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Case Report

Radiology Section

Endovascular Management of Ruptured Renal Angiomyolipoma: A Case Report

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ABSTRACT

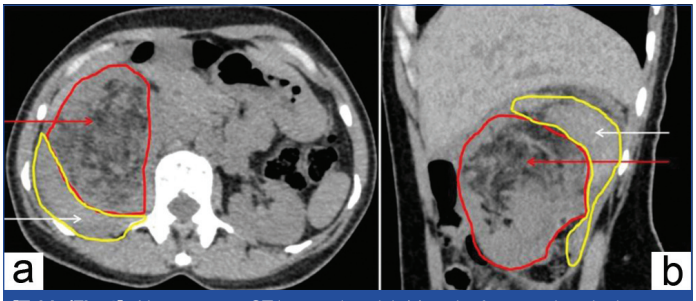
Renal angiomyolipoma is classified as a benign tumour comprising fat, smooth muscle cells, and vascular tissue. Renal giant angiomyolipoma with rupture is a relatively rare clinical emergency condition. Here, the authors present a case of a 37-year-old female patient who presented with complaints of right flank pain for six hours. The patient was anaemic and did not have haematuria. On non contrast Computed Tomography (CT) imaging, there was a right perirenal haematoma with a mixed-density, fat-containing tumour in the upper interpolar region of the right kidney. On Contrast-Enhanced Computed Tomography (CECT) imaging, the right renal tumour showed multiple abnormal tortuous vessels with moderate to intense enhancement, consistent with a large angiomyolipoma. She was managed with primary therapeutic endovascular embolisation for a ruptured right renal angiomyolipoma under local anaesthesia via a right transfemoral artery approach. The procedure was uneventful, and postembolisation syndrome was managed with medications. The patient was discharged in a stable condition. During the one-month follow-up, the patient's symptoms improved, and the need for major surgery like nephrectomy was avoided. Super selective embolisation to devascularise the tumour and preserve renal parenchyma was done under local anaesthesia via a right transfemoral approach without any complications. Postembolisation syndrome was managed medically.

Keywords: Embolisation syndrome, Nephrectomy, Perirenal haematoma

CASE REPORT

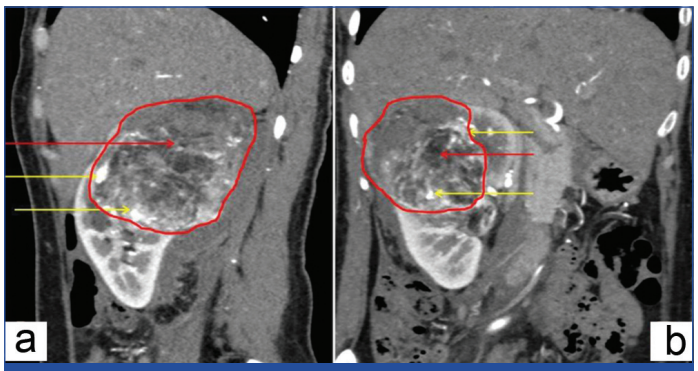
A 37-year-old woman was brought to the hospital with pain in her right flank for six hours. The pain had an acute onset and was a dull ache that worsened when walking but improved when in the supine position. There was no history of haematuria. On deep palpation, tenderness was observed in the right flank region. She had no history of diabetes, hypertension, or significant medical/surgical conditions. She was anaemic with a haemoglobin level of 9.1 g/dL, but her platelet count was normal. Her Prothrombin Time/International Normalised Ratio (PT/INR) was within the normal range (13.7 seconds/1.09). Urine examination showed no red blood cells.

Her Ultrasound (USG) evaluation revealed a large, heterogeneous, predominantly hyperechoic mass in the upper interpolar region of the right kidney. Contrast-Enhanced Computed Tomography (CECT) confirmed a ruptured angiomyolipoma measuring 10.5 cm [Table/Fig-1a,b], associated with multiple abnormal enlarged tortuous vessels [Table/ Fig-2a,b] and perirenal haematoma.



