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Case Report

Metachronous Giant Renal Angiomyolipoma: About One Case

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SUMMARY

Giant bilateral renal angiomyolipomas are exceedingly uncommon. Although the majority of renal angiomyolipomas are asymptomatic, symptoms may occur.

Hypovolemic shock can ensue in the event of a hemorrhagic rupture, the most lethal angiomyolipoma complication.

Tumors larger than 10 cm referred to as “giant” AMLs are exceptional. Only a few reported cases had giant tumours measuring more than 20 cm.

AML mortality and morbidity are correlated with the risk of bleeding and lesion invasion into adjacent normal renal parenchyma, which can progress to chronic kidney disease and potentially end-stage renal disease.

It is crucial for clinicians to recognize that when renal angiomyolipoma occurs on one side of a patient’s kidney, it is equally wise to pay attention to the opposite side of the kidney, because renal angiomyolipoma might reoccur.

We present a case of a very exceptional giant bilateral renal angiomyolipoma that required bilateral nephrectomy and hemodialysis.

Keywords: Bilateral nephrectomy; Giant angiomyolipoma ; Anephric patient

INTRODUCTION

Angiomyolipoma (AML) is the most common benign renal tumor diagnosed in clinical practice. It is composed of a mix of dysmorphic blood vessels, smooth muscle components, and mature adipose tissue (1).

AML can occur sporadically or may be associated with tuberous sclerosis complex or sporadic lung lymphangioleiomyomatosis (2).

The incidence rate of AML is about 0.3 % - 3.0%. Tumors more than 10 cm (referred to as “giant” AMLs) are rare. Only a few reported cases had giant tumours measuring more than 20 cm (3).

AML can range from asymptomatic to a hypovolemic shock caused by a hemorrhagic rupture, which is the most dangerous complication.

We present a case of a very exceptional giant bilateral renal angiomyolipoma that required bilateral nephrectomy and hemodialysis.

OBSERVATION

Patient A.E., 34 years old, has been in terminal chronic renal failure since 2015, receiving hemodialysis twice per week, and had a right nephrectomy in 2015 due to the rupture of a right renal angiomyolipoma.

Since 2015, the patient had no imaging of the left kidney.

The symptom began 8 months ago with the installation of a left lower back pain with complicated progressive abdominal distension, followed 10 days later by a total hematuria requiring transfusion in per-dialysis. The physical examination revealed abdominal distension as well as left lumbar tenderness.

Renal ultrasound revealed a heterogeneous ultrasound mass that had developed at the left renal lodge and had spread to the left iliac fossa. The Abdominal Computed Tomography (CT) revealed a left retroperitoneal mass with a hypervascularized renal appearance, which was locally advanced with a moderate amount of peritoneal effusion.

For a better characterization of the mass, a Computed Tomography Angiography (CTA) was indicated, which revealed a voluminous peritoneal and retroperitoneal mass occupying the entire left flank extended to the pelvis, heterodense poorly limited, measuring approximately 19 x 13 extended by 32 cm and seating three contingents: tissue, fat, and fluid, with integrity of the left renal pedicle probably related to a renal angiomyolipoma (Figure 1, 2). Because of the size of the mass, which put the patient at danger of bleeding and confusion regarding the tumor’s histological type, a left nephrectomy was performed. The post-operative care was straightforward. Angiomyolipoma of the left kidney was confirmed by the anatomopathological examination (Figure 3).

DISCUSSION

Angiomyolipoma is a rare benign tumor that affects only 0.3 % of the general population (4). Angiomyolipoma is typically unilateral; bilateral angiomyolipome is uncommon, accounting for only 15% of cases. (5).

In most cases, angiomyolipoma is asymptomatic. However, abdominal pain, hematuria, palpable mass, or other complications such as retroperitoneal hemorrhage may occur (6). According to

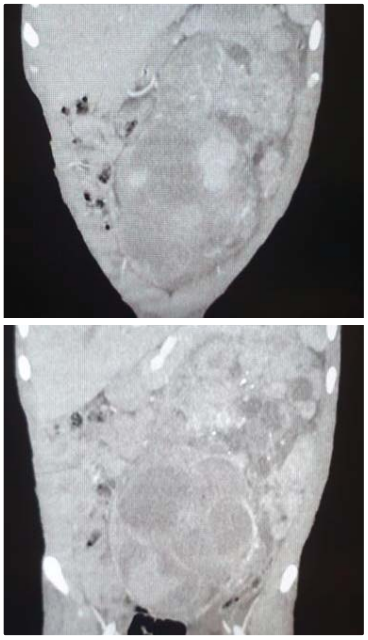

Figure 1: CT scan showing huge heterogeneous masses with fatty content (empty arrow) in left kidneys in accordance with giant AML.

