Initial Presentation of Tuberous Sclerosis Complex in an Adult Woman with Hemorrhage from Bilateral, Giant, Multi-Focal Renal Angiomyolipomas

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Abstract

Renal angiomyolipomas (AML’s) are benign tumors containing smooth muscle, adipose, and aberrant vasculature and can cause hypertension, renal failure and life threatening hemorrhage. In Tuberous sclerosis complex (TSC), AMLs are often bilateral and can be too numerous to count, up to 89% and 76% respectively. The disease is autosomal dominant and affects multiple anatomic systems. No single feature is diagnostic for TSC, making complete evaluation for major and minor signs requisite. We report a case of a hemorrhagic AML in a woman with giant, bilateral, multifocal AML and hypovolemic shock requiring emergent endovascular treatment of unilateral AML hemorrhage.

Keywords: Angiomyolipoma; Sirolimus; mTOR; Giant angiomyolipoma

Clinical Image

During workup for diminished appetite and weight loss, a 53 year-old woman was found to have bilateral giant AML’s. Severe anemia was discovered on labs drawn for symptoms of dizziness and fatigue; she was referred for emergency treatment. Because her eGFR was 20, she underwent a non-contrasted CT. This demonstrated mixed attenuation within an 11 cm superior, left AML and lymphangiomyomatosis, a second major and confirmatory sign of TSC (Figure 1).

After 3 units of packed red cells and fresh frozen plasma, her hematocrit did not respond. Angiogram demonstrated a large pseudoaneurysm with an arteriovenous fistula arising from the left renal artery, in the superior lateral AML. The pseudoaneurysm was embolized using Ruby® coils.

Figure 1: (a) Non-contrast coronal (b) axial CT showing bilateral angiomyolipomas essentially replacing her kidneys bilaterally with a suspected left retroperitoneal hematoma (arrow) (c) Scattered pulmonary cyst (arrows) suggestive of lymphangiomyomatosis, also seen in tuberous sclerosis.
Figure 2: (a) Angiography of the left kidney with pseudoaneurysm (arrow) arising from the superior pole of the left kidney with extravasation of contrast inferiorly. (b) Post-interventional angiography demonstrating coiled pseudoaneurysm with resolution of contrast extravasation. (c) Angiography of the left renal artery after embolization with alcohol and lipiodol showing decreased perfusion to left upper pole AML.

(Penumbra Inc., Alameda, CA). Subsequently, the artery feeding the AML of the left upper pole was embolized with alcohol and lipiodol. Post embolization imaging demonstrates the successfully treated pseudoaneurysm with appropriate blood flow to the left kidney (Figure 2).

Her creatinine, hemoglobin and hematocrit stabilized after embolization. Repeat CT demonstrated hematoma stability, and no further transfusions were required. Following discharge she was scheduled to see oncology for Sirolimus, an mTOR inhibitor, which is FDA approved to treat AMLs and may result in regression [1-4].

References


