

Renal Angiomyolipoma presenting with Wunderlich's Syndrome – A Case Report

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Abstract- INTRODUCTION: Angiomyolipoma (AML) is a benign neoplasm; in its classic form it consists of blood vessels, smooth muscle, and adipose tissue. The main complication of AML is retroperitoneal hemorrhage caused by tumor rupture, which can be severe and life threatening.

CASE REPORT: A 48 years male presented with complaints of left flank pain for the last 4 days. General examination revealed pallor and vitals showed signs of hypovolemia with a Blood Pressure (BP) of 70/50 mmHg & a pulse rate of 120/min. Resuscitative measures were started with monitoring in intensive care unit (ICU) and vitals were stabilized with IV fluids and other measures. Preliminary investigations showed HB-8.4 gm%, TC-7000 cells/dl, RBS- 94 mg/dl, Creatinine- 1.5 mg/dl, USG Abdomen showed enlarged Right kidney, showing numerous heterogeneous, diffuse lesions. A large heteroechoic mass like lesion was seen in left renal fossa and a hypoechoic area of size 7x5.4 cm in superior aspect of left kidney. CT Abdomen revealed B/L renal angiomyolipomas with Left perirenal collection. After resuscitation, selective embolization of the left upper pole branch of the renal artery was performed with coils. The patient was transferred to the ICU where he received a total of three units of packed red blood cells. Following the procedure, the patient's condition improved, and he was hemodynamically stable.

Index Terms- Angioembolization allows rapid patient stabilization in cases of acute hemorrhage and provides good renal preservation in cases of multifocal AML.

I. INTRODUCTION

Angiomyolipoma (AML) accounts for less than 10% of renal tumors, in autopsy series¹ and ultrasound-screened populations showing incidences of 0.3% and 0.13%, respectively, in the general population.¹ It is a benign tumor which consists of blood vessels, smooth muscle, and adipose tissue.² Most AML patients are usually asymptomatic and the diagnosis of AML is often incidental.^{3,4} The main complication of AML is retroperitoneal hemorrhage caused by tumor rupture, which can be severe and life threatening. This Urological emergency, also called Wunderlich's Syndrome, was how our case presented.¹

CASE: A 48 years male presented with complaints of left flank pain which was mild, dull aching, continuous, and non-

radiating, with no aggravating or relieving factors for the last 4 days. There was no significant past medical or family history. General examination revealed pallor and vitals showed signs of hypovolemia with a Blood Pressure (BP) of 70/50 mmHg & a pulse rate of 120/min. On Systemic examination, a palpable lump was felt in the left flank that was tender, firm, diffuse, ill-defined and extended three fingers above the left iliac crest, not crossing midline. Rest of the systemic examinations revealed no significant findings. Resuscitative measures were started with monitoring in intensive care unit (ICU) and vitals were stabilized with IV fluids and other measures. Preliminary investigations showed HB-8.4 gm%, TC-7000 cells/dl, RBS- 94 mg/dl, Creatinine- 1.5 mg/dl, Urine r/e: RBC-0-1/hpf, Pus cell-0-2/hpf. USG Abdomen showed enlarged Right kidney, showing numerous heterogeneous, diffuse lesions (Fig: 1). A large heteroechoic mass like lesion was seen in left renal fossa and a hypoechoic area of size 7x5.4 cm in superior aspect of left kidney. CT Abdomen revealed multiple enhancing soft tissue lesions of varying sizes in both kidneys with internal fat component (Left>Right). On post contrast study in arterial phase few dilated arteries noted in these lesions. Heterogeneous collection approx. 10x7x7cm was noted in upper pole of left kidney compressing and distorting renal parenchyma and left pelvicalyceal system (PCS). Features were suggestive of B/L renal angiomyolipomas (AML) with large lesion in the left kidney with active hemorrhage and perirenal collection (Fig: 2).

Interventional Radiology Opinion was taken. After resuscitation, selective embolization of the left upper pole branch of the renal artery was performed with coils. The patient was shifted to the ICU where he received a total of three units of packed red blood cells. Following the procedure, the patient's condition improved, and he was hemodynamically stable.

