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CASE REPORT

Ruptured Renal Angiomyolipoma Presenting with Isolated Flank Pain: A Case Report

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ABSTRACT

Introduction: Ruptured renal Angiomyolipoma (AML) is a life-threatening condition that may mimic other diagnoses in the clinical presentation, and if not diagnosed promptly, it can lead to life-threatening bleeding and shock.

Case report: In this paper, we report the case of a 56-year-old previously healthy Saudi man who presented with isolated left flank pain. The patient was initially treated as a case of renal colic, possibly caused by renal stones. However, the imaging revealed the clinical picture to be caused by a ruptured renal angiomyolipoma (AML) of the left kidney and successfully treated with angiographic embolization by an interventional radiologist.

Conclusion: Despite its rare occurrence, rupture of the renal AML may mimic other diagnoses and lead to life-threatening hemorrhagic shock. Prompt diagnosis and management can improve outcomes. By reporting this case, we want to highlight the importance of considering alternative differential diagnoses and including all the substantial life-threatening conditions while dealing with patients presenting with isolated flank pain, even with no other risk factor.

KEYWORDS: Ruptured renal Angiomyolipoma, Isolated flank pain, Emergency medicine, Heamorrhagic Shock, Renal Angiomyolipoma.

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INTRODUCTION

Angiomyolipoma (AML) is the most common benign mesenchymal tumor of the kidney. They are hormone-sensitive; therefore, they are more prevalent in females and tend to grow during pregnancy (1-3). Ruptured renal AML is a rare but serious complication and a life-threatening differential diagnosis of acute flank pain (4).

Less than half of the symptomatic patients with ruptured renal AML show manifestations of retroperitoneal bleed, and only half of those patients present with acute flank pain, but due to its rare occurrence, ruptured renal AML can be easily misdiagnosed as simple renal colic in patients presenting to the ED.

Despite limitations, when combined with appropriate clinical data, imaging modalities can provide valuable real-time diagnostic workup for abdominopelvic emergencies like ruptured renal AML. Maintaining a high suspicion index and considering a complete differential diagnosis can prevent devastating outcomes in such unexpected cases when evaluating patients.

The following case report presents a case of a patient with an acute onset of isolated left flank pain that was initially misdiagnosed as renal colic. CT scans later revealed the underlying pathology of ruptured Renal Angiomyolipoma managed by angiographic embolization.

CASE REPORT

A 57-year-old Saudi male patient complained of acute-onset left flank pain in the emergency department (ED). Her pain began at rest, reached maximum intensity within minutes, and was described as "dull aching pain." The patient denied fevers, nausea, vomiting, dysuria, and hematuria.

The patient had a similar episode a few months ago, and the pain was relieved with painkillers. His medical history was notable for an appendectomy. Initial vital signs in triage included: blood pressure of 127/53, heart rate of 73, respiratory rate of 16, oxygen saturation of 100% on room air, and a temperature of 36.0°C.

An abdominal examination revealed localized tenderness in the left lumbar area with a mildly distended abdomen. Otherwise, the reminder systemic physical examination was unremarkable.

Initial laboratory investigations were unremarkable and revealed a white blood cell count of 10.6 K/uL, hemoglobin 12.6 g/dl, PT 12 s, APTT 26.9 s, and INR 1.1. The urine studies and chest x-ray were unremarkable.

A non-contrast computed tomography of the Abdomen and pelvis without contrast was ordered for the renal colic workup. It showed a left renal exophytic fat-containing lesion with adjunct hyper-density, likely representing a left renal exophytic fat-containing lesion ruptured Angiomyolipoma with an adjunct hematoma. The left ureter could not be assessed. The right kidney and ureter were unremarkable, with a trace amount of abdominal fluid and a small left pleural effusion. Figure1

Although the patient remained vitally stable during his stay in the ED and there was no significant drop in his hemoglobin level, blood type and screen was ordered, and he was urgently referred to the urology and interventional radiology on call. Meanwhile, the patient was admitted to the intensive care unit and kept NPO under close monitoring of his vitals and hemoglobin level with four units of PRBC on standby.

