

INTERVENTIONAL RADIOLOGY

Hemorrhagic renal angiomyolipoma

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CASE SUMMARY

A 63-year-old female patient with history of end-stage renal disease (ESRD) and tuberous sclerosis presented to the emergency department (ED) with right flank pain.

IMAGING FINDINGS

A computed tomography (CT) scan in the ED (Figure 1) revealed a new hemorrhagic mass in the upper pole of the right kidney. Three days after initial presentation, the patient's hemoglobin levels fell by 3 units, and she was brought to IR for emergent embolization (Figures 2 and 3).

DIAGNOSIS

Hemorrhagic renal angiomyolipoma
There is a high-attenuation mass in the upper pole of the right kidney consistent with a hemorrhagic lesion. The differential diagnosis includes hemorrhage from an angiomyolipoma, renal cell carcinoma, or a metastatic lesion. The presence of bilateral fat-containing renal masses as well as a history of tuberous sclerosis make hemorrhagic

renal angiomyolipoma the most likely diagnosis.

DISCUSSION

Tuberous sclerosis is an autosomal-dominant disease with variable penetrance that affects approximately 40,000 individuals in the United States and 2 million people worldwide. There are 2 abnormal genes associated with tuberous sclerosis, the TSC1 gene on chromosome Q19 and the TSC2 gene located on chromosome P16. These genes have an important role in tumor suppression. The classic triad of tuberous sclerosis consists of adenoma sebaceum, seizures, and mental retardation, but is only seen in one-third of patients. Major features of tuberous sclerosis include facial angiofibromas, cortical tubers, subependymal nodules, giant cell astrocytomas, cardiac rhabdomyomas, lymphangiomyomatosis, and renal angiomyolipomas.¹

Angiomyolipomas (AML) are benign hamartomatous lesions composed of vessels, smooth muscle, and adipose tissue. Although 80% are



FIGURE 1. Non-contrast coronal CT image from initial presentation. There are bilateral masses replacing the normal renal parenchyma. These include a high-attenuation lesion in the upper pole of the right kidney most consistent with a hemorrhagic mass. There are also bilateral fat-containing renal masses.



FIGURE 2. Digital subtraction angiographic image of the right renal artery demonstrates increased renal vascularity.

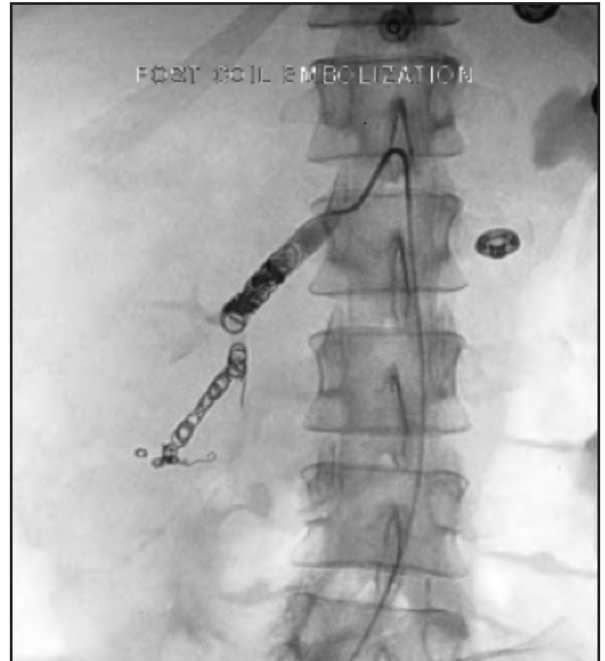


FIGURE 3. Digital subtraction image demonstrates complete embolization of the right renal artery with embosphere particles and coils.

sporadic and solitary, 20% are multiple and associated with syndromes such as tuberous sclerosis. These lesions are generally benign except for the rare epithelioid subtype. Vessels associated with AMLs are dysplastic in nature and therefore predisposed to aneurysmal dilatation and hemorrhage, particularly when an AML is >4 cm. Therefore, embolization of lesions >4 cm is recommended.² When there is spontaneous hemorrhage of AMLs, also known as Wunderlich's syndrome, management

options include conservative management, embolization, and nephrectomy depending on hemodynamic considerations. In the above mentioned case, the patient was actively bleeding and, therefore, could not be treated conservatively. Her hemodynamic status was stable enough to avoid surgical nephrectomy, and thus, she was an ideal patient for embolization.

CONCLUSION

Bilateral angiomyolipomas are very common in patients with tuberous

sclerosis. When hemorrhage is noted or when they are >4 cm, embolization is recommended.

REFERENCES

1. Castillo M. *Neuroradiology: Core Curriculum*. 1st ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2002.
2. Roth CG. *Fundamentals of body MRI*. Philadelphia, PA: Elsevier Saunders; 2012.

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