II. DISCUSSION

Renal AML is a benign tumor known to occur sporadically and in association with genetic syndromes like tuberous sclerosis (TS) and lymphangiomyomatosis.^{1,3} Genetic studies in patients with tuberous sclerosis complex (TSC) have shown discovery of two genes associated with angiomyolipomas: TSC1 on chromosome 9q (encoding for hamartin protein) and TSC2 on chromosome 16p (encoding for tuberin protein). In sporadic TSC patients, 10% had TSC1 mutations, 68% had TSC2 mutations, and 22% had no mutations detected.⁵

The typical sporadic presentation is of a middle-aged woman with a single asymptomatic tumor. Sporadic AML are usually detected incidentally.² In 10% of cases, AML can also present with features of acute retroperitoneal bleeding, manifesting with acute flank pain, abdominal tenderness and signs of internal bleeding such as hematuria.^{1,4} This Urological emergency is also called Wunderlich Syndrome and is the most significant complication of Renal Angiomyolipomas.³ Risk factors for rupture of angiomyolipoma includes aneurysm size of >5mm which strongly correlates with rupture followed by tumor size > 4mm, pregnancy and genetic abnormalities.⁶

On Grayscale Ultrasound, the lesion appears as a well-defined, markedly, hyperechoic mass relative to normal renal parenchyma with acoustic shadowing. However, a subset of minimal fat AMLs are isoechoic to slightly hypoechoic and lack shadowing. Color Doppler will detect pseudo-aneurysms which appear as a color-filled ovoid or round structures within or adjacent to an AML. Color flow appears as swirling or "yin and yang" sign.⁷ On CT scan, the lesion appears as a well-margined, heterogeneous mass with macroscopic fat arising from renal cortex, which originates from a triangular or rectangular notch-like defect in the cortex, called the "Notch Sign".⁷ The presence of fat, within a renal lesion confirmed on non-enhanced CT by a value of -20 HU or less, is considered the diagnostic hallmark. Findings of more than 20 pixels with attenuation of less than -20 HU and of more than 5 pixels with attenuation of less than -30 HU have been found to have a positive predictive value of 100%.⁸

On MRI, it appears typically as heterogeneous signal intensity. Macroscopic fat will show high signal intensity on T1WI and T2WI. Other findings include loss of signal fat suppression (frequency selective), signal loss in out of phase images on opposed phases imaging, India ink artifact and chemical shift artifact (alternating high and low signal bands on out of phase images). However, clear cell RCCs lack India ink artifact and do not lose signal on conventional T1 fat-suppressed sequences.⁹ Angiographic findings include highly vascular mass with disorganized, long, and tortuous vessels, sacculated pseudo-

aneurysms, absence of arterio-venous shunts, "Sunburst" appearance of capillary nephrogram.⁷

