

Renal Angiomyolipoma Presenting with Massive Retroperitoneal Hemorrhage

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Renal angiomyolipomas can occur sporadically as solitary tumors or can be associated with tuberous sclerosis and present as bilateral multifocal disease. Modern radiographic imaging with computed tomography (CT) scan or ultrasonography can often diagnose this lesion based on its characteristic fat content. Even though it is a benign lesion, angiomyolipomas can be associated with severe symptoms, including hemorrhage. Most can be managed with nephron-sparing approaches that include observation, angiographic embolization, or partial nephrectomy. This article presents the case of a woman with a renal angiomyolipoma and perirenal hematoma treated by angiography and selective embolization.

CASE PRESENTATION

A 49-year-old woman, with no significant past medical history, presented with an acute onset of severe left-sided flank pain associated with nausea and vomiting. The pain was constant, sharp, and radiated to the left anterior abdominal wall. There was no preceding history of trauma, hematuria, dysuria, or other lower urinary tract symptoms. On examination, she was afebrile with stable vital signs (temperature, 98°F [36.6°C]; blood pressure, 126/72 mm Hg; heart rate, 66 bpm; respiratory rate, 16 breaths/min). The patient appeared in moderate distress with left abdominal and flank tenderness and voluntary guarding. Results of pertinent laboratory tests included a blood hemoglobin level of 12.2 g/dL, serum creatinine level of 1.3 mg/dL, and 5 to 10 erythrocytes per high power field on urinalysis. CT scan identified a 2.8 × 1.7 × 2.8 cm fat-containing mass extending from the midpole of the left kidney, consistent with an angiomyolipoma. In addition, a large perirenal hematoma was displacing the kidney anteromedially (Figure 1). On the second hospital day, the patient's hemoglobin level decreased to 7.6 g/dL, and

she was transfused with 2 units of packed erythrocytes. An emergent angiogram was performed and revealed multiple tortuous, dilated, hypervascular vessels emanating from the superior left renal artery (Figure 2); the abnormal vessels were selectively embolized. Post-treatment contrast injection into the left renal artery confirmed adequate embolization with resolution of the abnormal vasculature (Figure 3). The patient was discharged home after 2 days of observation with no further evidence of bleeding.

DISCUSSION

Etiology and Epidemiology

Renal angiomyolipomas are benign neoplasms composed of thick-walled blood vessels, smooth muscle, and adipose tissue in varying proportions. These lesions have been isolated in extrarenal locations, including the liver, lymph nodes, inferior vena cava, bladder, and spleen.¹ Two phenotypes predominate. The overall female/male predominance is approximately 4:1. In the first phenotype, tumors present sporadically as a single unilateral renal lesion; the mean age of onset is in the fifth or sixth decades of life.² The second phenotype, associated with tuberous sclerosis, usually presents in the third decade of life as multifocal bilateral renal masses.³ The strong female prevalence suggests a hormonal component to tumor growth.^{2,4} This hypothesis is supported by documented cases of rapid angiomyolipoma growth during pregnancy and the identification of progesterone and estrogen receptors on smooth

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Figure 1. Abdominal computed tomography scan of the case patient showing an exophytic fat-containing tumor originating from the left kidney, consistent with an angiomyolipoma.

muscle nuclei of some angiomyolipomas using immunohistochemical staining.^{5,6}

Diagnosis

The classic presentation of angiomyolipoma includes flank pain, a palpable tender mass, and gross hematuria. Nausea or vomiting, fever, anemia, and changes in blood pressure are observed less frequently.⁴ In asymptomatic patients, angiomyolipomas usually present as incidental findings on CT or ultrasound. Prior to the widespread use of these diagnostic imaging modalities, radical nephrectomy was the treatment of choice to ascertain the pathology of these lesions and control potentially fatal bleeding.¹ Today, however, angiomyolipomas are easily diagnosed noninvasively on the basis of their fat density and the absence of calcifications.^{7,8} They appear hyperechoic on sonography and measure -10 Hounsfield units or less on CT scan. In some lesions, high content of smooth muscle or intratumoral hemorrhage may make diagnosis more difficult. In these cases, the use of unenhanced thin-section CT is advocated for detection of small amounts of high attenuation fat.⁹

Several studies have demonstrated that the frequency of symptoms and risk of bleeding increases with the size of the angiomyolipoma.^{1,2,4} Quantification of tumor size and the presence or absence of specific symptoms help to distinguish patients who should undergo an intervention from those who should be observed at regular intervals. In a review of the literature, Oesterling et al reported that 82% of patients with lesions 4.0 cm or larger were symptomatic compared

