

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Endovascular treatment of sporadic renal angiomyolipoma presenting with Wunderlich syndrome ☆☆☆★

Devta Viswam, MBBS, MD, DNB*, Keerthiraj B, MBBS, MD, DM, Anand Venugopal, MBBS, MD, Geetanjali Parmar, MBBS, MD

Department of Radiodiagnosis, Kasturba Medical College, Manipal Academy of Higher Education, Mangalore, Karnataka, India

ARTICLE INFO

Keywords:

Angiomyolipoma
Pseudoaneurysm
Wunderlich's syndrome
Selective embolization

ABSTRACT

Angiomyolipomas are benign renal hamartomas with - fat, smooth muscle, and abnormal blood vessel (tortuous, dysmorphic) components. The risk of hemorrhage is related to size of the tumor, presence of aneurysm, associations with tuberous sclerosis. In this case report we review the case of a 42-year-old woman presenting with acute flank pain and decreased hemoglobin, who was diagnosed with AML with right renal artery pseudo aneurysm on CT. Subsequent coil embolization of feeding vessels and using PVA particles was successfully done. Selective embolization remains a minimally invasive, attractive option, and a nephron sparing approach.

© 2023 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Renal angiomyolipoma (AML) is essentially a solid, benign, mesenchymal, hamartomatous neoplasm with well-established characteristics and consisting of 3 components: adipose tissue, abnormal blood vessels and smooth muscle. There are mainly 2 types: first, 80% cases are of singular sporadic occurrence in higher age groups of 50–80 years, the second can be a part of tuberous sclerosis complex (20%, usu-

ally bilateral). Both types have a female predominance. Ten percent of patients of tuberous sclerosis can have a lethal retroperitoneal hemorrhage, also irrevocable damage to the renal parenchyma. Thus, timely radiological imaging in renal complications of tuberous sclerosis is imperative [1,2].

Expanding AMLs have a tendency to bleed abruptly and painfully, owing to dysmorphic blood vessels developing macro and micro aneurysms that rupture spontaneously. Risk of hemorrhage is directly proportional to the size (> 4 cm) and warrants embolization prophylactically [3,4].

☆ Previous presentation: Organisation KARAVALI RADCON 36TH Annual IRIA Conference Karnataka Chapter, TMA PAI International Convention Centre, Mangaluru, Karnataka, Dated March 7 and 8, 2020.

☆☆ Acknowledgments: None.

★ Competing Interests: The authors declare no conflicts of interest.

* Corresponding author.

E-mail address: devtavismam@gmail.com (D. Viswam).

<https://doi.org/10.1016/j.radcr.2023.11.077>

1930-0433/© 2023 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Renal angiomyolipoma may result in an uncommon condition known as Wunderlich's syndrome consisting of hypovolemic shock, acute flank pain and palpable mass in the flank(s) – also, referred to as the Lenk's triad. Hence, hemorrhage with a life threatening potential occurs due to acute, nontraumatic rupture of the elastin-poor arteries into the subcapsular and peri-nephric spaces [4,5].

Characteristic radiological features of AMLs are attributed to the fat component, include echogenicity on sonography, hypodensity with negative attenuation on plain computed tomographic studies and magnetic resonance imaging [6].

Hemodynamically unstable patients with computed tomography features such as intratumoral bleeding, arterial extravasation, evidence of pseudo aneurysm, anemia, hematuria indicate immediate interventional embolization [7,8].

Here is a case of renal angiomyolipoma with pseudo aneurysm of segmental branch of right renal artery at the mid pole with sub capsular hematoma and its interventional coil embolization.

Case description

Forty-two-year female, a known case of right renal angiomyolipoma not associated with tuberous sclerosis presented with complain of right flank pain and hematuria. On systemic examination, patient had pulse rate of 118-Tachycardia, low blood pressure (90/60 mm Hg) and respiratory rate - 25. Lab investigations- Hb-7.2, patient's condition was deteriorating clinically with persistent tachycardia and decreasing blood pressure, repeat Hb after 24 hours was 6.8. blood sugar levels, renal function tests were in normal limits.

In view of history of renal angiomyolipoma, flank pain, falling hemoglobin levels and symptoms of hypovolemic shock, CT urogram was done.

