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Pathology

# A Giant Renal Angiomyolipoma: A Case Report

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**Abstract** | **Case Report**

***Background***: The most common solid benign renal tumors are angiomyolipomas (AMLs). They are highly vascularized and contain smooth muscle and adipose tissues. Perivascular epithelioid differentiation is characteristic for AML. Radiological imaging plays an important role for diagnosis of these tumors which usually are found incidentally, but sometimes symptoms like flank pain, gross hematuria, or severe retroperitoneal hemorrhage may also exist. ***Case Details*:** In this study, we will present a 20-year-old male patient who came to the emergency department with right sided abdominal pain for one day duration of sudden onset, associated with generalized weakness, vomiting and dysuria. The patient was diagnosed to have a huge right renal angiomyolipoma (sized 18 x 13.5 x 32 cm), compressing the liver, crossing the midline, and reaching below right iliac crest. The management was by right nephrectomy in our Prince Hussien Urology Center. The follow-up period was for 6 weeks in our urology clinics. ***Conclusion*:** In our case report the angiomyolipoma was larger than 30 cm. So, awaring about the presentation of huge angiomyolipoma and its complications is mandatory due to the rarity of these cases (> 20 cm). Therefore, in the future we can deal with them in a good manner.
**Keywords:** Angiomyolipoma, Nephrectomy, Angioembolization.


## INTRODUCTION

In clinical practice the most common benign renal tumors are Angiomyolipomas (AMLs). These tumors contain three components: 1- dysmorphic blood vessels, 2- smooth muscles, and 3- mature adipose tissues [1].

There are three types of AMLs: 1- classic type, 2- fat poor AML, and 3- epithelioid AML which may be fatal. Besides that, depending on the ratios of the three components of AMLs, the presentation of AMLs on radiological images could not be distinguished from renal cell carcinoma (RCC) [2].

The usual size of renal AMLs the was reported in most studies is 4 cm in maximum diameter, but when their size becomes more than 10 cm, they are called giant AMLs [3].

When AMLs become > 4 cm the risk of bleeding due enlarging and rupturing of aneurysms increases which may lead to life threatening hemorrhage, beside that the compressing symptoms may become more obvious [4]. So, in our case report we review the clinical course, diagnosis and the management option of the giant renal AML.

## PATIENT AND OBSERVATION

**Patient Information**:

In February 2022, a 20-year-old male patient came to the emergency department at Jordanian royal medical services with severe right sided abdominal pain of sudden onset for one day duration and associated with generalized weakness, recurrent vomiting, nausea, and dysuria for one month.

**Medical Psycho-Social History**: free

**Medical Family History**: his mother has diabetes milletus; his father has hypertension and chronic kidney disease.

**Surgical History**: free

**Clinical Findings:**

On physical examination the patient was stable (vital signs normal), looked pale, his abdomen was tense with voluntary guarding and mild tenderness.

**Timeline of Current Episode:**

The right flank pain started from 3 months ago then the pain disappeared, and the patients felt recurrent episodes of generalized crampy abdominal pain before one month and associated with recurrent vomiting, nausea, and generalized weakness, then the pain increased in intensity the day before presentation to the emergency room.

**Diagnostic Assessment:**

Laboratory blood and urine tests were as follows: (complete blood count (CBC): hemoglobin: 8.4 g\dl, Full chemistry was normal and urine analysis had microscopic hematuria: 6-8 RBCs). Then the patient underwent abdominal sonography and abdominal CT with contrast and was diagnosed to have huge right renal angiomyolipoma (sized 18 x 13.5 x 32 cm) compressing the liver and crossing the midline and reaching below right iliac crest (figures 1-3).

**Figure 1: Extension of the AML to the liver**

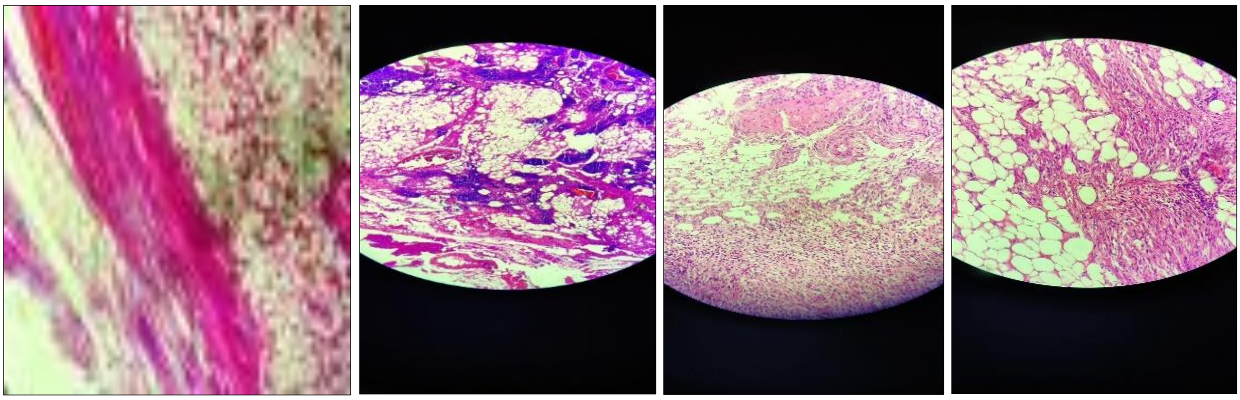
**Figure 2: The mid portion of the AML**

