

Angiomyolipoma rupture: does size always matter?

Adrianna Wojciechowska¹, Olga Grodzka¹, Maciej Stroczyński¹, Aras Almohammad¹, Krzysztof Śmigaj¹, Bartosz Żabicki², Maciej Salagierski¹

¹University of Zielona Góra, Collegium Medicum, Department of Urology, Zielona Góra, Poland

²University of Zielona Góra, Collegium Medicum, Department of Radiology, Zielona Góra, Poland

Citation: Wojciechowska A, Grodzka O, Stroczyński M, et al. Angiomyolipoma rupture: does size always matter? Cent European J Urol. 2021; 74: 48-50.

Article history

Submitted: Jan. 21, 2021

Accepted: Feb. 6, 2021

Published online: Feb. 25, 2021

Corresponding author

Maciej Salagierski
Faculty of Medicine
and Health Sciences
University Hospital
Urology Department
26 Zyty Street
65-046 Zielona Góra,
Poland
m.salagierski@cm.uz.
zgora.pl

Angiomyolipoma (AML) is the most common benign renal neoplasm. Although asymptomatic lesions rarely lead to clinical dilemma, the management of ruptured tumours can become challenging. The size of the tumour has been widely accepted as a prognostic factor for intervention but there exists some evidence against considering the size as the only prognostic factor for intervention in AML. In our study, we described three recent cases of ruptured AML which were treated with different approaches – in two cases radical nephrectomy was performed, and in one patient a minimally invasive approach was adopted.

Key Words: angiomyolipoma ↔ retroperitoneal bleeding ↔ nephrectomy ↔ selective arterial embolization

CASE PRESENTATION

A 40-year-old female was admitted to the urology department on October 28, 2020 with symptoms of left-sided renal colic. In the interview, the patient denied suffering from chronic diseases or having undergone any surgeries in the past. She also denied smoking and drinking alcohol. The patient initially consented for ureteroscopy (URS) treatment of the left kidney and a DJ catheter was inserted. No complications occurred. However, due to persistent discomfort, a computed tomography (CT) scan of the abdominal cavity was ordered. The CT examination revealed a heterogeneous, poorly demarcated lesion with a clear fat component within the left kidney, with a maximum size of about 97 mm (Figure 1). The image of the lesion enforced the diagnosis of angiomyo-

lipoma (AML) with secondary haemorrhagic changes. The lesion pressed against and narrowed the lower half of the left kidney, causing thickening of the renal fascia to approximately 13 mm. The patient underwent surgery on November 3, 2020. Left radical nephrectomy was performed. Pathology report confirmed AML. Further hospital stay was uneventful.

A 45-year-old female was admitted to the urology department on December 22, 2019 due to a massive 13 cm tumour of the right kidney with bleeding into the retroperitoneal space. The patient reported pain in the right lumbar region, which she rated as 5 on a 10-point visual analogue scale. A 5 cm AML of the right kidney was diagnosed in 2012, but the patient refused surgical treatment and did not attend any follow-up appointment. The patient denied smoking and drinking alcohol. The patient on admission was

hemodynamically stable and she underwent right radical nephrectomy on December 24, 2019. Histopathological examination supported the diagnosis of angiomyolipoma. The patient was discharged in good general condition.

A 23-year-old female was admitted to the urology department on December 17, 2020 due to bleeding into the retroperitoneal space. In the medical history, obesity and insulin resistance was noted. The patient denied smoking or drinking alcohol. CT revealed bleeding from a 3 cm AML (Figure 2). Right superselective embolization of the bleeding upper pole renal vessel was performed. During further stay, the patient received two units of concentrated red blood cells. Due to dyspnoea in the supine position, she underwent angio-CT and chest X-ray and was consulted by cardiologist and pulmonologist. No major complication occurred and the patient was discharged in good general condition with a planned outpatient appointment within one month.

DISCUSSION

Angiomyolipoma belongs to a rare group of tumors of mesenchymal origin. Angiomyolipoma may occur sporadically or develop in the course of tuberous sclerosis. The sporadic form is the most frequent subtype of benign kidney tumours and appears more often among women, as in the presented cases. Due to their fat content, AMLs are generally easy to identify using imaging methods such as ultrasonography (US), CT or magnetic resonance imaging (MRI). The majority of them are asymptomatic. However, some can cause pain and haematuria. A major complication may be a life-threatening retroperitoneal haemorrhage, which is a serious clinical problem and requires surgical intervention, often associated with a nephrectomy. Contemporary, active surveillance is an accepted treatment for small (<4 cm), asymptomatic masses. It is established that symptomatic masses, as well as masses greater than 4 cm should be treated, though, as minimally invasive techniques unfold, other relative indications may apply. Active treatments include embolization, ablation techniques, total or partial nephrectomy. For those with tuberous sclerosis complex mTOR inhibitors may represent a viable approach [1].