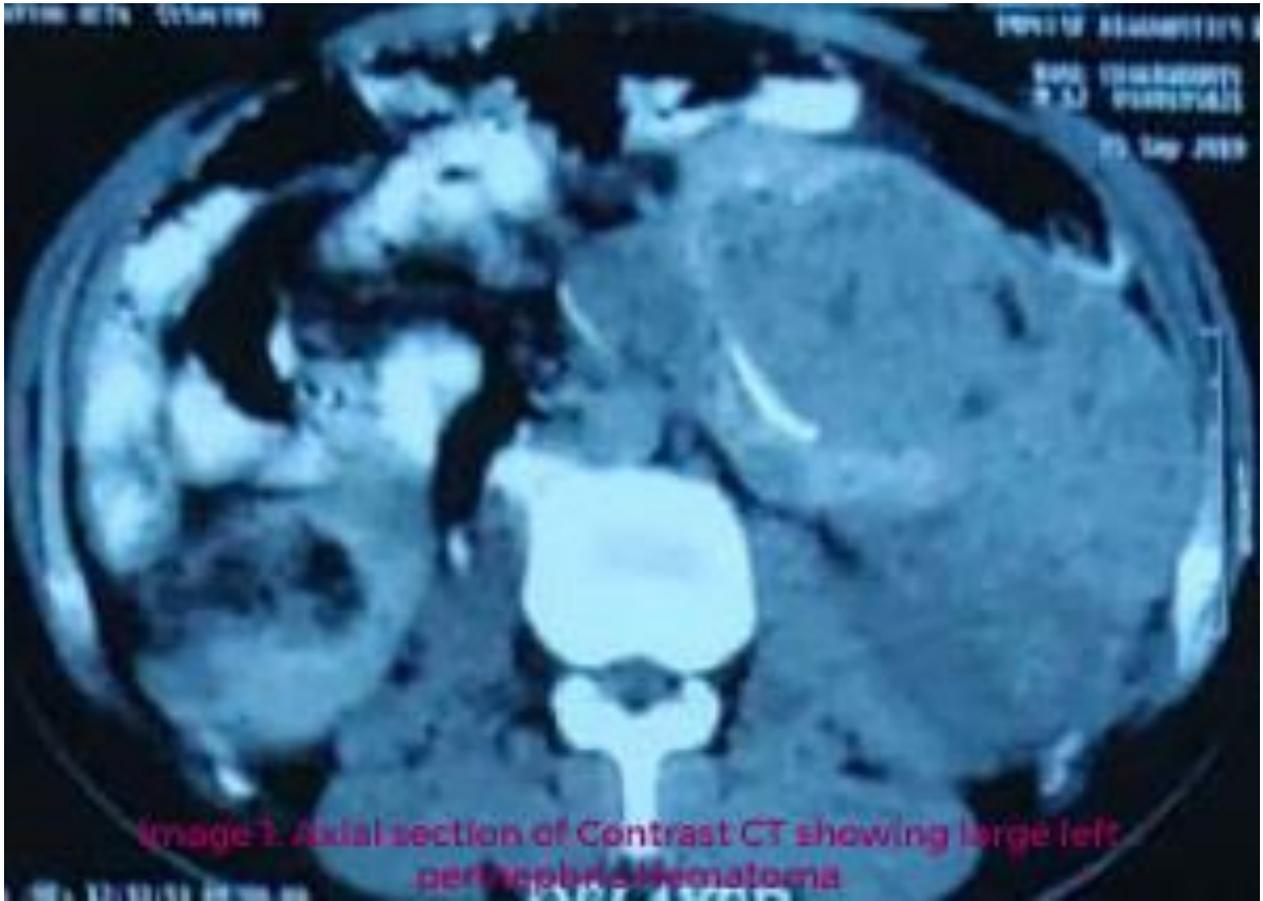
The management of AMLs has been correlated with symptoms. Patients with small tumors (<4 cm) that tend to be asymptomatic are managed conservatively, under periodic ultrasonographic follow up.¹⁰ Ablative therapies such as radiofrequency ablation and cryoablation have also been used for the treatment of angiomyolipoma, but follow-up remains short^{11,12}

Intervention should be considered for larger tumors, particularly if the patient is symptomatic, taking into account the patient's age, comorbidities, and other related factors.¹³ In women of childbearing age and patients with limited access to surveillance or to emergency care, a proactive approach should also be considered.¹³

A nephron-sparing approach, by either selective embolization or open or laparoscopic or robotic partial nephrectomy, is clearly preferred to radical nephrectomy in patients with angiomyolipomas requiring intervention.^{14,15}

Preservation of renal tissue remains a priority in those with TSC or multicentric angiomyolipoma and particularly in patients with underlying renal insufficiency. The overall complication rate with embolization in one series was 10% which is similar to rates of partial nephrectomy and included hemorrhage, abscess formation, or sterile liquefaction of the tumor requiring percutaneous drainage or surgical intervention.¹⁵ These data highlight the need for extended follow-up after selective embolization, which would not be required after partial nephrectomy.¹⁶ Selective embolization should be considered as first-line therapy¹⁷ in patients with acute or potentially life-threatening hemorrhage, because surgical exploration in this setting is often associated with total nephrectomy.¹⁶

