

Case Report

Wunderlich Syndrome in an Elderly Aged Woman Managed with Renal Angioembolisation - Case Report

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Abstract

Angiomyolipoma (AML) is rare and benign lesions and they have been frequently reported in young or middle aged women. We here present an interesting case of an old lady that presented with gross hematuria episode in emergency. She ultimately underwent renal artery Angioembolisation and was successfully managed for Wunderlich syndrome.

Keywords: Angiomyolipoma; Sebaceum; Wunderlich

Introduction

Angiomyolipoma (AML) is quite uncommon and benign lesions having a prevalence rate of 0.3% to 3%. They are comprised of three types of tissue lines: blood vessels, smooth muscle cells and adipose tissue. It is known that the sporadic form of AML is typically found in middle-aged patients. Furthermore, it's more commonly present in women. Apart from it, approximately twenty percent of AMLs occur in patients having tuberous sclerosis syndrome, which is an autosomal dominant abnormality having features of epilepsy, mental retardation and adenoma sebaceum, a distinctive skin lesion. It's pertinent here that massive retroperitoneal hemorrhage in AML cases occur in about 10% of patients. Furthermore, it's the most dreadful complication. In past young age women presenting with Wunderlich syndrome have been treated but old age lady presenting with it is a rare phenomenon. We discuss here an old lady who was being managed with conservation of bleeding kidney by interventional radiologist using Angioembolisation technique.

Case Presentation

A 60-year-old, previously healthy lady was brought with presentation of single episode of hematuria in emergency department at our hospital. She was in a state of shock following sudden severe right flank pain. She had no fever. On examination, she was drowsy, cold, pale, and sweating. She had a pulse rate of 111/minute and

blood pressure of 89/69 mmHg. While examining her, we found severe tenderness in her left flank. In emergency she underwent rapid resuscitation with IV fluids. She was having no edema, Chest, cardiovascular and central nervous system examination was unremarkable and she was having a GCS (Glasgow Coma Scale) of 15/15.

Her blood hemoglobin and hematocrit levels were sent to laboratory, which were 8.2 g/dL and 25.1% respectively, dropped to 7.1 g/dL and 21.4% in 12 hours of emergency presentation. Her white blood cells = 19000/dl while Platelets were 223000/dl, Urine routine exam showed PH of 6, specific gravity (1.05), Red Blood Cells (numerous). Hepatitis B and C status was negative. Her blood grouping and cross match was sent. Her Urea was 65.3 mg/dl, BUN (Blood Urea Nitrogen) was 30.5 mg/dl, Creatinine of 0.7 mg/dl, and eGFR (Estimated Glomerular Filtration Rate) equaling 94 mL/min/1.73 m². Her serum electrolytes were within normal range (sodium = 138 mEq/L, potassium = 4.47 mEq/L, chloride = 110 mEq/L, Calcium = 7.01 mg/dl, Phosphorus = 2.36 mg/dl, Magnesium = 1.99 mg/dl). Meanwhile her Liver function tests were also sent (Total bilirubin = 0.83, direct bilirubin = 0.46, indirect bilirubin = 0.37, ALT = 37, AST = 31, Alkaline phosphatase = 81, Albumin = 3.03 g/dl).

An ultrasound examination of the abdomen and pelvis was also done in emergency that revealed right kidney of normal size, parenchymal thickness and Echogenicity with intact corticomedullary demarcation. (Right kidney size measured 12.2 cm × 6.0 cm × 1.8 cm). Large heterogeneous area of bleed was seen extending from the kidney into perinephric tissue (Figure 1). This collection measured 10.1 mm × 1.0 mm × 9.7 mm with a collective volume computed to be 521 ml. A provisional diagnosis of retroperitoneal mass/collection/hemorrhage was made and a Computed Tomography (CT scan) of the abdomen pelvis was done for accurate evaluation. CT scan of the abdomen and pelvis revealed large right renal Angiomyolipoma. This measured 11.3 cm × 10.3 cm. internal areas of ectasia blood vessels were seen with suspected blur extravasation. These were the

