HSPI The Open Access Publisher

ACR ARCHIVES OF CASE REPORTS
ISSN 2637-3793

Case Report

A Life-threatening Case of Giant Bilateral Renal Angiomyolipoma: A Case Report

Mehdi Marrak*

Urology Department, Military Hospital, Tunis, Tunisia

Abstract

Renal angiomyolipoma (AML) is a rare tumor with an incidence of 0.3% - 3%.

We reported a case of a 41-year-old male patient who presented with gross hematuria and hemorrhagic shock, due to a right giant angiomyolipoma he underwent urgent right nephrectomy by subcostal laparotomy, total weight of the mass was 6 Kg, histological examination concluded in a renal angiomyolipoma.

Treatment of renal AML depends on the clinical presentation, tumor size, and single or multiple lesions: single small (< 4 cm) asymptomatic lesions require only clinical and radiological follow-up, however giant symptomatic (hematuria), life-threatening masses require urgent multidisciplinary treatment and especially surgery.

Giant renal bilateral AML is very rare, conservative treatment in the absence of hemorrhage should always be first proposed to preserve renal function as possible.

More Information

***Address for correspondence:** Mehdi Marrak, Urology Department, Military Hospital, Tunis, Tunisia, Email: mahdimarrag59@gmail.com

Submitted: June 06, 2024
Approved: June 17, 2024
Published: June 18, 2024

How to cite this article: Marrak M. A Life-threatening Case of Giant Bilateral Renal Angiomyolipoma: A Case Report. Arch Case Rep. 2024; 8: 060-062.

DOI: 10.29328/journal.acr.1001097

Copyright license: © 2024 Marrak M. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Keywords: Urology; Case report; Renal neoplasms

Abbreviations: AML: Angiomyolipoma; TSC: Tuberous Sclerosis Complex; LAM: Lymphangioleiomyomatosis

Check for updates

OPEN ACCESS

Introduction

Renal Angiomyolipoma (AML) is a rare tumor with an incidence of 0.3% - 3% [1]. Some 80% of renal angiomyolipomas are sporadic and the remaining 20% are associated with tuberous sclerosis complex [2].

It is the most common benign renal tumor and is defined by its 3 histological components: blood vessels, smooth muscle, and adipose tissue [3]. Giant AMLs associated with Tuberous Sclerosis Complex (TSC) represent a treatment challenge and are life-threatening [3].

Case report

We reported a case of a 41-year-old male patient with TSC and epilepsy, suffering from chronic right lower back pain, who presented with gross hematuria and hemorrhagic shock. After stabilization with vasoactive drugs and a massive transfusion of red blood cell units, a CT scan showed bilateral giant renal AMLs, a right renal hematoma of 4cm, and multiple intra-tumoral aneurysms (Figure 1). Hemodynamic status was stable, hemoglobin level was 7 g/dl. The patient was admitted to the intensive care unit of the urology department and blood pressure, pulse, and hemoglobin levels were monitored. On the next day of hospitalization, the haemoglobin level dropped to 5 g/dl with hypotension and tachycardia.

Angioembolization was not available because of the lack of a coil so an urgent right nephrectomy by subcostal laparotomy was performed. Intraoperative blood loss needed a transfusion of 3 red blood cell units.

The total weight of the mass was 6 kg (Figure 2).

The postoperative course was normal, the patient left the intensive care department after 3 days and the hospital after 7 days.

A histological examination of the mass revealed an angiomyolipoma (Figure 3).

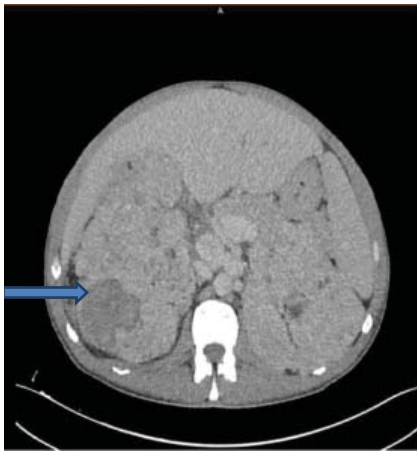

Figure 1: Bilateral giant renal AMLs + a 4 cm right renal hematoma.

Figure 2: A 6 kg right kidney.

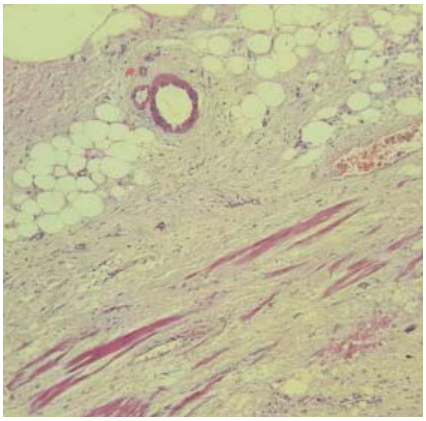

Figure 3: Histological examination showing blood vessels, smooth muscle, and adipose tissue. Hematoxylin-eosin stain and x 20 magnification.

