

SPONTANEOUS RUPTURE OF AN INTRATUMORAL PSEUDOANEURYSM IN A GIANT RENAL ANGIOMYOLIPOMA

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ABSTRACT:

We reported a rare case of a spontaneous rupture of an intratumoral pseudoaneurysm in a giant renal angiomyolipoma of a 52-year-old lady. The initial presentation was a sudden onset of right hypochondriac pain, nausea, and vomiting. CT scan revealed a large heterogenous exophytic enhancing mass with mixed solid and fat density within, arising from the right kidney likely representing a giant right renal angiomyolipoma. There is associated right perinephric hematoma and active bleeding within the mass. No features suggestive of tuberous sclerosis. A subsequent right renal angiogram revealed a pseudoaneurysm of an inferior segmental right renal artery and emergency embolization was done with successful obliteration of the aneurysmal sac and devascularization of the mass.

Keywords: Angiomyolipoma, Intratumoral hemorrhage, Pseudoaneurysm, Tuberous Sclerosis.

INTRODUCTION:

Angiomyolipoma (AML) is a soft tissue tumor involving the kidneys and other organs. Extrarenal angiomyolipomas are extremely rare and reported in the liver, nasal cavity, vagina, spermatic cord, skin, mediastinum, and GI tract. It is the most common benign mesenchymal neoplasm of the kidney and is composed of varying amounts of fat, smooth muscle, and abnormal thick-walled blood vessels that tend to hemorrhage [1]. AMLs are observed in 0.1–0.22% of the general population and are four times more common in females [2]. The inheritance pattern of renal AML is autosomal dominant. Two broad types have been described: sporadic isolated AML and AML associated with tuberous sclerosis. Approximately 80–90% of renal AML occurs sporadically [3]. Sporadic

isolated AML mainly occurs in women 40-70 years old while AML associated with tuberous sclerosis usually occurs at any age and in either sex. Aneurysm formation is usually noted in AMLs associated with tuberous sclerosis and is rare in the sporadic variety [4]. Most patients are asymptomatic and the tumor is often incidentally detected during ultrasonography (US) or CT [5, 6]. As many as 40% are symptomatic and these tend to be larger due to aneurysmal formation and rupture, which may cause life-threatening hemorrhage [7]. Tumor diameter, aneurysm diameter, and presence of tuberous sclerosis complex have been used as a criterion for predictors of rupture.

CASE PRESENTATION:

KZ is a 52-year-old lady with underlying obesity, diabetes, triple vessel disease, and hypertension. She presented to the emergency department with sudden onset of right hypochondriac pain associated with nausea and vomiting. Blood investigation showed rapid drop of hemoglobin from 12.2 to 4.3 g/dl during her first day of admission. Immediate resuscitation was instituted as she was in shock. The CT scan of the abdomen and pelvis revealed a large exophytic heterogenous fat-containing mass occupying the interpolar and lower pole of the right kidney with extension medially across the midline measuring 18.0 x 17.0 x 17.5 cm. There is evidence of active bleed within the mass and right perinephric hematoma. Radiological diagnosis of a large right renal angiomyolipoma with active intratumoral hemorrhage was made. She was not suitable for nephrectomy as she was a high-risk patient with multiple comorbidities. She then underwent an emergency embolization of the aneurysm of the right renal angiomyolipoma using coils and gel foam respectively, with successful obliteration of the aneurysmal sac and devascularization of the mass.



Figure 1: Abdominal x-ray showing soft tissue opacity at the right lower abdomen with loss of right psoas muscle silhouette.

DISCUSSION:

Angiomyolipomas are considered to be non-aggressive and benign. However, some AMLs may have alarming properties such as nuclear pleomorphism and mitotic activity, extension into the renal vein, vena cava, atrium, and spread to regional lymph nodes without malignant progression. While the vast majority of AML show benign biological behavior, a small proportion is malignant and may metastasize [4]. The features of malignancy include >70% atypical epithelioid cells, >2 mitotic figures per 10 high-power field (HPF), atypical mitotic figures, and necrosis [11].

The majority of tumors sized <4 cm are asymptomatic and the diagnosis of AML is often incidental. A giant AML is considered when the tumor reaches >10 cm in size. Giant renal AML is uncommonly reported in the literature with the largest renal AML (39 × 25 × 9 cm) reported in 2013 by Taneja et al [14, 15]. It was previously reported that renal AML may grow by 4 cm each year in its maximum dimension [16, 17].