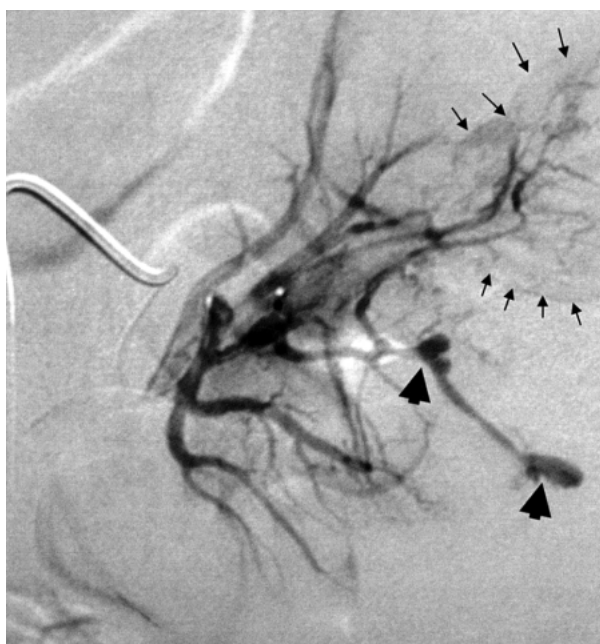


Figure 2. Selective left angiogram of the case patient demonstrating abnormal feeding vessels. An aneurysmally dilated vessel is noted inferiorly (thick arrows) with a diffuse hypervascular blush (thin arrows).

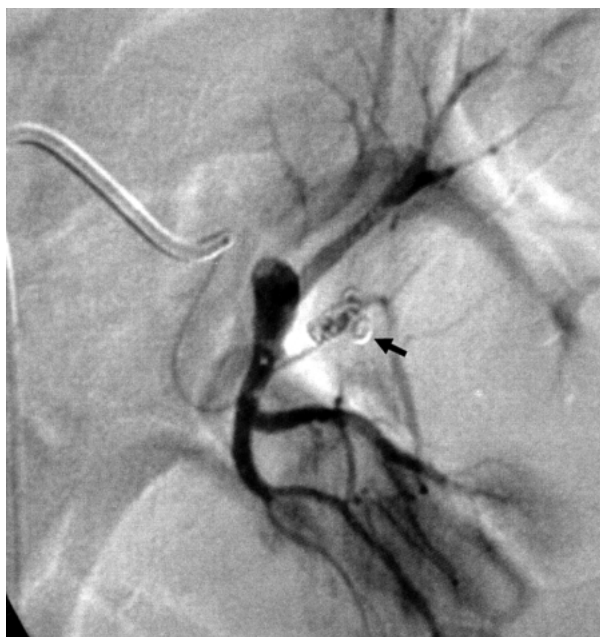


Figure 3. The lesion after embolization with microparticles (700–900 μm) and coils (3 and 4 mm) showing obliteration of the abnormal vessels.

with 23% of patients with lesions less than 4.0 cm in diameter.⁴ Signs and symptoms of angiomyolipoma included flank or abdominal pain (53%), presence of a palpable mass (47%), hematuria (26%), and hypovolemic shock (20%). Renal hemorrhage occurred in 51% of patients with lesions 4.0 cm or larger, but only 13% of patients with lesions smaller than 4.0 cm. The importance of surveillance in the management of an angiomyolipoma is underscored in the study by Steiner et al demonstrating documented growth of an angiomyolipoma in 27% of lesions smaller than 4 cm and 46% of lesions 4.0 cm or larger over a mean follow-up period of 4 years.¹⁰

Management

Current management guidelines for angiomyolipomas have been developed from observational studies and emphasize the importance of monitoring coexisting symptoms in addition to lesion size.⁸ For asymptomatic lesions smaller than 4.0 cm, intervention is not required and observation with annual CT scan is recommended. In asymptomatic lesions 4.0 cm or larger, follow-up should be more frequent, with serial CT scans every 6 months. Prophylactic embolization of asymptomatic lesions 4.0 cm or larger is recommended in select high-risk patients, including younger women who intend pregnancy or patients in which regular follow-up is difficult.² For symptomatic lesions, intervention is advised regardless of size. In all cases, the treatment goal is to preserve renal function. Currently, first-line intervention is angiography with selective embolization. Surgery is reserved for patients with persistent hemorrhage, suspicion of malignancy, and failed embolization.²

CONCLUSION

Angiomyolipomas are benign lesions of the kidney that are often asymptomatic but may be associated with severe symptoms. Large tumors and multifocal tumors, as seen in tuberous sclerosis, are more likely to present

with symptoms, including significant bleeding. First-line treatment for symptomatic or bleeding angiomyolipoma is angiography with selective embolization, as was performed on the case patient. The goal in managing angiomyolipoma is to limit intervention to tumors that are symptomatic or at high risk for bleeding, with the ultimate goal being renal preservation. **HP**

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