In our point of view, results of AML treatment, using technologies that have been introduced lately, like cryoablation, radiofrequency ablation (RFA), microwave (MW) ablation or selective arterial embolization (SAE) are very promising. One of the first trials with laparoscopic cryoablation were performed in 2006 and the results were encouraging [2]. Retrospective reviews from Denmark (2017) showed that



Figure 1. Computed tomography scan showing poorly demarcated lesion with a clear fat component within the left kidney.

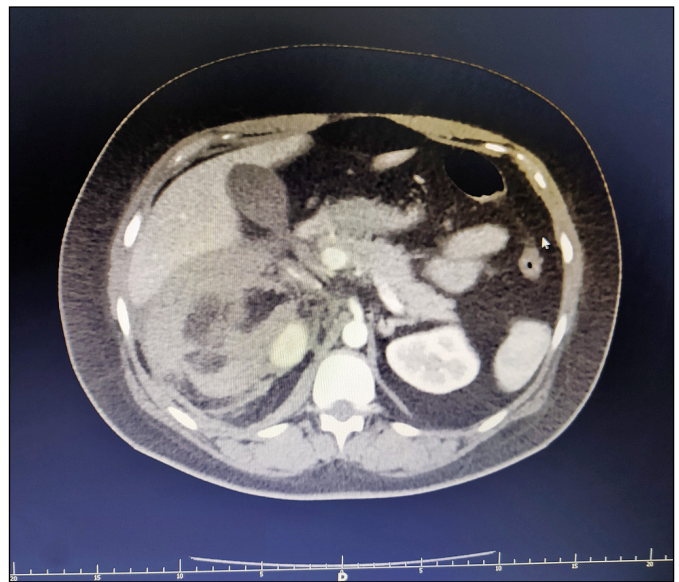


Figure 2. Computed tomography revealing bleeding to the retroperitoneal space from angiomyolipoma of the right kidney.

cryoablation is a safe and effective nephron-sparing approach and allows for treatment of even subclinical renal AMLs, minimizing the risk of potentially life-threatening hemorrhage. The complication rate is low, no patients presented with retroperitoneal hemorrhage or recurrence during follow-up [3]. Also, ablation techniques could be a good alternative. US-guided percutaneous microwave ablation demonstrates to be a safe and effective modality to devascularize and decrease the size of renal AMLs [4]. However, problems with complete ablation of some lesions in MW were recorded [5]. The study assess-

ing the feasibility of percutaneous radiofrequency ablation was also performed and showed that the tumors have become fattier with involution of the soft-tissue elements after the procedure, though no renal haemorrhage was detected [6]. Embolization is another possible approach in AML management. However, even though transarterial embolization of angiomyolipoma demonstrates low rates of mortality and serious complications, unplanned repeat embolization or surgery was required in almost 21% of cases, mainly due to revascularization [7]. Transcatheter arterial embolization (TAE) has proved to be effective procedure controlling severe hemorrhage and preservation of renal function following spontaneous ruptured renal AMLs [8]. Selective arterial embolization could be considered safe treatment option for patients with symptomatic or large AML. It represents a minimally invasive intervention with good long-term outcome. SAE may be offered as first-line treatment in most cases, though, it should be noted that the retreatment rate is high (over 41%)

[9]. In our group SAE was performed as an emergency procedure in a 23-year-old female which stopped bleeding and led to the preservation of the kidney which is of paramount importance. According to the literature, one may observe a post-embolization syndrome, meaning pain and fever after procedure and possibility of renal function impairment and developing a large artery lesion. A total of 30 studies from 2019, including 653 patients have proven that the occurrence of post-embolization syndrome is quite frequent (54%) [10].

In our study, we described three recent cases of ruptured AML. All treated patients were young females. Radical treatment was performed in tumours exceeding 10 cm. The significant bleeding from a 3 cm AML was managed with SAE. In our opinion, better AML prognostic markers to predict the chances for bleeding, especially from small AML lesions, are needed.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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