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likely sources of bleed. The right kidney was perfused with the one main renal artery and another accessory renal artery arising below the main renal artery. Tiny left simple cysts were seen with the larger one measures 9 mm × 8 mm. Furthermore, there was normal liver and spleen. Gallbladder and biliary system was normal. Rest of the solid abdominal viscera including adrenals and pancreas were unremarkable. Gut loops were unremarkable. Skeleton was grossly unremarkable. We can see that a large right perinephric collection (hematoma) is displacing the right kidney medially (Figures 1 and 2). A hypodense mass of fat density without calcifications was seen in the inter-polar region of the right kidney, thus these CT findings (Figures 2 and 3) were suggesting perinephric hematoma secondary to rupture of a renal Angiomyolipoma (AML). The patient received two packed cell red blood cells transfusions. That corrected her hemoglobin level. She later on underwent right selective angioembolisation of bleeding vessels and was then shifted to intensive care unit for observation and monitoring. She had improved vitals and relieved pain. She was

discharged home in stable condition after 6 days stay in hospital. On follow up she had no symptoms or paleness or drop of hemoglobin. Her hematuria had settled successfully.

Discussion

Renal AMLs are benign lesions that arise from renal mesenchymal cells. The common presenting feature for patients with AMLs is incidental discovery while performing radiological imaging studies. Most of them are not histologically confirmed as the imaging features of AML are enough to preclude tissue examination. They have characteristic lack of a complete elastic layer that predisposes vascular AML lesion to hemorrhage after minimal or no trauma. Clinically, WS should be suspected if patient presents with Lenk's Triad: acute flank pain, tenderness to palpation and symptoms of internal bleeding [1,2].

According to a classification system suggested by Harabayashi, AMLs are categorized as small (<4 cm), medium (4 cm to 10 cm), or large lesion (>10 cm). The largest ever AML being recorded in the literature reached a size of 36 cm × 15.5 cm × 18 cm and almost weighed 5,600 g.¹⁰ The more the size of an AML, the more is the risk for spontaneous hemorrhage. Beside this, risk factors for hemorrhage are vascular abnormalities, multifocality, aneurysm formation [3,4]. Spontaneous rupture of these lesions in kidney leads to retroperitoneal hemorrhage and it's known as the Wunderlich Syndrome (WS), named after a German physician (Carl Reinhold August Wunderlich).

As, high incidence of spontaneous bleeding is linked to large lesions, therefore early resection of large sized AML might avert situation of life-threatening Wunderlich Syndrome, Imaging techniques are mainly relied upon for their diagnosis. Its management is governed by the clinical presentation of patient. Many authors favor conservative management of WS in case hemorrhage is self-limiting and the patient shows adequate response to fluid resuscitation. However, hemodynamically unstable patients and those refractory to resuscitation measures and transfusion may need nephrectomy on emergency basis if angiographic embolisation is not available in hospital. Emergency nephrectomy (total or partial) controls hemorrhage swiftly and can be performed with low rates of peri-operative mortality. However, it unfortunately has a high incidence of morbidity in terms of renal function loss and later complications ascribed to hemodialysis treatment, using selective arterial embolisation of AML lesion has been suggested by clinicians, especially in those with tuberous sclerosis. It's because of the fact that they might have limited renal reserve due to replacement of the renal parenchymal tissue by multiple cysts and AMLs [5-7].

In our case, we picked up the diagnosis incidentally on ultrasound modality in an old age lady. Then initially she was resuscitated with fluids and transfusions. After that her hemoglobin dropped steadily despite transfusions. So we discussed the case with the interventional radiologists. They performed selective right renal angioembolisation and patient had a good recovery in hospital. Our case underscores the fact that Wunderlich syndrome can present even at such old an age and should ideally be conservatively managed initially otherwise renal selective angioembolisation can help in controlling the bleed and also preserving renal tissue at such an extreme of age where nephron tissue sparing might help in maintaining and preserving renal function of a patient.

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Figure 1: Ultrasound image of right kidney showing bleed extending from kidney into perinephric area.



Figure 2: CT scan image axial view of the right kidney perirenal hematoma displacing kidney.



Figure 3: CT scan image axial view of the right kidney prerenal hematoma extending down in retroperitoneal space.

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