Giant renal angiomyolipoma in a solitary kidney

Ericka Sohlberg, MD, Andrew Sun, MD, Rustin Massoudi, MD, Kris Prado, MD, Eila Skinner, MD
Department of Urology, Stanford University, Stanford, California, USA

SOHLBERG E, SUN A, MASSOUDI R, PRADO K, SKINNER E. Giant renal angiomyolipoma in a solitary kidney. *Can J Urol* 2018;25(6):9614-9616.

While renal angiomyolipomas (AMLs) generally remain small and asymptomatic, larger AMLs are more common in tuberous sclerosis patients. Giant AMLs over 20 cm are a rare entity and little is known about their management. We present a unique case of a 48-year-old woman with tuberous sclerosis and a 39 cm AML arising from a solitary kidney, after undergoing nephrectomy for a prior AML. Giant renal AMLs can occur in patients with tuberous sclerosis and resection should be considered even for large tumors. Renal sparing is often difficult and patients should be counseled about potential need for postoperative hemodialysis.

Key Words: AML, giant angiomyolipoma, tuberous sclerosis

Introduction

Renal angiomyolipomas (AML) are benign tumors consisting of poorly organized blood vessels, smooth muscle and adipose tissue. While rare in the general population, AMLs are more common in patients with tuberous sclerosis (TS). An estimated 30%-80% of tuberous sclerosis patients will develop AMLs during their lifetime.[1] AMLs are generally managed conservatively due to their benign nature and as lesions less than 4 cm are usually asymptomatic.[2] However, AMLs in tuberous sclerosis patients have been noted to be larger, faster growing, and more often multifocal compared to their sporadic counterparts.[3] Isolated cases of giant AMLs in these patients have been reported but there are limited descriptions of these large angiomyolipomas with no consensus on management. This case report provides an example of this rare entity.

Accepted for publication October 2018

Address correspondence to Dr. Ericka Sohlberg, 300 Pasteur Drive, Grant Building S285, Stanford, CA 94305 USA

Case report

A 48-year-old woman with a history of TS and bilateral renal and hepatic AMLs was admitted to the medicine service for abdominal pain and inability to tolerate oral intake. Other than her known AMLs, her only other manifestations of TS were pulmonary lymphangioleiomyomatoses. She had previously undergone a right nephrectomy with AML resection 15 years prior to admission, and later had multiple angioembolizations of her left renal and hepatic AMLs without success. More recently, she had been trialed on both Everolimus and Rapamycin with persistent growth of both hepatic and renal AMLs.

On admission, she was found to have a tender, asymmetric, and very distended abdomen with a palpable mass arising from the left side. A computed tomography scan of the abdomen and pelvis without contrast revealed a giant left-sided AML measuring 30 cm x 16 cm x 20 cm that surrounded and heavily involved the left kidney, Figure 1. The AML exerted a mass effect on her GI tract, likely leading to her abdominal pain, nausea and inability to tolerate oral intake. After discussion with the patient about her options and need for dialysis after removal, she was taken to the operating

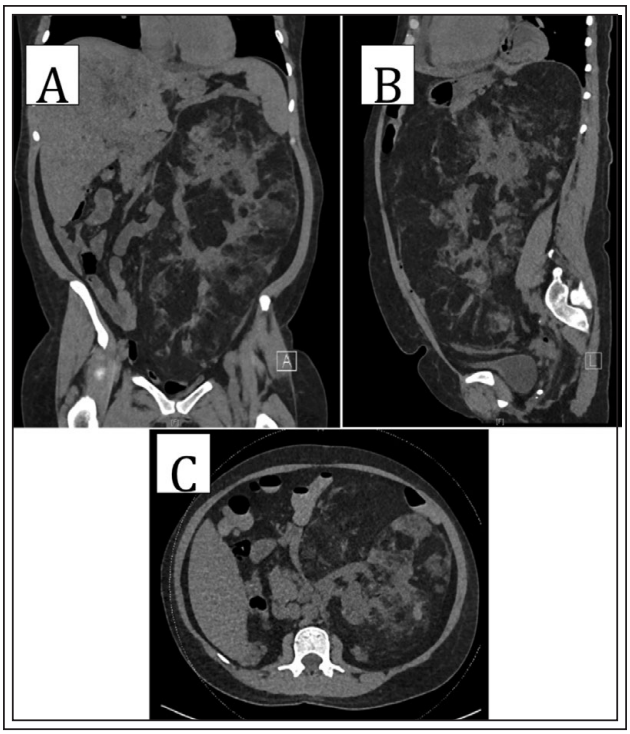

Figure 1. CT images in coronal (panel **A**), sagittal (panel **B**), and transverse (panel **C**) planes demonstrating a large left renal mass.

room and through an open, midline approach, left nephrectomy with AML resection was performed.

The kidney and AML was removed en bloc, measuring 39 cm x 18 cm x 14 cm in total. The total weight was 3.7 kg, Figure 2. Pathology review confirmed the diagnosis of AML. She recovered well after the procedure and there were no postoperative complications. She was started on hemodialysis, which she tolerated well, and was discharged home on postoperative day 7.

