Renal Angiomyolipoma With Cardiac Extension in Patient With Tuberous Sclerosis Complex

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A 26-year-old woman with a clinical diagnosis of tuberous sclerosis complex, presenting the classical triad of seizures, mental retardation, and facial angiofibromas and reporting tachycardia, dyspnea during moderate physical activity, and 3 episodes of syncope in the past 30 days, was admitted to our hospital.

Echocardiography revealed right cardiac chambers with increased dimensions related to a large echogenic tumor with an inferior component within the inferior vena cava (Figure 1). Computed tomography of the chest and abdomen without iodinated venous contrast media depicted a large tumor arising from the right kidney with adipose tissue, extending superiorly via inferior vena cava with right cardiac chambers involvement (Figure 2). MRI of the abdomen and heart showed the same large right kidney tumor with high signal intensity in T1- and T2-weighted images, signal intensity loss in T2-weighted images with fat saturation, and enhancement after the use of venous paramagnetic contrast media (Figure 3 and Movies I, II, and III in the online-only Data Supplement).

The patient underwent surgical treatment with right nephrectomy and tumor resection. Pathology showed a angiomyolipoma with a cylindrical shape measuring 16×4×4 cm, and there were no atypical or epithelioid features seen on histological examination (Figure 4).

Renal angiomyolipoma (AML) is a benign neoplasm of the group of perivascular epithelioid cell tumors, consisting of 3 elements: smooth muscle, adipose tissue, and blood vessels. It is predominantly seen in women, and its incidence is highest in the fifth and sixth decades of life. It is well associated with tuberous sclerosis, seen in 80% of the patients. Typically, AML is confined to the kidney, but, in rare circumstances, the tumor can extend beyond the kidney.1

Although renal AML is a benign lesion, it has the potential for significant sequelae. Retroperitoneal hemorrhage from the AML can lead to shock in up to 20% of the patients. In the tuberous sclerosis complex population, renal failure can occur because the patients often require multiple procedures to manage their AMLs. Finally, the extension of a renal AML into the renal vein, inferior vena cava, or right cardiac chambers is a significant point of morbidity in this disease.2 There have only been a few case reports of AML with tumor extension to the cardiac chambers.

Renal AMLs occur as isolated, sporadic entities in 80% of cases, most commonly manifesting in middle-aged women. The other 20% of angiomyolipomas develop in association with tuberous sclerosis complex. Although the histological appearance of AMLs in these 2 entities is identical, the disease progression may not be. Angiomyolipomas that occur in association with tuberous sclerosis complex manifest at a younger age (31.5 versus 53.6 years, P<0.05), are likely to be larger and bilateral, and are prone to grow and need surgical treatment.3

Disclosures

None.

References

Figure 1. Echocardiography imaging. A, Transverse subcostal view showing an echogenic mass (star) in the right atrium. B, Apical 4-chamber view depicting the mass within the right atrium with extension to the right ventricle through the tricuspid valve. Transverse (C) and long-axis (D) parasternal views depicting the mass in the right ventricle (star) and in the right ventricle outflow tract, respectively.

Figure 2. Computed tomography imaging without venous contrast media, axial images at the level of the right kidney (A) and at the level of the heart (B) demonstrating a fat-containing renal mass with extension to the right cardiac chambers involving the inferior vena cava showed in coronal (C) and sagittal (D) multiplanar reconstruction.
Figure 3. MRI, axial T1 GRE (A), axial T2 turbo spin echo with fat saturation (B), and axial T1 GRE with gadolinium showing the renal origin of the angiomyolipoma (C). Axial double-inversion recovery images at the level of the intrahepatic inferior vena cava (D), right cardiac chambers (E), and right ventricle outflow tract (F) showing a large tumor with high signal intensity within those structures. The same sequence of images in coronal (G) and sagittal planes (H) showing the inferior and superior extension of the tumor. Coronal triple-inversion recovery (fat saturation) image (I) demonstrating loss of signal intensity of the tumor.

Figure 4. Angiomyolipoma. A, Macroscopy. B, Hematoxylin and eosin 40x stain shows tumor composed of smooth muscle (box), fat (star), and thick-walled blood vessels (arrow head). Immunohistochemistry shows positivity for Desmin (C) and Melan-A (D).
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