Cardiac Lipoma in a Patient with Tuberous Sclerosis Complex

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Clinical History
A 14-year-old young woman with a longstanding history of seizures was diagnosed with tuberous sclerosis. During her work-up, an echocardiogram demonstrated an echogenic mass between the aorta and the main pulmonary artery, likely pericardial in location (Figure 1). A CT of the chest was requested to further evaluate this finding.

Findings
The CT images show an intra-myocardial hypo-attenuating mass consistent with a lipoma (Figures 2), which corresponds to the hyperechoic mass initially localized to the pericardium by echocardiogram. MR imaging confirmed that the lesion was composed of fat, given iso-intensity to fat on all sequences (Figure 3A) and intra-lesion signal loss on the fat-suppressed sequences (Figure 3B).

Discussion
Tuberous sclerosis complex (TSC), or Bourneville-Pringle syndrome, arises due to mutations in the TSC tumor suppressor genes, which cause abnormal cellular differentiation and proliferation. The classic triad in TSC is facial angiofibromas, mental retardation and seizures, although all three features are only seen in 30% of cases. Patients with TSC have an increased incidence of cardiac neoplasms with rhabdomyomas seen in 50-65% (1). Cardiac lipomas generally do not occur as part of a syndrome, although there have been reports of multiple cardiac lipomas in pediatric patients with TSC (2).

Cardiac lipomas are benign neoplasms most commonly located in the pericardial space or within any cardiac chamber. When myocardial, they occur most frequently in the right atrium, left ventricle, and interatrial septum. They are typically large, broad-based and do not demonstrate calcification, hemorrhage or necrosis (3). On echocardiography, they are usually hypoechoic when in the pericardial space but echogenic when located in a cardiac chamber. Cardiac lipomas demonstrate homogeneous fat attenuation on CT, display homogeneous high T1 signal intensity on MRI and do not enhance.

Cardiac lipomas are often an incidental finding on CT and are typically asymptomatic. Due to its superb wide field of view and spatial resolution, a dedicated cardiac CT may be useful in patients with suspected TSC. Cardiac CT may be particularly useful in defining the relationship of these masses to coronary vessels and valves, as well as the great vessels.

REFERENCES
3. Araoz PA, Mulvagh SL, Tazelaar HD, Jilsrud PR and Breen JF. CT and MR Imaging of Benign Primary Cardiac Neoplasms with Echocardiographic Correlation. Radiographics 2000;20:1303-1319