Renal Angiomyolipomatosis in a Patient With Tuberous Sclerosis Complex

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A man with tuberous sclerosis complex (TSC) presented with fatigue and dyspnea. His abdomen was firm to palpation, and his hemoglobin level was 7.7 g/dL. Results of physical examination for gastrointestinal hemorrhage and hemolysis were unremarkable. Computed tomography demonstrated extensive angiomyolipomatosis and hemorrhages within axial (image A) and coronal (image B) reconstructions. Enlarged kidneys filled the abdominal cavity and compressed the intestinal contents into the pelvis. Heterogeneous attenuation of the renal masses was consistent with the mixed tissues of angiomyolipomatosis intercalated with hemorrhages. Adipose-rich components (long arrow), vascularized soft-tissue tumor components (short arrow), and the largest of the hematomas (star) are denoted. The abundance of lesions precluded selective embolization. The patient was treated conservatively with transfusions of 3 U (500 mL) irradiated packed red blood cells for 24 hours. There was no recurrent bleeding at 6-month follow-up. Angiomyolipomatosis in TSC has greater hemorrhage risk than non-TSC–related angiomyolipomas (AML).1-3 Although computed tomography usually differentiates AML from other masses, fat-poor subtypes may be difficult to discern.4 Without bleeding, AML generally warrants conservative management; otherwise, transcatheter embolization is preferred.5 Partial or total nephrectomy may be required in patients ineligible for embolization or when severe bleeding persists.

References

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4. Kim JK, Park SY, Shon JH, Cho KS. Angiomyolipoma with minimal fat: differentiation from renal cell carcinoma at biphasic helical CT. 