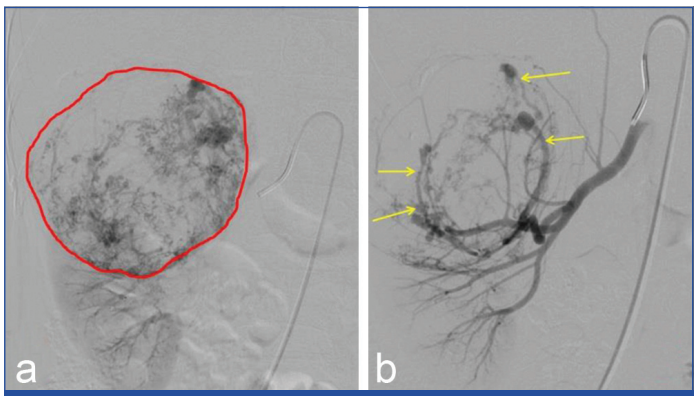
[Table/Fig-1]: Non-contrast CT images in axial: (a) and reformatted sagittal; (b) planes show fat (red arrow) containing a large tumour (red margin) in the upper interpolar region of the right kidney with retroperitoneal haematoma (white arrow and yellow margin).

She was taken for endovascular embolisation of angiomyolipoma via the right common femoral arterial approach. Right renal diagnostic angiogram with 5F Simmons 1 catheter revealed large hypervascular tumour, blush in the upper interpolar region of the right kidney with splaying of normal renal parenchyma. The arterial feeders supplying the tumour were hypertrophied and tortuous



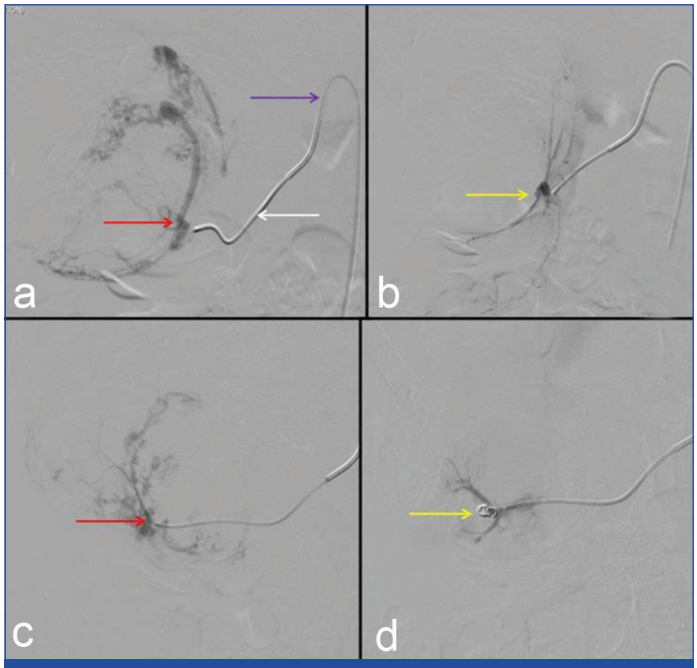
[Table/Fig-2]: Contrast-enhanced CT images in reformatted (a) Sagittal: and reformatted (b) Coronal: planes show fat (red arrows) containing the large tumour (red margin) in the upper interpolar region of the right kidney with multiple abnormal hypertrophied tortuous vessels (yellow arrows).