Clinical presentations like a large palpable mass, lower back pain, hematuria, and shock are the



Figure 2: CT abdomen showing large heterogenous lesion containing fat measuring 18 x 17 x 17.5cm arising from the right kidney.

main reason for renal AML patients seeking medical attention. The main complication of AML is intra-capsular or retroperitoneal hemorrhage due to the tortuous, thick-walled, and angiomatous arrangement of the blood vessels in AML. The characteristic absence of elastic tissue in the tumor vessels predisposes the patient to small, saccular aneurysms formation and spontaneous hemorrhage [8,9]. Hemorrhage can be intraparenchymal, intraperitoneal, or retroperitoneal and can be life-threatening. Tuberosus sclerosis complex associated angiomyolipoma tends to be multiple, larger, with aneurysmal formation, and more likely to cause spontaneous hemorrhage than sporadic forms of angiomyolipoma [10]. In a study that examined AML tumor size, aneurysm size, and the chance of rupture, aneurysms ranged from 2–7 mm in diameter and predictors of aneurysm rupture were a tumor size of 4 cm or more and an aneurysm diameter of 5 mm or more [7]. The common method to screen AML is by ultrasound (US). The typical appearance of AML

on US is a hyperechoic renal lesion with acoustic shadowing. However, owing to the mechanism of imaging, the US cannot clearly define AMLs with minimal fat components [12]. Moreover, isoechoic or hyperechoic evidence is often displayed in both fat-poor AML and epithelioid AML. Thus, the US is not very sensitive and accurate for differential diagnosis and AML subtype identification. CT imaging should be the first choice diagnostic tool for diagnosing AML. On CT, classical AML appears as predominantly fatty attenuation with various densities, whereas fat poor AML is iso- or hyperattenuating with homogeneous enhancing. Epithelioid AML displays a hyperattenuating image with heterogeneous enhancing or multilocular cystic appearance.

Due to the tendency of progressive tumor growth and aneurysmal rupture, Oesterling et al have proposed the following treatment protocol based on size and symptoms of AML. Patients may be followed conservatively with yearly Ultrasound or CT scans for those with isolated AML < 4 cm in

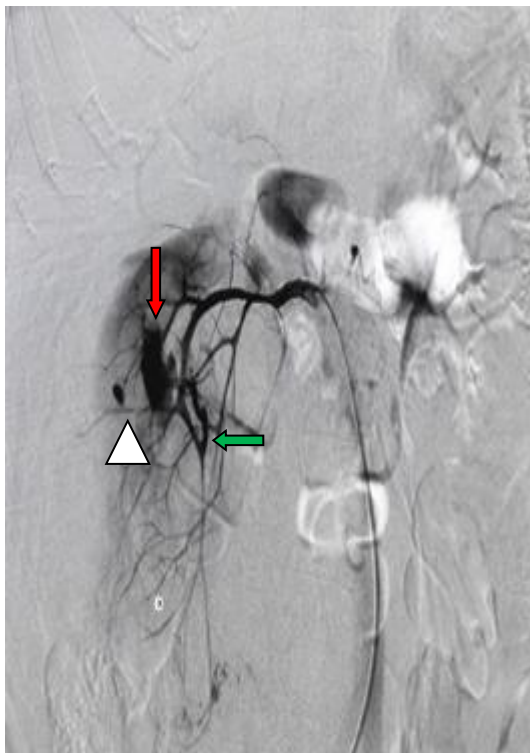


Figure 3: Selective renal artery angiogram showed evidence of contrast extravasation (red arrow) from abnormal inferior segmental artery (green arrow). Small tumor blush (black arrow) and pseudoaneurysm (arrowhead) seen from the proximal branches of inferior segmental artery supplying the inferior aspect of the mass.

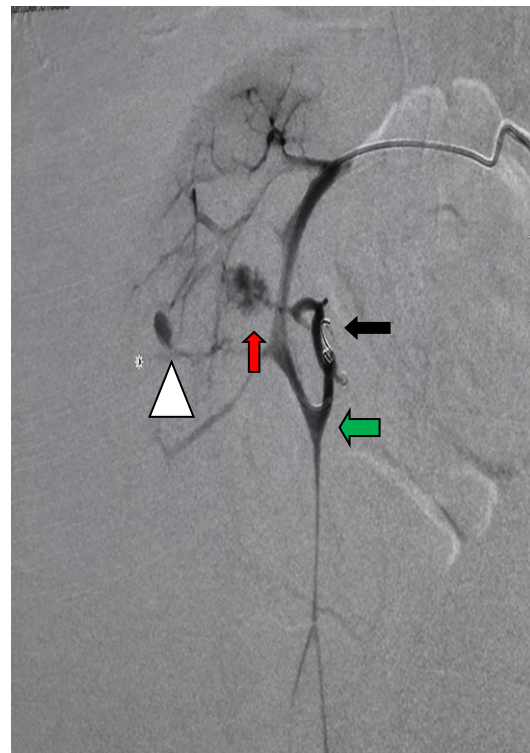


Figure 4: Angiogram following placement of first coil (black arrow) in the tortuous feeding artery of the AML. There is residual contrast extravasation (red arrow) with the persistent flow of the abnormal inferior segmental artery (green arrow) and pseudoaneurysm (arrowhead).

diameter and 6 monthly CT scans for those with lesions > 4 cm for assessment of growth. Patients with tuberous sclerosis complex and AML < 4 cm in diameter should be followed by a semiannual CT scan [13].

