

TREATING BILATERAL RENAL ANGIOMYOLIPOMAS IN TUBEROUS SCLEROSIS WITH MULTI-SESSION EMBOLIZATION AND FOLLOW-UP

Atif Rana, Abdur Raheem, Rayyan Pervez, Shahjehan Alam

Department of Radiology, Shifa International Hospital, Islamabad, Pakistan.

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ABSTRACT

We present a case of middle aged lady suffering from tuberous sclerosis (TS) and having symptomatic bilateral renal angiomyolipomas (AML) who was successfully treated with invasive embolization. The right AML was treated first and after 6 months the left AML was attempted. A follow-up with computerized tomogram (CT) scan after 6 months showed regression in size of bilateral lesions. Clinical follow-up after 2 years revealed that patient was asymptomatic after treatment.

Introduction

Angiomyolipoma (AML) has long been treated with partial or complete nephrectomy. Robust advancement in the field of interventional radiology led to better management of the symptomatic AMLs as well as salvage of the kidneys.¹ In Pakistan very few health centers have practiced the selective embolization of AML. We have found only one patient documented in literature who had undergone this procedure and ended in nephrectomy.² Embolization in our patient, first for right and then left with an interval of six months, was quite successful and their sizes regressed on follow up.

Case

A 35 year old female, diagnosed case of bilateral renal AMLs and TS, presented to urology department with gross haematuria on 24th September, 2009. Her ultrasound performed on 12th March, 2005 (outside institution) demonstrated a 6.5 x 5 cm AML in upper pole of right kidney and a 5.5 x

4 cm AML in lower pole of left kidney. Ultrasound performed on the day of presentation showed interval increase in right renal AML (9.3 x 5.6 cm). CT scan performed on the same day demonstrated hemorrhage in the right renal AML and clots in the right renal pelvis, upper ureter and bladder (Fig.1). Cystoscopy was performed and it showed clots in



Figure 1: Coronal image showing bleed in right AML with renal pelvis clot. Also left renal lower pole AML is shown.

the bladder which were evacuated. Patient underwent super selective embolization of right renal AML on 1st Oct, 2009 using a micro-catheter, absolute alcohol and PVA 300-500 μ (cook). Patient's symptoms settled and she was sent home

Correspondence : Dr. Atif Rana
Department of radiology,
Shifa International Hospital,
Islamabad, Pakistan.
Email: atifrana1@hotmail.com

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with preserved post procedure renal functions. Follow up CT scan 6 months later showed the right renal AML had regressed to 3.5 x 3.1 x 4.4 cm (AP x Tr x CC) (Fig. 2). Elective embolization of left renal AML was done on 10th April, 2010 in similar fashion. Again at follow up of 6 months, right AML had further regressed to size of 3.3 x 2.4 x 3 cm and left renal AML had shrunk to 2.4 x 2 x 4 cm (Fig. 3). No renal functions impairment was observed on follow up.



Figure 2: Coronal image after first follow up of 6 months showing marked decrease in right renal AML.



Figure 3: Coronal image of second follow up of another 6 months showing further decrease in right renal AML and decrease in left renal AML.

Discussion

AML is a benign renal neoplasm composed of fat, vascular, and smooth muscle elements. It has an incidence of about 3.2%.³ It can occur in an isolated form or in association with TS.⁴ AML associated with TS constitutes 20% of all cases of renal AML and these are usually symptomatic with multiple bilateral lesions.¹ When associated with TS, these lesions occur at a younger age (53.6 vs 31.5 yrs) and have larger size.⁵

The majority of AMLs are asymptomatic and development of symptoms is mostly related to the

size of tumor. The classic Lenks triad of renal AML includes flank pain, palpable tender mass, and signs of internal bleeding.⁶ The presence of massive retroperitoneal bleeding, known as Wunderlich syndrome, constitutes the most serious complication and occurs in up to 50% of patients with tumors larger than 40 mm.⁷

Currently imaging modalities are sufficient for the diagnosing AMLs and biopsy is rarely required.¹ Based on the presence and amount of detectable fat within the lesion, AMLs were classified in 4 distinct radiological patterns: Pattern-I, predominantly fatty (intrarenal): 54%; Pattern-II, partially fatty (intrarenal or exophytic): 29%; Pattern-III, minimally fatty (most exophytic and perirenal): 11%; and Pattern-IV, without fat (most exophytic and perirenal): 6%.⁸ The imaging technique of choice for confirming the presence of AML is CT. Magnetic resonance imaging (MRI) is more sensitive than CT in identifying the fatty component and is used in some cases where the rest of techniques prove ambiguous. Although ultrasound may suggest the diagnosis, its main usefulness is as a follow-up technique.⁹

Risk of bleed increases with tumor size and it is generally agreed that patients with asymptomatic AML larger than 4 cm should be treated, as should those with symptomatic lesions of any size.¹ Van Baal et al. followed 20 TS patients for 5 years and found that in 20% of them the AMLs enlarged, 35% had renal hemorrhage requiring hospitalization, 10% required nephrectomy, and 5% died.¹¹ Elective procedure may also be offered to women of child bearing age and those patient who are in remote areas and don't have immediate access to the health facility.¹

Traditional procedures such as partial or complete nephrectomy are outdated. In current day practice nephron sparing surgery (NSS) or selective arterial embolization are the procedures being used. Selective arterial embolization gained grounds as the treatment of choice as AMLs are benign, patient symptoms usually result from hemorrhage, and selective embolization preserves some normal renal parenchyma.¹² Furthermore, it is not easy to perform a NSS by delayed elective surgery due to severe tissue adhesion after bleeding.¹³ Oesterling JE et al. have formulated a guideline to go for NSS and

embolization for symptomatic AMLs and 6-12 months follow-up imaging for asymptomatic AMLs.¹⁴ Multiple variables may be considered for choosing the appropriate intervention technique, such as isolated AML or in association with TS, laterality, radiologic features (number, size, location of tumor and associated hemorrhage) and failure of the previous intervention. In case of embolization and NSS failure or location of tumor close to pedicle, radical nephrectomy is considered.¹⁵

Multiple embolic agents are used such as ethanol & PVA, ethanol & ethiodized oil, coils and embolospheres.¹³ In our case we successfully opted for superselective embolization and used absolute alcohol and PVA as embolic agent.


Complications are uncommon and our patient also tolerated the procedure well. Although she did experience mild post embolization syndrome which is a known entity after embolization.¹

Conclusion

Detection of bilateral AML is concerning for underlying tuberous sclerosis. When considering a first line nephron sparing procedure, embolization can serve as the ideal minimally invasive treatment option in expert hands. An added advantage of using super-selective catheter embolization for multiple lesions is that it can be used effectively on more than one occasion. To assess post-treatment response and follow-up CT-scan serves as an effective modality.

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