Journal of the
Bahrain Medical Society

Sharaf M, J Bahrain Med Soc. 2022;34(1):66-70

CASE REPORT

Giant Renal Angiomyolipoma: A Case Report

Mahmoud Sharaf[1*], Deyari Henari[2], Veena Negarat[3], Hani Alfadhel[4], Abdulla Darwish[5], Khalid Ahmed[6]

[1]Urology Department of Bahrain Defence Force Hospital
[2]Resident Department of Radiology, Bahrain Defence Force Hospital
[3]Chief Resident, Department of Clinical Pathology, Bahrain Defence Force Hospital
[4]Consultant, Department of Radiology, Bahrain Defence Force Hospital
[5]Consultant, Department of Clinical Pathology, Bahrain Defence Force Hospital
[6]Consultant, Department of Urology, Bahrain Defence Force Hospital

Corresponding author:
Dr. Mahmoud Sharaf, Senior Resident, Department of Urology, Bahrain Defence Force Hospital;
E-mail: sharafmahmoud81@gmail.com

Received date: March 19, 2021; **Accepted date:** September 06, 2021; **Published date:** March 31, 2022

Abstract

Renal Angiomyolipoma (AML), moreover known as a renal hamartoma, is a solid tumor with no malignant characteristics. The inheritance pattern of renal AML is autosomal dominant. If the lesion grows to a large size, a series of clinical manifestations and important complications might also occur. This research paper presents a case of huge renal AML in a 47-year-old lady, who visited the Emergency Department of Bahrain Defence Forces Hospital with right-sided abdominal pain of sudden onset. The patient underwent a total right nephrectomy. The resected mass was sized 10.5 x 13 x 14 cm. Postoperative histopathological examination confirmed the lesion as a huge renal AML. Due to the huge size of the tumor, it is crucial to record similar cases, along with their diagnosis and treatment.

Keywords: Abdominal pain, Angiomyolipoma, Hamartoma, Kidney Neoplasms, Nephrectomy

Introduction

Renal Angiomyolipoma (AML) is an unprecedented benign renal neoplasm including mature adipose tissue, thick-walled blood vessels, and smooth muscle fibers in distinct proportions. AML is likewise known as a renal hamartoma. Previous research stated that the renal AML may be also developed through four cm every year in its most dimension, while renal AMLs develop to a length of >10 cm, they're known as 'massive' AMLs.[1,2]

Due to the hemorrhagic aneurysms that expand in the enlarging AML, the hazard of bleeding from rupture and the prevalence of compression signs of close-by systems increase. Giant renal AML is uncommonly cited in the literature.[3]

We herein file the clinical course, prognosis and remedy of a patient with a massive renal AML.

Case Presentation

In November 2019, a 47-year-old female patient without any past medical illnesses, no known allergies, and no active problems was admitted through the Emergency department and presented with right flank pain of sudden onset increasing gradually and radiating to involve the whole abdomen with no history of increasing body temperature and no history of gross Haematuria.

On examination, the patient looked ill, vital signs were as follows, blood pressure was 110/70 mmHg, pulse was 90 bpm, and temperature was 37ºc with generalized abdominal tenderness especially the

right flank with hemoglobin (Hb) 9.6 mg/dL. The patient underwent initial abdominal Ultrasound scan (USS) which revealed a heterogenous hyperechoic lesion arising from the lower pole of the right kidney with perinephric extension and extended inferiorly to the right iliac fossa and measured approximately 13 x 14cm (Figure 1).

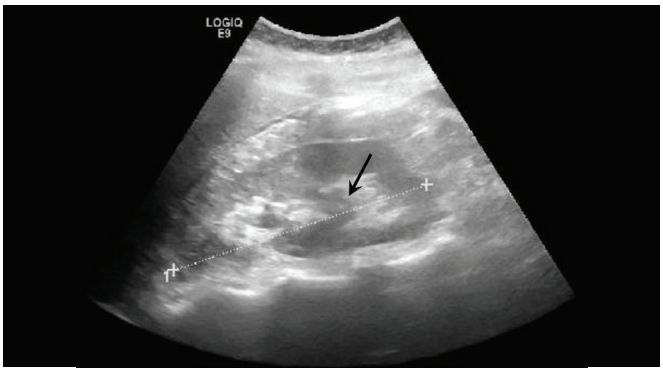

Figure 1: Grey-scale image of the right kidney, longitudinal view reveals a heterogenous hyperechoic lesion involving the lower pole with perinephric extension

The patient underwent Computed tomography (CT) study without and with intravenous contrast that confirmed the presence of a large well defined mixed fat and solid attenuated lesion measuring approximately 10.5 x 13 x 17 cm which was arising from the right kidney with evidence of peri-renal collection and extravasation of contrast. The mass effect of the lesion was appreciated, crossing the midline, with a displacement of major abdominal vessels, right ureter and bowel to the left side of the abdomen. No internal or peripheral calcification was seen. Features were consistent with a large angiomyolipoma with acute hemorrhage within the mass and extended retroperitoneally (Figure 2).