[Table/Fig-3a,b]. Superselective angiograms of two major arterial feeders via Progreat microcatheter confirmed the sole supply to the tumour from these abnormal arteries [Table/Fig-4a,b] and these abnormal arteries were not supplying normal renal parenchyma. These abnormal tumour arteries were super selectively embolised

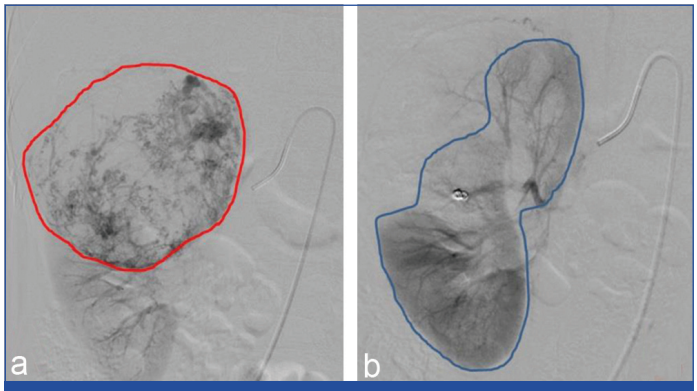


[Table/Fig-3]: Digital subtraction angiogram of the right renal artery shows hypervascular tumour blush (red margin in 'a') in the upper interpolar region of the kidney with abnormal hypertrophied branches (yellow arrows in 'b') of the renal artery supplying the tumour.

[Table/Fig-4c,d] using Polyvinyl Alcohol (PVA) particles of size 300-500 µm injected via Progreat microcatheter. This was to subserve the purpose of occluding distal tumour bed microvasculature. The proximal main stem of the abnormal artery was embolised using coils. Postembolisation angiogram showed complete devascularisation of the tumour with preservation of normal renal parenchymal arteries [Table/Fig-5a,b]. There was no procedure-related complication. Postembolisation syndrome was treated with medical management in the form of antibiotics, analgesics, anti-inflammatory and antiemetic medications and maintaining adequate hydration. Patient was discharged in stable condition. On one month follow-up, there was a significant improvement in the patient's symptoms with almost complete resolution of flank pain.



[Table/Fig-4]: Super selective (red arrow in a&c) digital subtraction angiograms with microcatheter (white arrow) through Simmons (purple arrow) diagnostic catheter show abnormal branches of the right renal artery supplying the tumour with tumour blush. Super selective (b&d) postembolisation angiograms show complete devascularisation of tumour with no opacification of intra-tumoural abnormal arteries (yellow arrow).



[Table/Fig-5]: Pre-embolisation right renal digital subtraction angiogram: (a) shows large tumour blush of angiomyolipoma in upper interpolar region of kidney (red margin). Postembolisation right renal digital subtraction angiogram; (b) shows complete devascularisation of tumour with preservation of normal renal parenchyma (blue margin).

DISCUSSION

Renal angiomyolipoma is classified as a benign tumour comprising fat, smooth muscle cells, and vascular tissue. In cases of rupture, it presents with back/flank pain, haematuria, and retroperitoneal haematoma [1]. This emergency condition can be managed by nephrectomy as the primary treatment modality. Endovascular embolisation is a minimally invasive treatment modality with equal efficacy and early patient recovery [2]. Imaging is crucial in diagnosing angiomyolipomas, as they can be difficult to distinguish from malignancy. Accurate diagnosis is important to avoid unnecessary surgery and preserve renal function [3]. Radiological classification CT and Magnetic Resonance Imaging (MRI) classifies angiomyolipomas into fat-rich, fat-poor, and fat-invisible based on the degree of fat content in the lesion [3]. In the present case, the tumour was classified as a fat-rich renal angiomyolipoma due to the macroscopic fat content revealed by CT imaging.

Symptomatic patients with angiomyolipomas usually present with flank pain and gross haematuria. A few patients can present with nausea, vomiting, or anaemia [4]. Sometimes the presentation is confusing and confounded by high blood pressure or shock [5,6]. In this case, the patient presented with flank pain without evidence of haematuria. Symptoms are more likely to be present in tumours with a size of 4 cm or greater, while patients with tumours smaller than 4 cm are more likely to be asymptomatic [4]. Tumour size, the multiplicity of lesions, and significant neovascularity are risk factors in angiomyolipoma [7,8].

