SUNTEXT REVIEWS

SunText Review of Medical & Clinical Research

Open Access
Case Report
Volume 1:2

Case Study: Embolization of an Angiomyolipoma in a Pediatric Tuberous Sclerosis Patient

Mousa M[1,*], Alzate D[1], Bisht R[1], Towbin R[2] and Aria D[2]

[1]University of Arizona College of Medicine, Phoenix, USA
[2]Phoenix Children's Hospital, USA

[*]**Corresponding author:** Mousa M, University of Arizona College of Medicine, Phoenix, USA; E-mail: momousa22 (at) email (dot) arizona (dot) edu

Received date: 12 September 2020; **Accepted date:** 22 September 2020; **Published date:** 29 September 2020

Citation: Mousa M, Alzate D, Bisht R, Towbin R, Aria D (2020). Case Study: Embolization of an Angiomyolipoma in a Pediatric Tuberous Sclerosis Patient. SunText Rev Med Clin Res 1(2): 111.

DOI: https://doi.org/10.51737/2766-4813.2020.011

Abstract

We present a case in which a coil embolization was used to treat an active hemorrhage from an angiomyolipoma in a pediatric patient. Angiomyolipomas most commonly occur in patients with tuberous sclerosis and often bleed once the tumor is larger than 4 cm. Traditional treatments such as partial or complete nephrectomies can result in significant complications. An arterial embolization was performed to target the hemorrhagic tumor.

Keywords: Angiomyolipoma; Pediatric Tuberous Sclerosis

Case Summary

A 16-year-old female presented to an outside institution with syncope, right upper quadrant abdominal pain, and pallor. Past medical history was significant for tuberous sclerosis, developmental delay, and hepatic cavernous malformation. Physical exam demonstrated ash leaf patches over the torso. Laboratory findings revealed a haemoglobin level of 9.6 g/dL while a CT abdomen/pelvis demonstrated an acute, large, intra-abdominal haemorrhage. The patient was transferred to our emergency department and active bleeding was confirmed with a repeat haemoglobin of 6.6 g/dL. Vitals signs were stable but significant for tachypnea with a respiratory rate of 40 breaths/min, tachycardia with a heart rate of 109 bpm, and normotensive with a blood pressure of 129/98 mmHg. The patient was transfused pRBCs while interventional radiology was consulted for embolization.

Imaging Findings

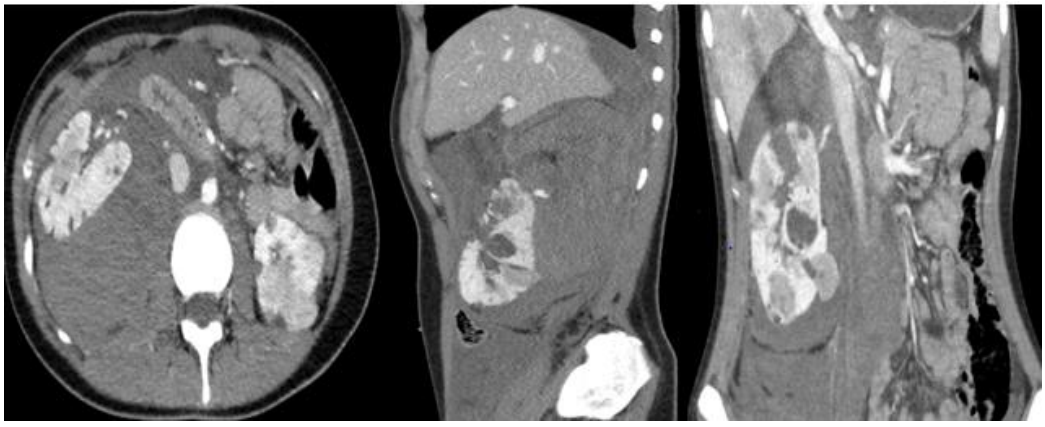

Figure 1: *Contrast-enhanced CT abdomen/pelvis.*

Axial, Sagittal, and Coronal images demonstrate multiple bilateral renal masses demonstrating fat-attenuation, consistent with angiomyolipomas. There is a large right prerenal hematoma displacing the kidney anteriorly with active contrast extravasation (black arrow) along the posteromedial aspect of the right upper pole (Figure 1).

