A 61-year-old woman with no risk factors for cardiovascular disease presented to her local hospital with symptoms of exertional angina. The resting ECG was unremarkable, but during exercise treadmill testing, the patient had her usual symptoms associated with >2-mm downsloping ST-segment depression in the inferolateral leads, which took 13 minutes to resolve. Urgent coronary angiography revealed a normal left main stem, left anterior descending artery, and circumflex artery (Figure 1; online-only Data Supplement Movie I), but right coronary artery cannulation proved impossible. The patient had chest pain during the procedure, and an aortogram was performed both to exclude dissection.

Figure 1. A, Coronary angiogram showing normal left main stem, left anterior descending artery, and circumflex artery (right anterior oblique view). B, Aortagram showing subtle circular filling defect adjacent to the aortic valve (white arrow). C, D, and E, Contrast-enhanced 64-slice spiral computed tomography scan. A low-attenuation, pedunculated mass with a mean of 56 Hounsfield units is seen attached to the left coronary cusp of the aortic valve in close proximity to the origin of the left main stem (arrows). F, Small right coronary artery arising above the aortic sinuses between the aorta and pulmonary artery (arrowheads).
and to attempt to identify the right coronary artery (Figure 1; online-only Data Supplement Movie II). There was no dissection, but the right coronary artery was still not seen. As a result, contrast-enhanced 64-slice computed tomography coronary angiography was requested. This showed a small, anomalous right coronary artery but also revealed a well-defined low-attenuation filling defect consistent with a mass adjacent to the left coronary cusp of the aortic valve (Figure 1). To obtain better tissue characterization of this mass, cardiovascular magnetic resonance was performed. This demonstrated a mobile, noncalcified spherical structure attached to the aortic valve with intermediate signal intensity on T1, high signal on T2-weighted sequences, and no suppression using fat saturation. There was enhancement after gadolinium contrast injection (Figure 2; online-only Data Supplement Movies III and IV). The mass was seen on a subsequent transthoracic echo study and clearly visible on 3-D transesophageal echocardiography before surgical removal (Figure 3; online-only Data Supplement Movies V and VI), which confirmed its mobile nature, the close proximity to the left main stem, and additionally identified Lambl excrescences on the tip of the valve leaflet.

During surgery to remove the tumor, direct coronary cannulation was required to achieve cardioplegia because the mass had prolapsed deep into the left main stem. The right coronary artery was confirmed to be an anomalous small vessel supplying only the right ventricle. The tumor, a cream-colored mass with numerous outer papillary projections, was excised with a thin slither of the valve leaflet. Further small Lambl excrescences were removed from the

Figure 2. Cardiovascular magnetic resonance scan images (Siemens Avanto, 1.5 T) showing a spherical mass attached to the left coronary cusp of the aortic valve (arrows). A and B, Steady-state free precession (TrueFISP) images. C, Intermediate signal intensity on T1-weighted spin-echo sequence; D, High signal intensity on T2-weighted spin-echo sequence with fat saturation (short τ inversion recovery–STIR T2). E and F, Enhancement of the mass after intravenous injection of gadolinium DTPA.

Figure 3. Three-dimensional transesophageal echocardiography images. A, Short-axis view of the aortic valve showing the proximity of the mass (white arrow) to the origin of the left main stem (black arrow). B, Long-axis view of the aortic root showing the mass (arrow) and a large Lambl excrescence attached to the tip of the aortic valve leaflet (arrowhead).
right and noncoronary cusps. Histology revealed finger-like processes containing elastic tissue and dense collagen, covered by a single endothelial layer (Figure 4), typical of a fibroelastoma. The patient made a complete recovery and has remained symptom-free.

Papillary fibroelastoma, the third most common primary cardiac tumor after myxoma and lipoma, is a benign tumor of the endocardium that primarily affects the valves. It represents 10% of all primary cardiac tumors.\(^1\) The aortic valve is the most often involved (44.5%), followed by the mitral valve (36.4%) and left ventricular outflow tract (18%). There is a high propensity for systemic embolization leading to neurological symptoms (transient ischemic attack, stroke, retinal emboli), coronary ischemia (including coronary occlusion and sudden death), or peripheral embolization. The gross specimen has branching fronds reminiscent of a sea anemone, and the characteristic histology gives the definitive diagnosis. Each frond has an outer layer of endothelial cells with layers of mucopolysaccharides and an inner core of connective tissue (containing collagen, smooth muscle cells, and elastic fibers). No clear underlying causative process has been identified, but the histological findings highlight the similarity between Lambl excrescences and papillary fibroelastoma.\(^2\)

This case demonstrates the power of state-of-the-art multimodality imaging not only regarding the anatomic and tissue characteristics but also the functional aspect of a pedunculated tumor causing obstruction to coronary blood flow. The sequence and number of imaging modalities used are particular to this case, and although fibroelastomas can often readily be identified using echo alone, specific imaging characteristics distinguish them from other cardiac lesions (highly echo-dense on 2D echocardiography and reported to have a computed tomography attenuation of 52 to 69 Hounsfield units). On cardiovascular magnetic resonance, the typical features are of intermediate signal on T1 and intermediate-to-high signal on T2-weighted spin-echo sequences, no suppression with fat saturation, and strong enhancement after gadolinium contrast injection.\(^3\) Once detected, surgical excision is recommended, especially for left-sided lesions in symptomatic patients because of the risk of emboli. Complete resection is curative, and conservative, valve-sparing operations may be performed with “shave excision” of part of the valve leaflet. If the patient is not a surgical candidate, then long-term anticoagulation is suggested.

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References


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