INTRODUCTION

Wunderlich syndrome (WS) secondary to angiomyolipoma (AML) clinically represents with acute flank pain, palpable mass abdomen, and hypovolemic shock, called Lenk’s triad. Urgent contrast-enhanced computerized tomogram (CT) is diagnostic. Urgent selective renal angioembolization or nephrectomy after resuscitation is lifesaving.

CASE REPORT

A 60-year-old female, known to have AML, with no clinical evidence of tuberous sclerosis (TS), presented in the emergency medicine department, with complaints of acute right flank pain, visible gross hematuria, dizziness, and vomiting, for 1 day duration. She was a known diabetic on insulin therapy. On examination, she was pale, her blood pressure (BP) was 111/70 mmHg, and heart rate (HR) was 111/min with a palpable tender mass in the right upper quadrant abdomen. Labs revealed hemoglobin (Hb) of 7 g/dl and her other labs including coagulation screen were normal. On admission in ward, her BP fell to 70/30 mmHg with HR 130/min. She was shifted to intensive care unit where her Hb had decreased to 3 g/dl. She was resuscitated with intravenous fluids and packed red blood cells. Once stabilized, emergency contrast-enhanced CT was done, which revealed 11 cm renal AML with bleeder fistulating into pelvicalyceal system. Emergency angiogram and transarterial selective embolization was done [Figures 1-3]. Postprocedure, the patient’s recovery was uneventful and she was discharged home after 1 week. On follow-up after 1 month, she is doing well, and on ultrasound, AML had interval decrease in size.

Abstract

Wunderlich syndrome is a rarely entity. We report our case of a 60-year-old female, who presented in the emergency medicine department with acute right flank pain, tender mass right upper quadrant abdomen, hypotension, and visible hematuria. Urgent computerized tomogram confirmed bleeding in the right renal angiomyolipoma. Selective angioembolization was done. The patient recovered and was sent home after 1 week. On follow-up after 1 month, she is doing fine, and on ultrasound, AML had interval decrease in size.

Keywords: Angiomyolipoma, renal hemorrhage, spontaneous

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DISCUSSION

Spontaneous renal hemorrhage was first reported by Bonet in 1679[1] and later by Wunderlich in 1856.[2] This syndrome is associated with various conditions including renal tumors, vascular diseases, coagulation disorders, and idiopathic causes.[3] It can present with acute abdomen or with Lenk’s triad of acute flank pain, abdominal mass, and hypotension.[4] Contrast-enhanced CT is considered the gold standard for diagnosis and can provide the detailed description of hemorrhage as well.[5] Angiography can finally confirm the bleeding and can do simultaneous selective embolization of the bleeding vessel.[6] AML is the most common benign renal tumor causing WS, especially when AML size is larger than 4 cm. AML is sporadic in 80% or in 20% secondary to genetic mutations in TSC1 and TSC2 genes causing TS.[7] WS is very rare and can be fatal, as was seen in our case which was managed in time, to save life. If interventional radiology services for embolization is not available, emergency nephrectomy is the only other option.[3,5]

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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