CT abdomen and pelvis with contrast were arranged for further evaluation. It revealed multiple renal Angio-Myo-Lipomas, with the largest one located in the mid-pole with adjunct surrounding high-density fluid likely representing contained hematoma secondary to underlying rupture with no evidence of extravasation of the contrast or active bleeder. Figure 2

Despite the slow drop in the hemoglobin level, the patient did not require a blood transfusion.

On the following day, angiographic embolization was successfully performed by the interventional radiologist; after reviewing the patient's overall condition.

After reviewing the patient's condition, interventional radiology successfully performed angiographic embolization the next day. Afterward, the patient was transferred to the surgical ward under the urology service. The patient stayed hemodynamically stable during the postoperative period and was later discharged home for close follow-up with the out-patient urology clinic.

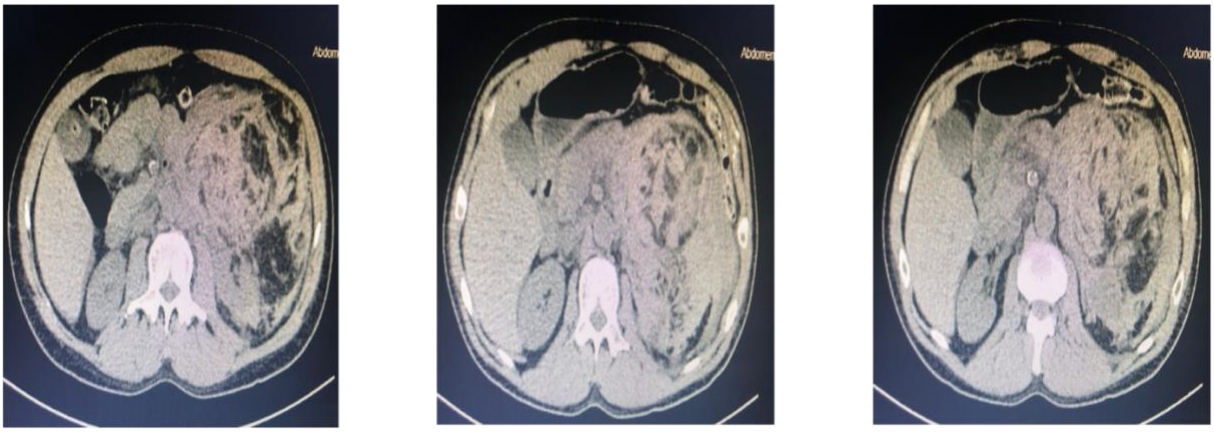

Figure 1: CT scan of the Abdomen and Pelvis without contrast showing left renal exophytic fat containing lesion with adjunct hyper density representing ruptured Angiomyolipoma with adjunct hematoma.