CT urogram showed exophytic mixed density (with focal areas of fatty attenuation and hyperdense areas) lesion measuring $4.7 \times 4.7 \times 4.9$ cm (AP $\times$ TR $\times$ CC) showing subtle arterial post contrast enhancement arising from the posterior aspect of renal parenchyma of mid pole of right kidney extending into the adjacent perinephric space with breach in the renal cortex overlying the lesion, however no obvious e/o active contrast extravasation.

Evidence of aneurysm in the segmental artery supplying the renal mass-suggestive of angiomyolipoma with rupture and perinephric hematoma (Fig. 1C).

Sub capsular collection (40-60 HU) posterior to right kidney $8.2 \times 10.2 \times 14.7$ cm (AP $\times$ TR $\times$ CC) causing anterosuperior displacement of the right kidney with extensive perinephric fat stranding S/o acute-subacute hematoma with perinephric fat stranding (Fig. 1A,B).

Diagnosis of Wunderlich syndrome was made.

In view of imaging findings, patient was subjected to the DSA with endovascular coil embolization of pseudo aneurysm.

Under local anesthesia, under all aseptic precautions right CFA puncture was done.

Selective right renal artery angiogram was done using 5F SIM-1catheter. Angiogram showed mid pole right renal artery

branch pseudo aneurysm with active contrast extravasation and tortuous corkscrew vessels supplying angiomyolipoma from mid polar another branch of right renal artery (Figs. 2A,B and 4A).

Using 5F SIM 1 as guiding catheter placed in the right main renal artery, Micro catheter was taken into the mid polar right renal artery branch distal to the feeder feeding the pseudo aneurysm and fiber coils were placed into the distal artery first and then coils of various sizes were placed into the feeder pedicle and then proximally.

Later micro catheter was taken into the feeder feeding the angiomyolipoma and embolized using PVA particles 250-350 microns and later coil was placed proximally.

Check angiogram showed complete exclusion of pseudo aneurysm from the circulation with no further contrast extravasation and obliteration of the feeders feeding the angiomyolipoma and normal filling of rest of right renal artery branches (Figs. 3A,B and 4B).

After the procedure the vitals and hemoglobin level of the patient came back within normal limits (HR – 76, RR -16, BP – 130/80; and Hb -10.8 gm %). There was a significant improvement in the symptoms of flank pain and hematuria after the procedure.

At the time of discharge (4 days later), the patient had no symptoms.

Discussion

Angiomyolipoma is a benign, mesenchymal, triphasic tumor with dysmorphic blood vessels, fat, and smooth muscle components in varying proportions.

Retroperitoneal hematoma remains the most potentially lethal complication of angiomyolipoma due to nontraumatic rupture with a number of commonly known risk factors namely: tumor size (>4 cm). However up-to-date evidences point towards the multifactorial dependency of rupture, with strongest association with presence of intratumoral aneurysms – others include coagulopathies, hormone levels, pregnancy, trauma and association with tuberous sclerosis (mutation of TSC gene) and lymphangioleiomyomatosis. In our case there was a pseudo aneurysm involving the mid segmental branch of right renal artery which was picked up on CT urogram and confirmed on DSA which showed active contrast extravasation: suggestive of rupture. The tumor size was $4.7 \times 4.7 \times 4.9$ cm. Hence, risk factors contributing to the most lethal complication of angiomyolipoma, i.e. retroperitoneal/sub capsular hematoma were present in our case. Non-traumatic sub capsular hematoma with – acute flank pain, flank mass, and hypovolemic shock were noted in this case, representing Wunderlich syndrome.

Wunderlich syndrome is a life threatening emergency condition owing to spontaneous hemorrhage confined to perirenal and sub capsular spaces of kidney and may lead to hypovolemic shock. Hence, early diagnosis is of utmost value, also to salvage the kidney from a nephrectomy indicated complication like persistent hemorrhage.



Fig. 1 – (A, B, C): (A,B)-axial and coronal CT urogram depicting subcapsular hematoma. C-coronal section of CT urogram in arterial phase depicting pseudoaneurysm.



Fig. 2 – (A, B): Shows anteroposterior (AP) and left anterior oblique (LAO) views of preprocedure digital subtraction angiography (DSA) showing right renal midpolar branch pseudoaneurysm.

Interventional radiological techniques like selective embolization are minimally invasive, thus, highly recommended for bleeding angiomyolipoma and preventive to reduce risks of rupture of aneurysm of AML. Other advantages include lesser complication frequency, preserved kidney function, less

trauma and recovery time. Embolization also aids in providing a blood-less field prior to surgery. However, disadvantages include possibility of relapse and recurrence of bleed, even on shrinkage of tumor size, also post embolization syndrome consisting of leukocytosis, slight fever, and pain [9].