**Figure 3: The extension of the AML to the iliac crest**

**Diagnosis:** Giant right renal angiomyolipoma.

**Therapeutic Intervention**:

The diseased right kidney was managed by open right nephrectomy in our Prince Hussien Urology Center after the preoperative management of the patient by (intravenous fluids, 2 units of blood, intravenous antibiotics). Postoperatively, the patient was given supportive treatment (intravenous fluid, antibiotics, and pain killers). The Postoperatively, report (gross description and microscopic examination) revealed that the diagnosis is angiomyolipoma with hemorrhagic mass, (5) reactive lymph nodes and the thin cortex kidney with its components measuring 24 x 16 x 32 cm (figures 4-8).



**Figures 4, 5, 6, 7: The microscopic appearance of the AML components**

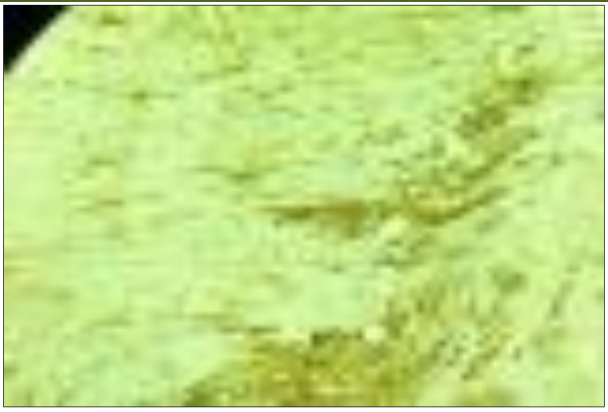


**Figure 8: HMB45 Immuno-histochemical stain is positive in tumor cells**

**Follow-Up and Outcomes**:

Then the patient was discharged after 5 days of hospitalization and followed for 6 weeks of follow-up period then every 6 months to look for the function of the other kidney in our urology center clinics.

**Patient Perspective:**

The patient was satisfied from the procedures were done in the emergency room, surgical department pre and post operatively, the good decision that was taken to remove the affected kidney without any delay, the high quality of surgery (experienced urologists who did the surgery), and from the follow-up protocols in our clinics.

**Informed Consent:**

Prince Hussein Urology Center in Jordanian Royal Medical Services is considered educational hospital (has residency programs and medical students), which means the patients receiving medical care within this hospital agreed indirectly for the use of their data in studies. So, the data was obtained from urology center registry system and despite that, the ethical committee approval was gained from our Royal Medical Services institution for publication of this study.

## Discussion

Renal AMLs are benign tumors (called hamartomas due to its varying composition) and usually treated by conservative management or radiofrequency ablation when they are asymptomatic and < 4 cm in size. But when they grow to larger sizes and cause compression symptoms and risk of bleeding like Wunderlich's syndrome (lower back pain, hematuria, and shock due to retroperitoneal bleeding), the preferred management is surgical excision or angioembolization regarding the clinical situation of the patient [5]. In our case due to the emergency presentation of the patient, huge angiomyolipoma (> 20 cm), and the eligibility for general anesthesia the preferred method was surgical excision.

Giant renal AMLs could be sporadic as in our case or within the context of Tuberous Sclerosis Complex (TSC) [6].

Due to the morbidity and mortality of giant AMLs (invasion of adjacent renal parenchyma and causing renal impairment and therefore could lead to end stage renal disease) and the suspicion of malignancy, surgical excision is preferred [7]. Like in our case.

Bilateral giant AMLs were reported and raised the tuberous sclerosis occurrence. Also, the management option for these cases to preserve the renal functions gives priority for renal angioembolization or partial nephrectomies more than total surgical excision [8]. In the contrary of our case.

## Conclusion

Giant AMLs are carrying a high risk of bleeding, and the compressing symptoms especially alimentary system lead the patients for seeking of medical attention, therefore the surgical management by total or partial nephrectomy even the angioembolization regarding the case is preferred than other management options to save the patients before the serious complications occur.

**Competing Interests:** the authors declare no competing interest.

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