Polyarteritis Nodosa, non traumatic hemorrhage

Introduction

Wunderlich syndrome (WS) is an extremely rare condition that involves spontaneous non- traumatic haemorrhage within the subcapsular and perirenal spaces of the kidney. This is a potentially life-threatening emergency condition. Although the first glimpse of this condition was observed by Bonet dates back to 1700, it was first described by Wunderlich in 1856 as 'Spontaneous renal capsule apoplexy.' The actual term 'Wunderlich syndrome' was later coined by Coenen in 1910.[1,2] Clinically this condition presents with acute flank pain, palpable flank mass and hypovolemic shock together named as 'Lenk's triad'.[3] Here we present a case of a 57 year old male patient who presented with sudden onset hypotension and an eventual diagnosis of Wunderlich Syndrome secondary to Polyarteritis Nodosa.

Case Report

A 57 year old male patient presented to the hospital with history of fever with chills and rigor associated with cough and mucopurulent expectoration; significant weight loss since 1.5 months. The patient had a history of Pulmonary Tuberculosis in 1993 for which he had completed CAT 1 DOTS and declared cured. He was diagnosed 3 months back with with Type 2 Diabetes Mellitus and was initiated on oral hypoglycemic drugs for the same. On examination, he was febrile with an otherwise normal systemic examination. He was admitted and underwent a detailed clinical and investigative work up [Table 1].

At this point he was considered to have bacteremia secondary to pyelonephritis and his blood and urine

cultures were awaited. He was treated with antibiotics (Carbapenems-according to the hospital microbiology profile), packed red blood cell transfusion and supportive treatment. Since the patient remained Afebrile, demonstrated clinical improvement and improving lab parameters, the patient was discharged at request to follow up with the culture reports, repeat complete haemogram and renal function tests after 3 days.

The patient lapsed his follow up and a week after discharge the patient was readmitted at night with slurring of speech and altered sensorium. On detailed history taking, the patient had a history of reduced intake of food associated with regular intake of oral hypoglycemic drugs. He was dehydrated other vitals were stable with no localizing signs. Primary evaluation demonstrated symptomatic hypoglycemia with a glucometer reading of 36mg/dl and he was febrile. 25% Dextrose was administered in the casualty, his sensorium improved and he was transferred to the high dependant unit to be monitored. Repeat investigations showed a worsening profile of renal function tests [Table1], thus he was planned for a Non Contrast CT Abdomen and renal biopsy the next morning. Early next morning, the patient developed sudden pain abdomen and multiple episodes of vomiting. On examination, he was conscious and oriented with rapidly falling blood pressure and tachypnea. Systemic examination was normal with no localizing signs. A bedside ABG demonstrated fall of Hb to 4.9 with severe metabolic acidosis and elevated lactate. The pt was immediately shifted to the Intensive Care Unit, stabilized with ionotropes and oxygen support and blood transfusion was initiated. An emergency CT Abdomen was done which demonstrated a large right perinephric and sub capsular hematoma with a small left renal cortical and sub capsular hematoma and moderate free fluid. [Figure 1]

Comprehensively, after radiological investigations it was concluded that the patient's anemia had worsened secondary to acute blood loss with worsening of renal function tests.

He was transfused with packed red blood cell units and planned for renal angiogram with embolisation vs nephrectomy. The renal angiogram revealed bilateral multiple renal arterial microaneurysms with intervening narrowed segments which was suggestive of **Polyarteritis Nodosa.** No active bleeder in the artery was seen. Significant vasospasm was seen in the renal vein and could not be visualized. Before the procedure could be completed and images could be taken, the patient had intraoperative hypotension and collapsed. The procedure was stopped, he was intubated, started on vasopressors, blood products were transfused and one session of Sustained low efficacy dialysis was completed. Yet,the patient's condition continued to deteriorate in spite of 3 vasopressors, he suffered a cardiac arrest and could not be revived. Finally a diagnosis of Wunderlich Syndrome-Secondary to Polyarteritis Nodosa was made.

Discussion

Wunderlich syndrome (WS) is characterized by acute onset of spontaneous haemorrhage which is confined to the subcapsular and perirenal spaces of the kidney with no previous history of trauma.[4] Approximately 450 cases of spontaneous perirenal haemorrhage have been published in literature till 2000 and a further 266 cases more since 2000.[5] The various etiologies of WS include renal neoplasms, vascular diseases, infection, cystic renal diseases, haematological diseases, idiopathic and other rare causes .In literature approximately 5 to 10% of all cases of WS are idiopathic, which refers to the absence of any underlying pathology within the kidney.[6] Of these, malignancies are the most common cause with vasculitis

