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Oncology

A rare case of giant renal angiomyolipoma in a woman with tuberous sclerosis

Charalampos Fragkoulis[a], Konstantinos Stasinopoulos[a], Georgios Theocharis[a], Georgios Stathouros[a], Georgios Papadopoulos[a,*], Akrivi Kostopoulou[b], Mariana Bethany-Michaelides[b], Konstantinos Ntoumas[a]

[a] Athens General Hospital "G. Gennimatas", Department of Urology, Greece
[b] Athens General Hospital "G. Gennimatas", Department of Pathology, Greece

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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] It mainly occurs sporadically affecting 0.3–3% of the general population but it is encountered quite often in patients suffering from tuberous sclerosis, Von Hippel-Lindau syndrome and neurofibromatosis type 1.[1] The aim of this case report is to present a very rare case of a giant AML in a woman treated in our department in an open surgical approach.

Case report

A 35-year old female was referred to our department with complaints of abdominal distention, constipation and palpable abdominal mass. Patient had been diagnosed during childhood with tuberous sclerosis (TS) and presented with full manifestation of the disease including facial angiofibromas, mental retardation, epilepsy and retinal lesions. Clinical examination revealed a giant palpable mass occupying the whole abdomen (Fig. 1). A contrast enhanced computed tomography was performed revealing a left renal mass with diameter of 40cm with radiological characteristics of AML (Fig. 2). Renal scan showed a 20% partial contribution of the left kidney to total renal function.

Subsequently, patient underwent left nephrectomy in an open transabdominal approach. Due to close correlation of the AML with the spleen and intraoperative hemorrhage, a splenectomy was performed simultaneously. As it was not technically feasible to extract the whole AML intact, we removed it in three pieces with the largest being 30 cm × 26 cm X 10 cm in diameter. The total weight was 7200gr (Fig. 3). Pathological report confirmed the diagnosis of AML (Fig. 4). Patient was transfused with two units of red blood cells during the operation. No complications were noted postoperatively, peritoneal drainage was removed on 5th day and the patient was discharged on 8th day with normal renal function.

Discussion

Although AML is quite rare among general population and generally remaining small and asymptomatic, it is quite often encountered in patients suffering from tuberous sclerosis. It is estimated that 26%–80% of patients diagnosed with tuberous sclerosis present with AML, while almost 80% of patients diagnosed with AML do not have TS. In addition, patients with TS present AML in younger age, with larger tumors.[1]

Clinical manifestations of AML include abdominal pain, distention and haematuria, although small masses are often asymptomatic. The most serious and dangerous complication with high mortality rates is spontaneous bleeding into the retroperitoneal space from pathological vessels, referred to as Wünderlich syndrome, straightly associated with diameter.[2]

Close monitoring is the best option for most cases of AML and indications for surgical treatment include tumor size > 4cm and presence of symptoms at the time of diagnosis.[1] If a surgical approach is decided, nephron-sparing surgery is the method of choice whenever technically

* Corresponding author. 1B, Kefallinias str, Maroussi, Athens, PC: 15126, Greece.
E-mail address: gipapadopoulos@yahoo.gr (G. Papadopoulos).

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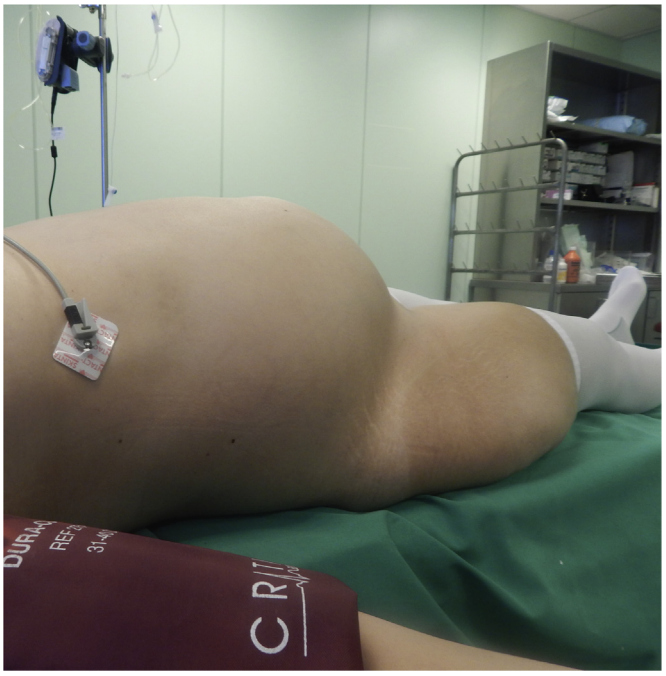

Fig. 1. Abdominal distention due to AML.