As a benign lesion that is usually asymptomatic, angiomyolipoma may often not require intervention [10]. Indications for intervention include suspicion of malignancy, spontaneous hemorrhage causing significant symptoms, pain, haematuria, and risk of rupture or other complications as the formation of an intrarenal aneurysm [7]. Most symptomatic angiomyolipoma can be managed by nephron-sparing approaches, including angiographic embolization or partial nephrectomy, nevertheless, some selected patients may require complete nephrectomy [10].

The occurrence of an aneurysm in sporadic AML, such in our case, is a rare phenomenon as the large majority tend to be associated with tuberous sclerosis. Our case was striking because of its large size and associated intratumoral pseudoaneurysm. A further interesting aspect of this case was the radiological appearance of the small intratumoral pseudoaneurysm that caused

extensive hemorrhage. As the patient is a high-risk patient and not suitable for operation, selective transcatheter embolization was successfully done as an emergency life-saving procedure instead of urgent nephrectomy.

CONCLUSION:

Spontaneous hemorrhage from an aneurysm in renal AML is potentially a life-threatening event. A quick assessment to verify vascularity of the mass and the presence of an aneurysm is important for preventive treatment against catastrophic bleeding.

STATEMENT OF ETHICS:

Informed consent was obtained from the patient for the publication of this work.

CONFLICTS OF INTEREST:

The authors have no potential conflicts of interest to disclose.

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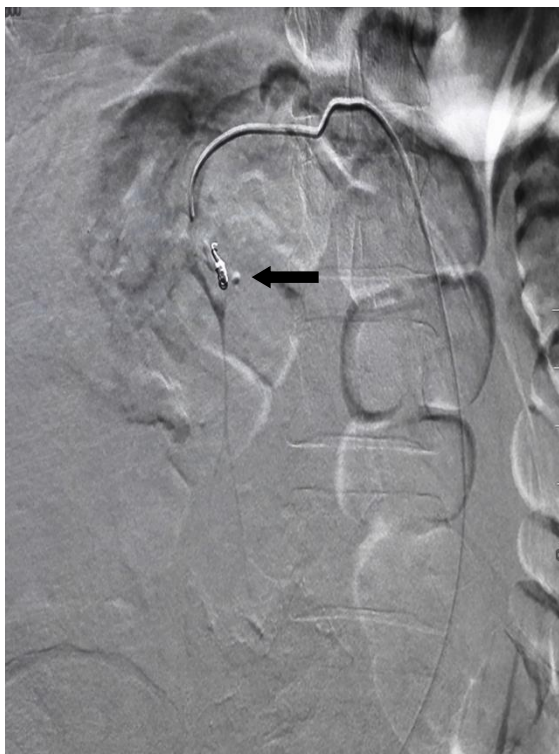


Figure 5: Second coil (black arrow) was inserted and check angiogram showed no active contrast extravasation or tumor blush.

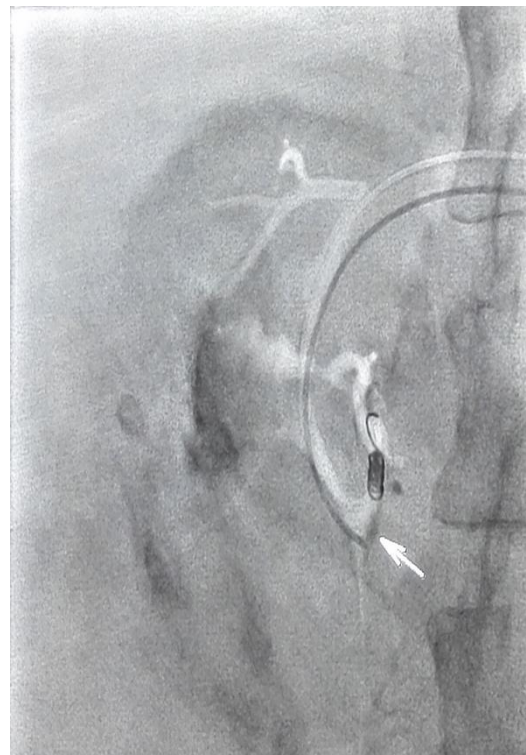


Figure 6: Coils deployed at the distal abnormal inferior segmental artery and gelfoam slurry (white arrow) infusion given proximally for added control of bleeding.

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