Fig. 3 – (A, B): Shows left anterior oblique (LAO) and right anterior oblique (RAO) views of post coil embolization digital subtraction angiography (DSA) showing no evidence of pseudoaneurysm filling.

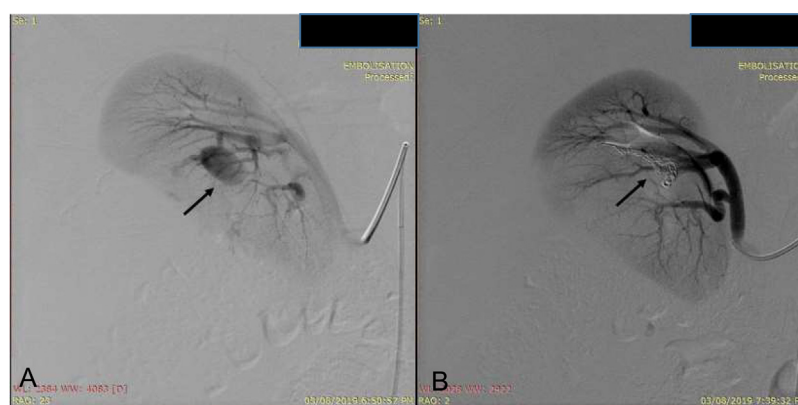


Fig. 4 – (A,B): (A) Shows right anterior oblique (RAO) view of preprocedure DSA showing right renal midpolar branch pseudoaneurysm, (B) shows anteroposterior view of postprocedure DSA showing complete obliteration of aneurysm.

Conclusion

Urgent trans arterial embolization in cases of ruptured renal angiomyolipoma is a parenchyma sparing alternative to partial/total nephrectomy leading to significant tumor size reduction. Monitoring of growth of AML size is important, especially in cases with tuberous sclerosis. Interval follow-ups are essential to look for recurrence. Long term efficacy of embolization is significant, with advantages of feasibility, safety and being minimally invasive.

Patient consent

The authors have obtained written informed consent for the publication of this case report.

REFERENCES

- [1] Rimón U, Duvdevani M, Garniek A, Golan G, Bensaid P, Ramon J, et al. Ethanol and polyvinyl alcohol mixture for transcatheter embolization of renal angiomyolipoma. *Am J Roentgenol* 2006;187(3):762–8.
- [2] BujPradilla MJ, MartíBallesté T, Torra R, VillacampaAubá F. Recommendations for imaging-based diagnosis and management of renal angiomyolipoma associated with tuberous sclerosis complex. *Clin Kidney J* 2017;10(6):728–37.
- [3] Chronopoulos PN, Kaisidis GN, Vaiopoulos CK, Perits DM, Varvarousis MN, Malioris AV, et al. Spontaneous rupture of a giant renal angiomyolipoma—Wunderlich's syndrome: Report of a case. *Int J Surg Case Rep* 2016;19:140–3.
- [4] Katabathina VS, Katre R, Prasad SR, Surabhi VR, Shanbhogue AK, Sunnapwar A. Wunderlich syndrome: cross-sectional imaging review. *J Comput Assist Tomogr* 2011;35(4):425–33.
- [5] Dhanapal V, Ramachandran R, Radhan P, Vivekanandan B, Jeevanandham B, Jacob P. The Many Facets of Wunderlich Syndrome: A Multidetector Computed Tomography based Review.
- [6] Philip CH. Interventional radiological treatment of renal angiomyolipoma. *Med Bulletin* 2010;15(7).
- [7] Andersen PE, Thorlund MG, Wennevik GE, Pedersen RL, Lund L. Interventional treatment of renal angiomyolipoma: immediate results and clinical and radiological follow-up of 4.5 years. *Acta radiologica Open* 2015;4(7):2058460115592442.
- [8] Salik AE. Transarterial Embolization of Symptomatic Renal Angiomyolipomas. *Med J Bakirkoy* 2019;15(3):304–9.
- [9] Jinzaki M, Silverman SG, Akita H, Nagashima Y, Mikami S, Oya M. Renal angiomyolipoma: a radiological classification and update on recent developments in diagnosis and management. *Abdom Imaging* 2014;39(3):588–604.