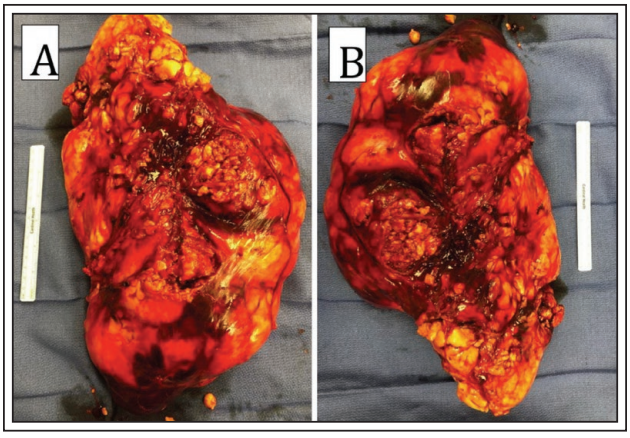

Figure 2. Gross specimen on medial (panel **A**) and lateral (panel **B**) views.

Discussion

Giant renal AMLs are a rare occurrence, with few similarly large tumors documented in the literature. Fragkoulis et al reported a case of a 30 cm renal AML weighing 7200 g (fragmented),[4] while Katz et al reported a length of 45 cm and weight of 3500 g.[5] Our case similarly reports a length of 39 cm and weight of 3700 g. The largest known renal AML was documented by Taneja et al, weighing 7500 g.[6] The majority of these cases occurred in tuberous sclerosis patients, supporting known data that tuberous sclerosis patients are at risk for larger and faster-growing AMLs.[2,3] This potential for rapid growth is critical to consider when planning surveillance for tuberous sclerosis patients.

It is important to recognize the entity of giant AML in order to prevent misdiagnosis. Large AMLs can mimic Wilms tumors[7] or retroperitoneal sarcomas,[8] leading to vastly different treatment implications. The acknowledgement of giant AMLs both facilitates accurate diagnosis and suggests more cautious management of tuberous sclerosis patients with AML.

While the typical renal AML is small and asymptomatic, the rarer giant AML poses its own set of complications. The main risk of a large AML is retroperitoneal hemorrhage, as the risk of bleeding has been shown to increase with the size of an AML.[9] Hemorrhage is thought to occur due to dysmorphic vessels that may develop aneurysms and can rupture. Giant AMLs additionally carry the risk of impingement on adjacent organs. In our patient, the sheer size of the tumor had resulted in bowel compression with food intolerance requiring TPN dependence. In this case, the extreme nature of this compressive effect ultimately prompted surgical resection.

Management of these giant lesions is challenging due to limited data. Similar to our case, the previously documented giant AMLs described above[4,6] also required radical nephrectomy as tumor had essentially replaced the entire kidney. However, it is interesting to note that embolization has been successful in giant renal AMLs previously.[10] Neoadjuvant embolization is also an option in giant AMLs to minimize intraoperative blood loss. It is unclear if mTOR inhibition, as attempted in our patient, has successfully been used to manage giant AMLs.

Particularly in the tuberous sclerosis population where bilateral AMLs are common, treatment must be tailored to the individual patient. Our patient was not

offered resection for many years due to her solitary kidney and grew increasingly symptomatic, as she failed both embolization and mTOR inhibition. This case demonstrates that for giant AMLs, surgical resection is still a safe option after exhausting the more conservative treatments, even in the setting of a solitary kidney. □

References

1. Lendvay TS, Marshall FF. The tuberous sclerosis complex and its highly variable manifestations. *J Urol* 2003;169(5):1635-1642.
2. Steiner MS, Goldman SM, Fishman EK, Marshall FF. The natural history of renal angiomyolipoma. *J Urol* 1993;150(6):1782-1786.
3. 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(5):1077-1082.
4. Fragkoulis C, Stasinopoulos K, Theocharis G et al. A rare case of giant renal angiomyolipoma in a woman with tuberous sclerosis. *Urol Case Rep* 2018;20:41-42.
5. Katz DS, Poster RB. Massive renal angiomyolipoma in tuberous sclerosis. *Clin Imaging* 1997;21(3):200-202.
6. Taneja R, Singh DV. Giant renal angiomyolipoma: unusual cause of huge abdominal mass. *J Clin Imaging Sci* 2013;3:56.
7. Dhua A, Ranjan A, Agarwala S, Bhatnagar V, Mathur S, Devasenathipathy K. Giant renal angiomyolipoma masquerading as a Wilms tumor. *Indian J Urol* 2017;33(1):73-75.
8. Kori C, Akhtar N, Vamsidhar PN, Gupta S, Kumar V. Giant exophytic renal angiomyolipoma mimicking as retroperitoneal sarcoma; a case report with review of literature. *J Clin Diagnostic Res* 2015;9(4):XJ01-XJ02.
9. De Luca S, Terrone C, Rossetti SR. Management of renal angiomyolipoma: a report of 53 cases. *BJU Int* 1999;83(3):215-218.
10. Bishay VL, Crino PB, Wein AJ et al. Embolization of giant renal angiomyolipomas: technique and results. *J Vasc Interv Radiol* 2010; 21(1):67-72.