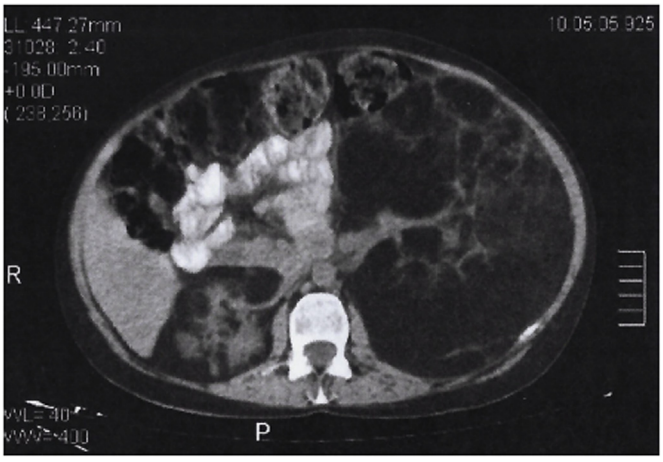

Fig. 2. AML as shown in Computed Tomography.

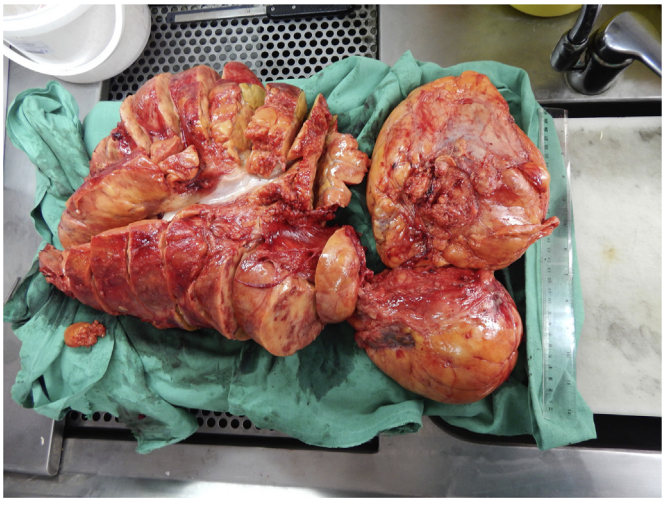

Fig. 3. The whole bulk of AML divided in 3 pieces.

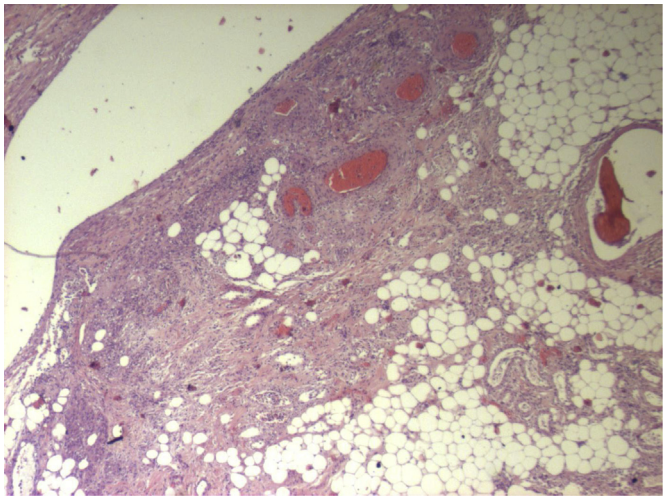

Fig. 4. AML with classic triphasic histology: an admixture of mature fat, thick walled blood vessels and smooth muscle (HE x25).

feasible. In other cases nephrectomy is performed. The decision for open approach, laparoscopic or robot-assisted surgery is based on surgeon's experience.

The open approach for nephrectomy in our patient was preferred mostly due to the massive size of the AML and the close correlation with the spleen. To our knowledge, this AML is one of the largest reported in literature and the biggest reported to be treated in a Greek hospital.[3,4] Giant AML of proximal size has been reported by Katz and Poster, where the mass measured 45 cm × 20 cm × 15 cm and weighed 3500 gr.[5] In review of the literature, the heaviest AML is the one reported by Taneja et al. with total weight of 7500 gr.[4]

Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.eucr.2018.06.006.

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