The follow-up over 2 years showed stability of the left angiomyolipoma on ultrasound and did not show any recurrence of hematuria.

Discussion

Affecting generally middle-aged women with an incidence of 0.3% in the normal population [4], AMLs of the kidneys remain usually asymptomatic, and symptoms like gross hematuria, and low back pain usually reveal masses > 4 cm.

Three types of AML were described in the literature: Sporadic, associated with tuberous sclerosis complex which is the case of our patient, and those that are associated with lung lymphangioleiomyomatosis (LAM) [4].

Some 80% of renal angiomyolipomas are sporadic and the remaining 20% are associated with tuberous sclerosis complex [2].

AML is the only benign renal tumor that can be diagnosed by CT scan: The presence of fat, confirmed by a negative attenuation value of -25 HU or less in CT, within a renal lesion is considered the diagnostic hallmark [5].

AMLs associated with TSC have generally bigger sizes and so more symptomatic than sporadic ones.

Treatment of renal AML depends on the clinical presentation, tumor size, and single or multiple lesions: single small (< 4 cm) asymptomatic lesions require only clinical and radiological follow-up, however giant symptomatic (hematuria), life-threatening masses require urgent multidisciplinary treatment and especially surgery [6].

Bilateral giant AML which is a benign lesion poses a therapeutic challenge: treatment principles are the resolution of symptoms and prevention of complications such as hemorrhage without comprising renal function.

These principles can be more and more respected due to the development of radiologic embolization and the ability of surgeons to perform partial nephrectomy [6].

Conclusion

Giant renal bilateral AML is very rare, conservative treatment in the absence of hemorrhage should always be first proposed to preserve renal function as possible.

Statement of ethics

Ethical approval is not required for this study in accordance with local or national guidelines.

Written informed consent was obtained from the patient for publication of the details of his medical case and any accompanying images.

Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Acknowledgement

We are grateful to all the staff of the Urology and Pathology Department of our hospital who contributed to this project.

Methods

The work has been reported in line with the SCARE criteria.

Agha RA, Franchi T, Sohrab C, Mathew G, Kirwan A, Thomas A, et al. The SCARE 2020 guideline:

Updating consensus Surgical Case Report (SCARE) guidelines. International Journal of Surgery. 2020; 84(1): 226-30.

References

1. Ghaed MA, Daniali M, Motaghi P, Sohi HJ. Huge renal epithelioid angiomyolipoma - A case report of a giant, benign renal mass. Int J Surg Case Rep. 2020; 66:374-378. doi: 10.1016/j.ijscr.2019.12.032. Epub 2019 Dec 28. PMID: 31945544; PMCID: PMC6965190.
2. Flum AS, Hamoui N, Said MA, Yang XJ, Casalino DD, McGuire BB, Perry KT, Nadler RB. Update on the Diagnosis and Management of Renal Angiomyolipoma. J Urol. 2016 Apr; 195(4 Pt 1):834-46. doi: 10.1016/j.juro.2015.07.126. Epub 2015 Nov 21. PMID: 26612197.
3. Fragkoulis C, Stasinopoulos K, Theocharis G, Stathouros G, Papadopoulos G,

Kostopoulou A, Bethany-Michaelides M, Ntoumas K. A rare case of giant renal angiomyolipoma in a woman with tuberous sclerosis. Urol Case Rep. 2018 Jun 18; 20:41-42. doi: 10.1016/j.eucr.2018.06.006. PMID: 29942788; PMCID: PMC6011037.

4. Hatano T, Egawa S. Renal angiomyolipoma with tuberous sclerosis complex: How it differs from sporadic angiomyolipoma in both management and care. Asian J Surg. 2020 Oct;43(10):967-972. doi: 10.1016/j.asjsur.2019.12.008. Epub 2020 Jan 17. PMID: 31959574.

5. Ryan MJ, Francis IR, Cohan RH, Davenport MS, Weizer A, Hafez K, Kunju LP. Imaging appearance of renal epithelioid angiomyolipomas. J Comput Assist Tomogr. 2013 Nov-Dec;37(6):957-61. doi: 10.1097/RCT.0b013e3182a77674. PMID: 24270119.

6. Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. J Urol. 2002 Oct;168(4 Pt 1):1315-25. doi: 10.1016/S0022-5347(05)64440-0. PMID: 12352384.