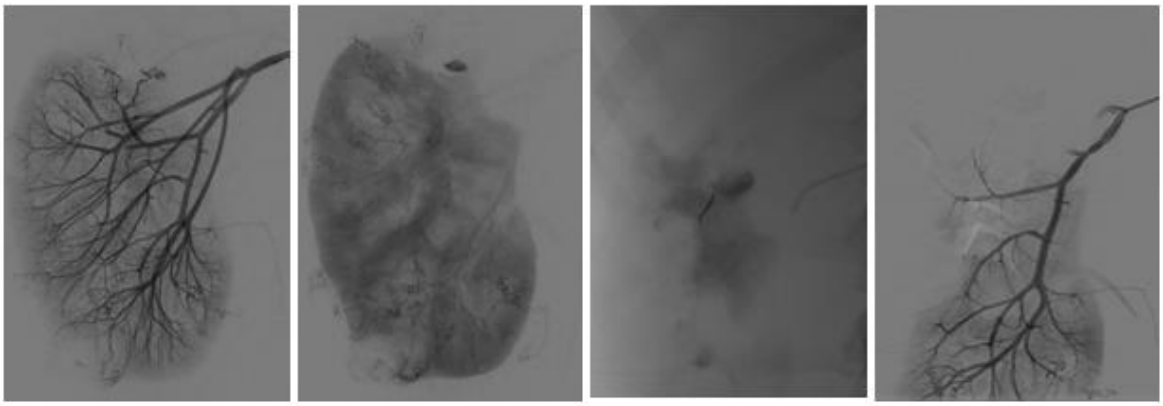

Figure 2: *Right renal angiography.*

A. Arterial phase DSA image demonstrates extravasation and pooling of contrast along the medial upper pole (black circle) consistent with site of active haemorrhage.
B. Parenchymal phase DSA image demonstrates persistence of contrast (black arrow).
C. Coil embolization was performed with deployment (black arrow) immediately proximal to the site of active haemorrhage.
D. Post-embolization arterial phase DSA image demonstrates complete occlusion of the feeding artery with resolution of active haemorrhage (Figure 2).

Diagnosis

Active haemorrhage secondary to angiomyolipoma in patient with tuberous sclerosis.

Discussion

Renal angiomyolipoma (AML) is a benign mesenchymal soft tissue tumour that arises in the cortex or medulla of the kidney. AML is considered "triphasic" in that it consists of adipose tissue, blood vessels, and smooth muscle [1]. AML can arise sporadically or through an inherited disorder. AML most commonly arises due to tuberous sclerosis compared to sporadic [2]. The most dominant type of AML present in tuberous sclerosis is of epithelioid origin.

Histologic fat composition of the tumour allows a diagnosis of AML to be made readily by abdominal CT or MRI [3]. Most sporadic AMLs are found incidentally, however in a patient with tuberous sclerosis, a CT scan is ordered to look for potential renal AMLs. AMLs may contain macroscopic or microscopic fat. In order to distinguish between the two, an MRI may be ordered to assess for fat-suppression. Due to their vascular nature, AMLs are prone to aneurysm formation and rupture. Complications such as haemorrhage can result in a symptomatic drop in haemoglobin and anaemia. Up to one-third of patients can present with hypovolemic shock [4]. Symptomatic AMLs can present with Lenk's triad, which consists of flank pain, abdominal tenderness and internal bleeding. Bleeding typically occurs when tumours are greater than 4 cm. Patients are typically asymptomatic when tumours are less than 4 cm [5,6]. Traditional treatments of AML included nephrectomies or partial nephrectomies. However, these therapeutic options are associated with significant complications and risks [7]. Today, the preferential therapy is angiography with selective arterial embolization [8].

Conclusion

Renal angiomyolipomas are commonly seen in patients with tuberous sclerosis. Despite their multiplicity, they rarely are prone to aneurysm formation or rupture when less than 4 cm. Small lesions can therefore be closely monitored via imaging. AMLs larger than 4 cm have greater propensity for acute bleeding and are therefore amenable to angiography and embolization as opposed to traditional surgical therapeutic options even when asymptomatic. When symptomatic, acute arterial bleeding commonly requires urgent intervention.

References

1. Eble JN, Sauter G, Epstein JI, Sesterhenn IA. World Health Organization classification of tumors: pathology and genetics. Tumors of the urinary system and male genital organs. IARC Press. 2004.

2. Seyam RM, Bissada NK, Kattan SA, Mokhtar AA, Aslam M, Fahmy WE, et al. Changing trends in presentation, diagnosis and management of renal angiomyolipoma: comparison of sporadic and tuberous sclerosis complex-associated forms. Urology. 2008; 72:1077-1082.
3. Schieda N, Hodgdon T, El-Khodary M, Flood TA, McInnes MDF. Unenhanced CT for the diagnosis of minimal-fat renal angiomyolipoma. AJR Am J Roentgenol. 2014; 203: 1236-1241.
4. Rimon U, Duvdevani M, Garniek A, Golan G, Bensaid P, Ramon J, et al. Large renal angiomyolipomas: digital subtraction angiographic grading and presentation with bleeding. Clin Radiol. 2006; 61: 520-526.
5. Oesterling JE, Fishman EK, Goldman SM, Marshall FF. The management of renal angiomyolipoma. J Urol. 1986; 135: 1121-1124.
6. Yamakado K, Tanaka N, Nakagawa T, Kobayashi S, Yanagawa M, Takeda K, et al. Renal angiomyolipoma: relationships between tumor size, aneurysm formation, and rupture. Radiol. 2002; 225: 78-82.
7. Faddegon S, So A. Treatment of angiomyolipoma at a tertiary care center: the decision between surgery and angioembolization. Can Urol Assoc J. 2011; 5: 138-141.
8. Bissler J, Cappell K, Charles H, Song X, Liu Z, Prestifilippo J, et al. Rates of interventional procedures in patients with tuberous sclerosis complex-related renal angiomyolipoma. Curr Med Res Opin. 2015; 31: 1501-1507.