The following factors are considered indications for treatment in angiomyolipoma: unbearable pain, ruptured tumour, significant haematuria, and large tumour size. Symptoms presented by the patient, as well as the size of the tumour, are predominant factors in deciding the type of treatment [9]. Previously, surgery was the mainstay of treatment for such patients. Endovascular embolisation has the benefits of being less invasive, protecting renal parenchyma, and having lower procedure-related morbidity/mortality [2]. A small tumour (<4 cm) needs yearly imaging follow-up, while large tumours (>4 cm) can be managed by either endovascular embolisation or surgery [9].

Super selective arterial embolisation shows a mean size reduction of the tumour by 43% [9]. The procedure-related complications during endovascular embolisation are very few or none relative to open surgery like nephrectomy. There was no rehaemorrhage observed during a five-year follow-up period [9]. In the present case, the authors performed super selective endovascular embolisation of two feeding arteries supplying the right upper interpolar renal angiomyolipoma. Authors used a microcatheter to navigate as distally as possible to selectively embolise only the tumour-related arteries while sparing the arteries supplying normal renal parenchyma. Authors used PVA particles and coils to embolise the tumour bed and arteries feeding the tumour, respectively.

The frequency of postembolisation syndrome in cases of angiomyolipoma is lower. Super selective embolisation preserves renal parenchyma, and there were no reported deaths in cases of super selective embolisation for renal angiomyolipoma [9]. The patient also experienced symptoms of postembolisation syndrome, which were managed with medical treatment.

A study by Andersen PE et al., has shown that super selective embolisation for angiomyolipoma results in a reduction in tumour volume in addition to devascularising the tumour [10]. Super selective endovascular embolisation in cases of renal angiomyolipoma prevents rebleeding during long-term follow-up. It is a minimally invasive and nephron-sparing procedure, with the preservation of renal function being an important consideration [10]. In the present case, there was no evidence of tumour rebleeding. However, authors do not have long-term follow-up data, so authors cannot comment on the percentage reduction in tumour size after embolisation, which is a limitation of the present case report.

When considering surgical treatment, partial nephrectomy has the advantage of lower recurrence compared to endovascular embolisation. Complications associated with nephrectomy are also more common compared to embolisation. Radical nephrectomy is indicated when there is a high suspicion of malignancy [9]. In the present case, the family consented to endovascular embolisation.

Successful urgent arterial embolisation of ruptured renal angiomyolipoma was described by Kervancioglu S et al., in a case

report of angiomyolipoma with rupture and pseudoaneurysm [11]. The authors used liquid embolic (glue) for embolisation, as the renal angiogram demonstrated a pseudoaneurysm with active extravasation. In the present case, there was no pseudoaneurysm or active extravasation, so authors opted to use PVA particles for deep penetration into the tumour bed vasculature and coils to occlude the main-stem of abnormal feeding arteries for optimal devascularisation of the tumour.

Altuwayr RM et al., discussed successful endovascular embolisation followed by radical nephrectomy in a 42-year-old patient with a ruptured renal angiomyolipoma, beta-thalassaemia minor, and a family history of glucose-6-phosphate dehydrogenase deficiency [12]. In contrast to this, the authors in the present study did not choose to perform nephrectomy as the patient was stable after endovascular embolisation.

The success rate for endovascular embolisation is almost 100%, as mentioned by various authors [2,5,6,13,14]. However, complete embolisation may not always be achieved. There are rare complications associated with embolisation, such as allergy, non targeted embolisation, respiratory complications, retroperitoneal haemorrhage, and complications at the femoral artery puncture site, as elaborated by Murray TE et al., [15]. However, no deaths were reported. In the present case, there was no evidence of contrast allergy, non target embolisation, or pseudoaneurysm of the punctured femoral artery.

CONCLUSION(S)

Ruptured renal angiomyolipoma is a relatively rare clinical emergency condition. Most of the time, the presentation of a ruptured renal angiomyolipoma includes flank pain, haematuria, or retroperitoneal haemorrhage. Endovascular super selective embolisation is an effective, minimally invasive treatment with a high success rate, lower complications, and faster patient recovery while preserving renal parenchyma. Whenever expertise is available, it should be the treatment of choice.

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