In a systemic review of 524 cases of angioembolization, at a mean follow-up of 39 months, the mean size reduction was 3.4 cm. Unplanned repeat embolization or surgery was required in 20.9% of cases during this period.¹⁷





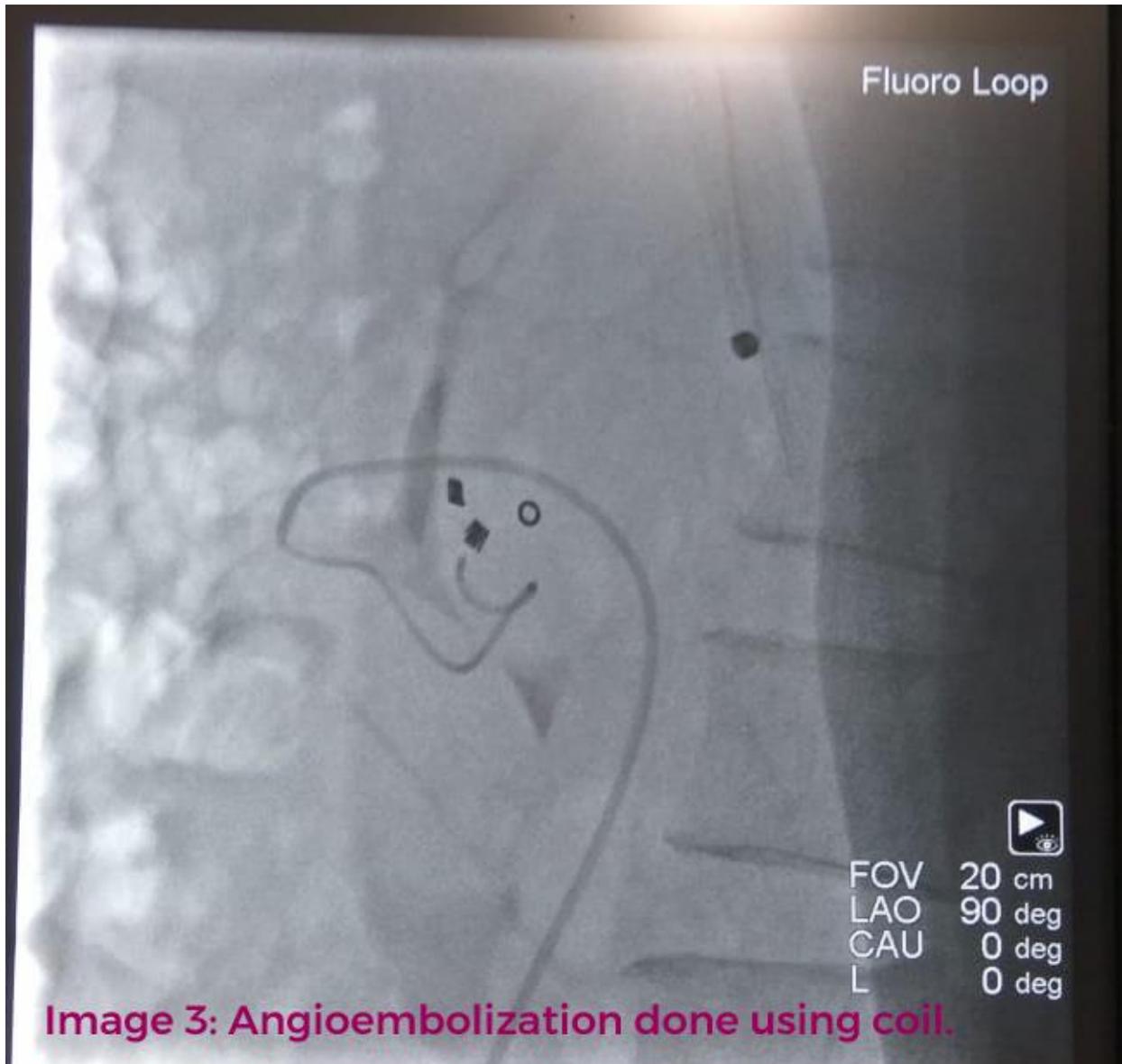


Image 3: Angioembolization done using coil.