being the next most common cause, and of these polyarteritis nodosa (PAN) accounts for most cases.[7] PAN is a multi-system necrotizing vasculitis that involves small and medium sized vessels. The kidneys are affected in 80% cases with hypertension, proteinuria and ultimately renal functional deterioration being the most common manifestations.[8] Spontaneous perinephric hemorrhage is a rare, but potentially devastating complication of PAN which is what was the unfortunate scenario in our patient. Only about 60 cases have been reported in the literature. [8] Ours is one such enigmatic case. It is secondary to the rupture of arterial aneurysms that usually involve the renal artery, and its segmental and interlobar divisions. [9] Nephrectomy is associated with a high mortality rate (50%) in the acute setting. [8] Angiography with selective embolization allows preservation of renal parenchyma and is now the treatment of choice in cases of active bleeding.[8] The diagnosis is ideally made by means of biopsy of involved tissue in a patient with the appropriate clinical symptoms and laboratory data, but an angiogram provides the proof in some cases. Most patients with PAN have positive angiographic evidence of their disease, predominantly in the visceral arteries but also in arteries of the extremities and in small branches of the aorta. The most well-known angiographic feature is the presence of so-called microaneurysms in medium or small arteries which visualized in our patient. Arterial occlusive lesions are also a feature, but their frequency is not reported. The presence of aneurysms increases the specificity of the diagnosis of PAN, but in their absence other arterial lesions such as luminal irregularities, stenoses, and occlusions can suggest the diagnosis.[10] Unfortunately before we could complete the procedure and take a biopsy, the patient collapsed and the angiography and planned embolization could not be completed. Based on the evidence of leukocytosis, elevated ESR and CRP, classical microaneurysms on renal angiogram a diagnosis of Wunderlich Syndrome secondary to Polyarteritis Nodosa was made.

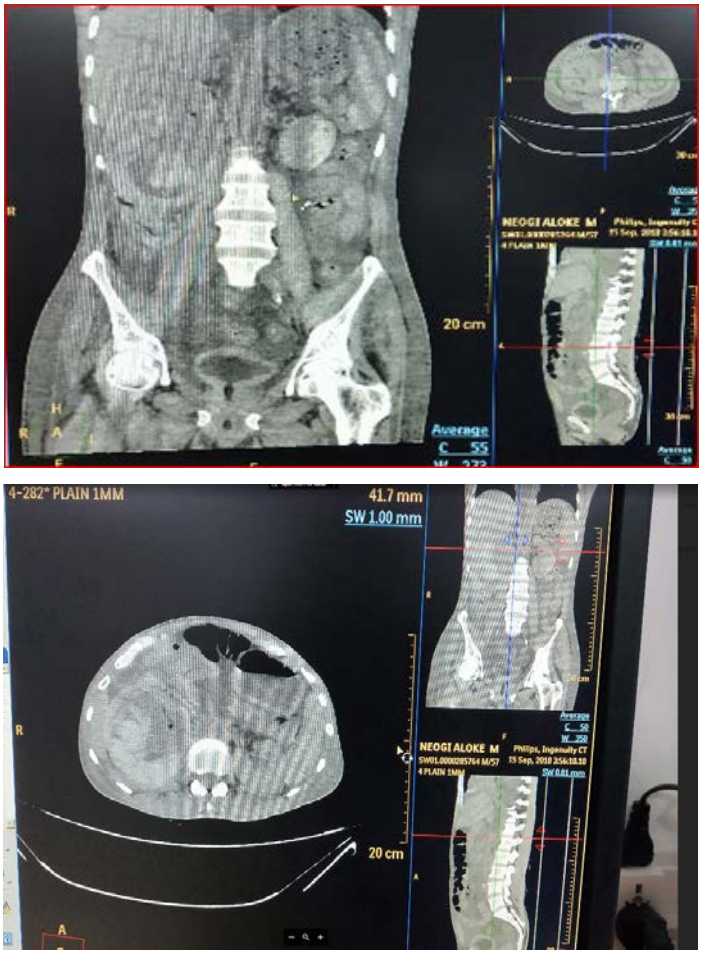

Figure 1A & 1B: Evidence of a large right perinephric and subcapsular hematoma with a small left renal cortical and subcapsular hematoma andmoderate free fluid.

	At 1st Admission	At 2nd Admission
Complete Blood Count	Anemia And Neutrophilic Leukocytosis(18,000cells/Cumm) With Elevated Esr And Crp	Worsening Anemia And Neutrophilic Leukocytosis(22,300cells/Cumm) With Elevated Esr And Crp
Serum Creatinine(Mg/Dl)	1.54	2.89
Urine Routine	Normal	Normal
Serology For Hiv And Hbsag	Negative	
Chest X Ray	Bilateral Upper Zones Fibroreticular Opacities	
Peripheral Smear	Combined Nutritional Anemia	
Ana By Immunofluroscence	Negative	
Sputum Afb (2 Samples)	Negative	
Hrct Chest	No Acute Changes ; Suggestive of Old Tb Sequelae	
Usg Abdomen	Bilateral Grade 1 Renal Parenchymal Changes, Bulky Right Kidney With Perinephric Fluid-S/O Acute Pyelonephritis	
Ecg And 2decho	Normal	
Blood Culture	Negative	Negative
Urine Culture	Negative	Negative
Serum Protein Electrophoresis For M Band	Negative	
Bone Marrow Aspiration And Biopsy	Hypercellular Marrow(Non Specific)	
Upper Gi Scopy (For Anemia Evaluation)	Normal	

Table 1-Investigation Work Up of the Patient.

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