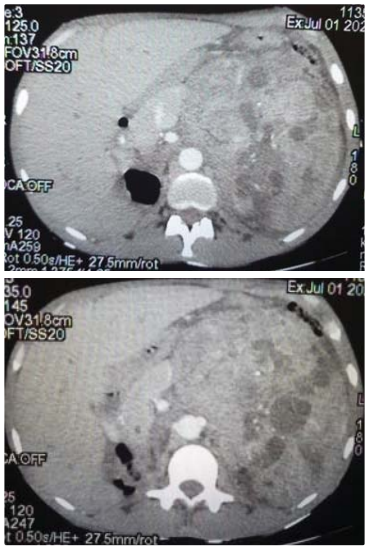

Figure 2: An axial section of a CT scan demonstrating a left renal angiomyolipoma.

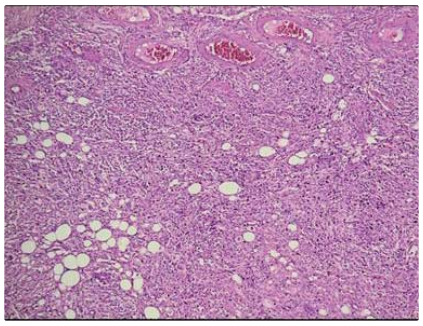

Figure 3: Adipose, fusocellular, and vascular quotas in renal angiomyolipoma.

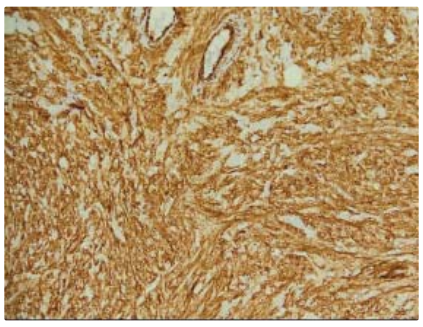

Figure 4: Spindle cell cluster diffuse expression of renal angiomyolipoma.

research, the risk of bleeding and the frequency of symptoms are related to the size of the tumor (greater than 4 cm) and the presence of tuberous sclerosis (6,7).

This is consistent with the case of our patient, who had a 19 x 13 extended by 32 cm. Patients with bilateral angiomyolipoma are generally at risk for high blood pressure and kidney failure (8). Proteinuria is an unfavorable occurrence, and further investigation is required (9).

Contrary to our case, the patient had a complicated unilateral form with significant bleeding, indicating a nephrectomy, and then developed another giant angiomyolipoma on the contralateral side complicated of abundance hematuria.

Ultrasound can be used to determine whether a tumor is solid or cystic. The CT scan is the reference exam; it allows the differentiation of the various components: vascular, adipose, and tissue (muscle) based on density. However, its use for monitoring is limited due to the accumulation of radiation and in cases of kidney function impairment (10).

On the other hand, MRI does not emit radiation, so it can accurately characterize lesions. Its spatial resolution is lower than that of the CT, whose speed prevents abdominal movement artifacts. It is adequate for determining the size of angiomyolipomas and other kidney lesions.

There are no current statistics or solid reasons to biopsy adipose angiomyolipomas. Intra-tumor calcifications, solitary adipose angiomyolipomas, or any uncertain MRI image are the most common indications (11).

In our case, a biopsy was not required. The CT provided enough informations to make the diagnosis. The primary treatment is surgery. Surgical excision is recommended for symptomatic tumors that are large and have a higher risk of spontaneous bleeding (12,13).

Furthermore, some authors discuss the lack of a clearly established relationship between tumor size and the risk of bleeding. The famous 4cm threshold recommended for deciding on a curative treatment and that it should no longer be used in isolation; other factors such as age, growth rate, and patient preferences must now be considered (14).

Surgical treatment is not recommended as a first-line therapy for AML. The indications for surgical treatment include suspicion of malignancy, symptoms, and a risk of hemorrhage (2).

AML mortality and morbidity are correlated with the risk of bleeding and lesion invasion into adjacent normal renal parenchyma, which can progress to chronic kidney disease and potentially end-stage renal disease (15).