III. CONCLUSION

A case of Wunderlich's Syndrome with massive retroperitoneal hemorrhage who was hemodynamically unstable presented in the Urology emergency. Patient was resuscitated and selective Renal Angioembolization was performed. Angioembolization allows rapid patient stabilization in cases of acute hemorrhage and provides good renal preservation.¹⁴

REFERENCES

- [1] Eble JN. Angiomyolipoma of kidney. *SeminDiagnPathol* 1998;15(1):21–40.
- [2] Bissler and Kingswood,2004) Bissler JJ, Kingswood JC. Renal angiomyolipomata. *Kidney Int* 2004;66(3):924–34.
- [3] Seyam RM, Bissada NK, Kattan SA, et al. Changing trends in presentation, diagnosis andmanagement of renal angiomyolipoma: comparison of sporadic and tuberous sclerosis complex–associated forms. *Urology*. 2008;72(5):1077–1082.
- [4] Steiner MS, Goldman SM, Fishman EK, et al. The natural history of renal angiomyolipoma. *Urol*.1993;150(6):1782–1786.
- [5] Dabora SL, Jozwiak S, Franz DN, Roberts PS, Nieto A, Chung J, Choy YS, Reeve MP, Thiele E, Egelhoff JC, Kasprzyk-Obara J. Mutational analysis in a cohort of 224 tuberous sclerosis patients indicates increased severity of TSC2, compared with TSC1, disease in multiple organs. *The American Journal of Human Genetics*. 2001 Jan 1;68(1):64-80.
- [6] Wang C, Li X, Peng L, GouX, Fan J. An update on recent developments in rupture of renal angiomyolipoma. *Medicine*. 2018 Apr;97(16)
- [7] Aal AA, Moawad S, Hanaoka M, Abouarab A, Mahmoud K, Oser R, Gunn A, Ertel N, Saddekni S, Birmingham AL. Endovascular management of renal angiomyolipoma using a unique sclerotic-embolic mixture. *European Congress of Radiology* 2017.
- [8] Simpfendorfer C, Herts BR, Motta-Ramirez GA, et al. Angiomyolipoma with minimal fat on MDCT: can counts of negative-attenuation pixels aid diagnosis? *AJR Am J Roentgenol*.2009;192(2):438–443.
- [9] Israel GM, Hindman N, Hecht E, Krinsky G. The use of opposed-phase chemical shift MRI in the diagnosis of renal angiomyolipomas. *American Journal of Roentgenology*. 2005 Jun;184(6):1868-72.
- [10] Matin S, Tamboli P, Wood C. Renal angiomyolipoma. *Bukowski RM, Novick AC. Clinicalmanagement of renal tumors. Humana: Totowa, NJ; 2008.*

- [11] Caviezel A, Terraz S, Schmidlin F, et al. Percutaneous cryoablation of small kidney tumours under magnetic resonance imaging guidance: medium-term follow-up. *Scand J Urol Nephrol*.2008;42(5):412–416.
- [12] S Prevo W, van den Bosch MA, Horenblas S. Radiofrequency ablation for treatment of sporadic angiomyolipoma. *Urology*. 2008;72(1):188–191
- [13] Nelson C, Sanda M. Contemporary diagnosis and management of renal angiomyolipoma. *J Urol*.2002;168(4 Pt 1):1315–1325.
- [14] Fazeli-Matin S, Novick A. Nephron-sparing surgery for renal angiomyolipoma. *Urology*.1998;52(4):577–583.
- [15] Boorjian SA, Frank I, Inman B, et al. The role of partial nephrectomy for the management of sporadic renal angiomyolipoma. *Urology* 2007;70(6):1064–8.
- [16] Pappas P, Leonardou P, Papadoukakis S, et al. Urgent super selective segmental renal artery embolization in the treatment of life-threatening renal hemorrhage. *Urol. Int.* 2006;777 (1):34–41.
- [17] Murray TE, Doyle F, Lee M. Transarterial embolization of angiomyolipoma: a systematic review. *The Journal of urology*. 2015 Sep 1;194 (3):635-9.