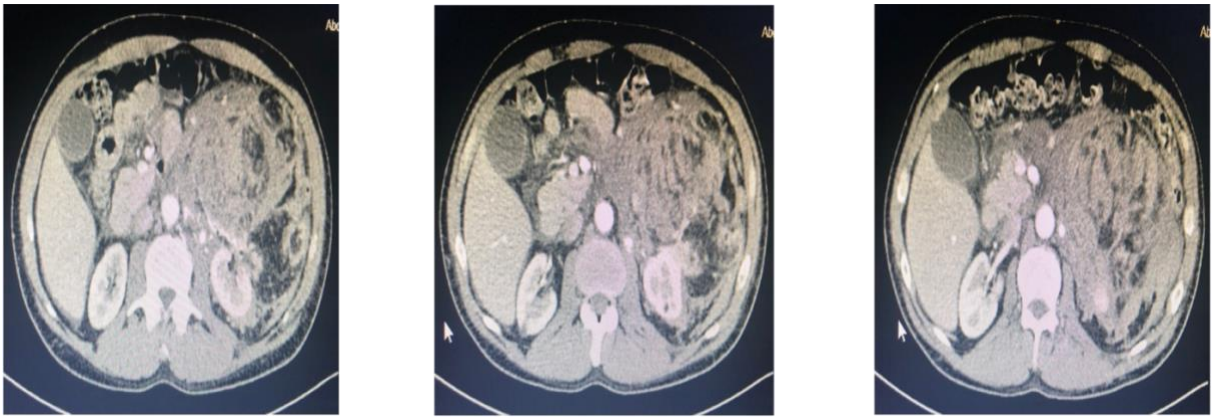

Figure 2: CT scan of the Abdomen and Pelvis with contrast shows left kidney with multiple variable sized heterogenous masses representing multiple renal Angiomyolipoma with adjunct surrounding high density fluid representing contained hematoma.

DISCUSSION

Acute flank pain is a frequently encountered clinical presentation in the emergency department (ED), and it can mimic any abdominopelvic condition.

Although evidence suggests that most patients who present to ED with acute flank pain have ureterolithiasis, typical presentations of severe pathology may share many symptoms with more benign causes of flank pain (1), such as ruptured renal Angiomyolipoma.

Renal AML is known to be a relatively rare benign neoplasm. The incidence of AML is estimated to be 0.13% in the general population, and it is more prevalent in women than in men (2,3).

As in our case, most patients present later in life, during their fifth or sixth decade (4).

It is histologically composed of perivascular epithelioid cells, smooth muscle, and fat tissue (2). Due to their fat content, AMLs are generally readily identifiable on computerized tomography and magnetic resonance imaging (3).

Even though ultrasound cannot define AML, renal AML will appear as a hyperechoic renal lesion with acoustic shadowing (5).

Renal AML occurs as isolated sporadic entities in 80% of cases; the remaining 20% of AMLs develop in association with tuberosclerosis complex (TSC) or pulmonary lymphangioleiomyomatosis (LAM) (4,6).

While 90% of these neoplasms are asymptomatic and found incidentally by imaging, symptomatic presentation is seen in less than 15% of cases and is most frequently related to spontaneous retroperitoneal hemorrhage (6).

Previous studies have indicated that renal AMLs larger than 4 cm have a significantly higher risk of rupture than others, reaching up to 60%. However, rupture can occur at smaller sizes, and larger AMLs may remain stable (7,8).

Acute bleeding of renal AML can manifest in a classic triad known as Lenk's triad, consisting of flank pain (53%), a palpable tender mass (47%), and gross hematuria (23%). When accompanied by hypovolemic shock, it is considered a urological emergency, known as Wunderlich syndrome (9,10).

Wunderlich syndrome is a life-threatening condition that refers to spontaneous nontraumatic bleeding confined to the perinephric space (10).

However, unusual presentations have also been reported in the literature. Renal angiomyolipomas are interesting tumors with diverse potential presentations.

Although Lenk's triad has been reported in less than half of patients diagnosed with ruptured renal AML, flank pain with renal AML on the same side was highly associated with tumor rupture and hemorrhage (11).

In our case, the patient presented with isolated acute flank pain and remained hemodynamically stable throughout his stay in the hospital.

Fortunately, the imaging performed for other indications guided us to the proper diagnosis before any catastrophic deterioration.

If presented with a similar scenario, there are higher chances of missing a life-threatening condition in another clinical setting.

Ruptured renal AML poses a challenging diagnosis, and this case reinforces the reminder to consider it among the other differential diagnoses when evaluating flank pain, haematuria, palpable tender mass, or any abdominopelvic complaint.

CONCLUSION

Emergency medicine physicians should maintain a high suspicion of ruptured renal Angiomyolipoma in patients with isolated flank pain.

Despite its rare occurrence, rupture of the renal AML may mimic other diagnoses and lead to life-threatening hemorrhagic shock. Prompt diagnosis and management can improve outcomes. The present case confirms the importance of considering a complete differential diagnosis when evaluating ED patients and reminds physicians to consider other life-threatening differential diagnoses of an acute flank pain patient with no risk factors.

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COMPETING INTERESTS

The author declares no competing interests with this case.

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PATIENT'S CONSENT

Written informed consent was obtained from the patient for the publication of this case report.

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