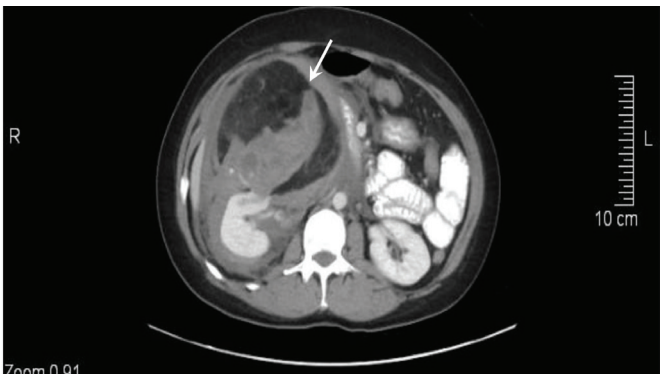

Figure 2: Axial delayed CT for abdomen shows the consequent mass effect, the mixed fat-solid lesion displacing the important abdominal vessel, right ureter, and bowel towards the left side of the abdomen

There have been no symptoms or radiological findings suggesting tuberous sclerosis complex (TSC). The adrenal glands bilaterally and the left kidney seemed to be normal. Furthermore, the results of blood biochemistry checks and coagulation profile have been in the normal range.

The patient was kept nothing by mouth or nil per os (NPO), on intravenous (IV) fluids and IV Tazocin and underwent right renal selective Angioembolization. The right renal angiogram was performed via the right common femoral artery (CFA) access using 5F. A sheath that delineated the right Kidney's homogenous enhancement with aberrant artery is arising from the lower renal branch. The aberrant artery was very tortuous and appeared to be descending inferiorly around the hematoma, likely, representing the AML feeding vessel. The attempt to cannulate this feeding vessel failed owing to its tortuosity and angled origin. The feeding vessel showed sluggish flow most likely due to spasm. No active blush was demonstrated. No embolization was performed (Figure 3).

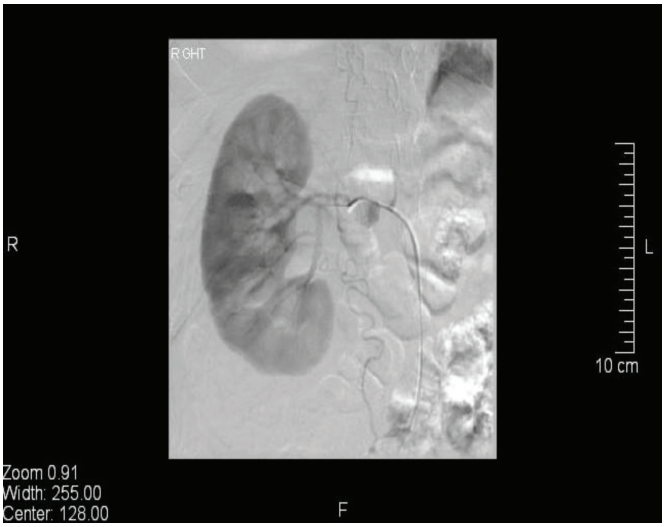

Figure 3: Right Renal Angiogram performed via right CFA access using 5F. The Sheath demonstrated a very tortuous artery arising from the inferior segment of the right Renal Artery

The patient's clinical condition was worsening blood pressure was 100/60, pulse was 110 bpm and temperature 37ºc. The patient appeared unwell with decreasing Hb reaching 7 mg/dL and increasing abdominal pain and tenderness, and so, was prepared to undergo right sided open nephrectomy after right sided renal angioembolization of the main right renal artery on 22nd of November 2019.

The patient received a total of 4 packed red blood cells (PRBCs) units during her hospital stay. Her clinical condition improved. She went home in a good condition on 28th of November 2019. On the date of the last follow-up 13/01/2020, the patient remained disease-free, and was feeling much better.

Macroscopic examination showed right kidney measuring 14 X 4 X 4 cm with an attached 5 cm long ureter with minimal perinephric fat. The lower pole showed 21 X 15 X 14 cm a large brownish lobular mass which was seen extending from the right kidney. The whole specimen weighed 1907 gm and the mass weighed 1740 gm.