The indication for nephrectomy was largely posed in our case to avoid severe and recurring bleeding.

CONCLUSION

The treatment of giant renal angiomyolipoma is complex and challanging.

Options ranging from *surveillance* to bilateral nephrectomy. To select an effective and safe therapeutic approach, a thorough assessment of patient symptoms, tumor size, and kidney function must be performed.

It is crucial for clinicians to recognize that when renal angiomyolipoma occurs on one side of a patient's kidney, it is equally wise to pay attention to the opposite side of the kidney, because renal angiomyolipoma might reoccur.

REFERENCES

1. 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.

2. 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.

3. 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.

4. Redkar N, Patil MA, Dhakate T, Kolhe P. Tuberous sclerosis complex presenting as bilateral large renal angiomyolipomas. BMJ Case Rep. 2012 Aug 8;2012:bcr2012006412. doi: 10.1136/bcr-2012-006412. PMID: 22878994; PMCID: PMC4543694.

5. Ziadie M. Kidneytumor-cysts, children, adultbenignBenign (usually) adult tumors Angiomyolipoma. PathologyOutlines.com. 2015.

6. Shankar S, Hulikanthimatt KS, Awatti SM, Narayanrao ST. Bilateral multifocal renalangiomyolipoma associatedwithWunderlich's syndrome in a tuberoussclerosis patient. J Clin DiagnRes. 2015;9(8):ED01. doi: 10.7860/JCDR/2015/13349.6289

7. Wright T, Sooriakumaran P. Renal angiomyolipoma presenting with massive retroperitoneal haemorrhage due to deranged clotting factors: a case report. Cases J. 2008 Oct 4;1(1):213. doi: 10.1186/1757-1626-1-213. PMID: 18834543; PMCID: PMC2567302.

8. Eijkemans MJ, van der Wal W, Reijnders LJ, Roes KC, van Waalwijk van Doorn-Khosrovani SB, Pelletier C, Magestro M, Zonnenberg B. Long-term Follow-up Assessing Renal Angiomyolipoma Treatment Patterns, Morbidity, and Mortality: An Observational Study in Tuberous Sclerosis Complex Patients in the Netherlands. Am J Kidney Dis. 2015 Oct;66(4):638-45. doi: 10.1053/j.ajkd.2015.05.016. Epub 2015 Jul 10. PMID: 26165440.

9. Pirson Y. Tuberous sclerosis complex-associated kidney angiomyolipoma: from contemplation to action. Nephrol Dial Transplant. 2013 Jul;28(7):1680-5. doi: 10.1093/ndt/gft009. Epub 2013 Feb 14. PMID: 23413089.

10. Valentı´nLo´pez B, Blasco Amaro JA. Plan of appropriate use of imaging diagnosis technologies in abdominal pathology in primary care and specialities. Madrid, Spain. 2010 : Unidad de Evaluacio´n de Tecnologı´asSanitarias, AgenciaLaı´nEntralgo.

11. Chung MS, Choi HJ, Kim MH, Cho KS. Comparison of T2-weighted MRI with and without fat suppression for differentiating renal angiomyolipomas without visible fat from other renal tumors. AJR Am J Roentgenol. 2014 Apr;202(4):765-71. doi: 10.2214/AJR.13.11058. PMID: 24660704.

12. Murphy DP, Glazier DB, Chenven ES, Principato R, Diamond SM. Extrarenal retroperitoneal angiomyolipoma: nonoperative management. J Urol. 2000 Jan;163(1):234-5. doi: 10.1016/s0022-5347(05)68015-9. PMID: 10604357.

13. Tseng CA, Pan YS, Su YC, Wu DC, Jan CM, Wang WM. Extrarenal retroperitoneal angiomyolipoma: case report and review of the literature. Abdom Imaging. 2004 Nov-Dec;29(6):721-3. doi: 10.1007/s00261-004-0179-2. Epub 2004 Jun 8. PMID: 15185030.

14. Oesterling JE, Fishman EK, Goldman SM, Marshall FF. The management of renal angiomyolipoma. J Urol. 1986 Jun;135(6):1121-4. doi: 10.1016/s0022-5347(17)46013-7. PMID: 3520013.

15. Afriansyah A, Yusuf AM, Nusaly H. Bilateral Giant Renal Angiomyolipoma in a Patient with Tuberous Sclerosis Complex: A Case Report. Acta Med Indones. 2018 Jan;50(1):61-65. PMID: 29686177.