On cutting, the mass measured 17 X 14 cm with a large area of hematoma at the medial aspect measuring 9.5 X 5.5 cm with mild extension to the peritumoral area.

The histopathological microscopic examination revealed that the tumor was largely composed of mature adipose tissue containing many large, thick-walled blood vessels throughout and smooth muscle fibers in different proportions with a large area of intramural and the peritumoral hemorrhage was seen (Figure 4).

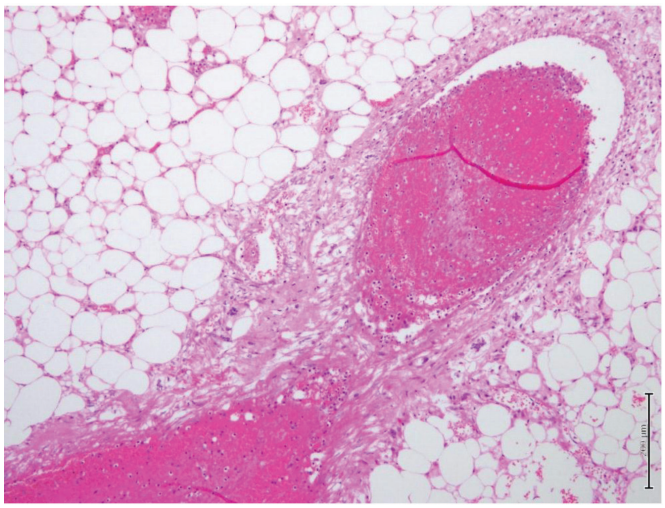

Figure 4: High power view of the tumor (AML) displaying medium-size blood vessels full of blood and surrounded by smooth muscle fibers with mature adipose tissue by Hematoxylin and Eosin staining (H&E stain)

Immunohistochemical staining of the tumor revealed that the tumor cells had been positive for human melanoma black-45 (HMB-45) and negative for Pankeratin, smooth muscle actin (SMA), desmin, S-100 and CD 10. Based on those findings, the patient was diagnosed with a huge renal AML which was resected completely, and the included kidney, ureter, and perinephric fat had been unremarkable.

Contrary to other benign renal masses, the diagnosis of AML can be made on imaging. The presence of macroscopic fat on CT or magnetic resonance imaging (MRI) is diagnostic of AML. Ultrasonography showed, hyperechoic masses, like some renal cell carcinomas (RCCs), making Ultrasound less reliable in the diagnosis. On CT the presence of intralesional fat (−15 to −20 Hounsfield Units [HU]) on non-enhanced series is diagnostic.[4]

Although the diagnosis may be confidently made on imaging in most circumstances, fat-poor AML (which resembles RCC), fat-containing RCC, and liposarcoma are unique situations in which the diagnosis can be problematic on imaging. Approximately 4% to 14% of AML do not contain radiographically identifiable fat and appear similar to RCC on standard imaging (Hyperdense on non-contrast imaging and enhancing on contrasted series). In this setting, MRI may be helpful because lesions appear hyperintense on T1 sequences with subsequent hypointensity on fat suppression and appear hypo intense on T2 because of the preponderance of smooth muscle.[5]

When fat-poor AML is considered, percutaneous biopsy has been useful in determining the diagnosis. Fat-containing RCC is a rare situation; however, when described, intralesional calcification is common, although not universally, described, a finding not routinely seen in AML.[6]

Finally, very large AMLs may have the appearance of retroperitoneal liposarcomas, with the key difference relating to the impact on the renal parenchyma. As AML arises from the parenchyma, an indentation in the parenchyma can be observed from this site of origination, whereas the liposarcomas begin in the retroperitoneum and subsequently envelop and compress the renal parenchyma.[7] The authors declare that the patient consent was obtained for treatment.

Discussion

Renal AML is a benign neoplasm arising from mesenchymal elements and was first reported in

1951.[6] AML is also referred to as 'Hamartoma' due to its varying composition, including adipose tissue, smooth muscle fibers, and blood vessels.[8]

These tumors are usually divided into two types, namely in the context of TSC or as sporadic; the case discussed herein belongs to the latter category.[1]

It was previously reported that renal AML may grow by 4 cm each year in its maximum dimension, when the diameter of renal AML reaches >10 cm, the tumor is referred to as 'giant'.[1,2]

Reports of giant renal AML in the literature are uncommon, with the largest renal AML (39 x 25 x 9 cm) reported in 2013 by Taneja.[9]

This giant tumor is rich in the thick-walled blood vessels that are prone to rupture and bleeding. Due to the hemorrhagic aneurysms that develop within the enlarging AML, the incidence of compression symptoms of nearby structures and the risk of bleeding from rupture increase.[10]

Lower back pain, hematuria, and shock are the most important clinical manifestations suggesting the occurrence of retroperitoneal bleeding, referred to as Wunderlich's syndrome. These clinical manifestations are the main reason for renal AML patients seeking medical attention.

The majority of tumors sized <4 cm are asymptomatic and patients may be managed conservatively or treated by radiofrequency ablation.[11,12]

As the tumor grows, compression of the gastrointestinal tract may cause alimentary symptoms; in addition, the presence of a mass in the abdominal area may be easily confirmed by palpation. The mass appears to be echogenic on ultrasound and has the density of fat on CT scanning.

When the diameter of renal AML becomes >4 cm, particularly for giant tumors with persistent hemorrhage or suspicion of malignancy, partial or a total nephrectomy is the treatment of choice.[13,14]

In the present case, total right nephrectomy was performed.

Histopathological examination of the renal AML revealed that the main component was normal/mature adipose tissue.[12] HMB-45 positivity on Immunohistochemical staining may be used to distinguish renal AML from other renal tumors.[15]

To summarize, in light of the above-mentioned analysis, it can be stated that giant renal AML may be identified by imaging and the diagnosis is confirmed by histological and immunohistochemical examination. As this tumor is associated with a high risk of hemorrhage, partial or total nephrectomy, rather than conservative treatment, is the treatment of choice in order to reduce the risk of bleeding and relieve the compression symptoms caused by the giant mass.

Financial support and Sponsorship

Nil

Conflict of Interest

No.

References

1. Ewalt DH, Sheffield E, Sparagana SP, *et al*. Renal lesion growth in children with tuberous sclerosis complex. *J Urol*. 1998; 160:141-145.
2. Seyam RM, Bissada NK, Kattan SA, *et al*. Changing trends in presentation, diagnosis and management of renal angiomyolipoma: Comparison of sporadic and tuberous sclerosis complex-associated forms. *Urology*. 2008; 72:1077-1082.
3. Serrano Frago P, Del Agua Arias Camisón C, Gil Sanz MJ, *et al*. Controversies related to epithelioid variant of renal angiomyolipoma: A review of the literature. Urology. 2006; 67:846. e3-e5.
4. Morgan GS, Straumfjord JV and Hall EJ. Angiomyolipoma of the kidney. *J Urol*. 1951; 65:525-527.
5. Steiner MS, Goldman SM, Fishman EK, *et al*. The natural history of renal angiomyolipoma. *J Urol*. 1993; 150(6): 1782– 1786.
6. Hayn MH, Cannon GM Jr, Bastacky S, *et al*. Renal cell carcinoma containing fat without associated calcifications: two case reports and review of literature. Urology. 2009; 73(2):443 [e445-447].
7. Wang LJ, Wong YC, Chen CJ, *et al*. Computerized tomography characteristics

that differentiate angiomyolipomas from liposarcomas in the perinephric space. *J Urol*. 2002; 167(2 Pt. 1):490–493.

8. Kulkarni B, Desai SB, Dave B, *et al.* Renal angiomyolipomas-a study of 18 cases. *Indian J Pathol Microbiol*. 2015; 48:459-463.
9. Taneja R and Singh DV. Giant renal angiomyolipoma: Unusual cause of huge abdominal mass. *J Clin Imaging Sci*. 2013; 3:56.
10. De Luca S, Terrone C and Rossetti SR. Management of renal angiomyolipoma: A report of 53 cases. *BJU Int*. 1999; 83:215-218.
11. Bora A, Soni A, Sainani N, *et al.* Emergency embolization of a bleeding renal angiomyolipoma using polyvinyl alcohol particles. *Diagn Interv Radiol.* 2007; 13: 213-216.
12. Gregory SM, Anderson CJ and Patel U. Radiofrequency ablation of large renal angiomyolipoma: Median-term follow-up. *Cardiovasc Intervent Radiol*. 2013; 36:682-689.
13. Faddegon S and So A. Treatment of angiomyolipoma at a tertiary care centre: The decision between surgery and angioembolization. *Can Urol Assoc J*. 2011; 5:E138-E141.
14. Zhou Y, Tang Y, Tang J, *et al.* Total nephrectomy with nephron-sparing surgery for a giant bilateral renal angiomyolipoma: A case report. *Oncol Lett*. 2015; 10:2450-2452.
15. Vitaly M, Surena FM and Christopher GW. Campbell-Walsh, Urology. Benign renal tumors. 10th edition. Philadelphia, Saunders Elsevier. 2012 pp1498-pp501.