Papillary Fibroelastoma and Lambl's Excrescences: Echocardiographic Diagnosis and Differential Diagnosis

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papillary fibroelastoma, *Lambl's excrescences*

A previously healthy 44-year-old male was admitted with a history of two transient ischemic attacks (TIA) during the previous month. Transthoracic echocardiography (TTE) revealed normal cardiac dimensions and left ventricular function, however, a 1-cm mobile mass was noted attached to the tip of the posterior mitral valve leaflet (Fig. 1). This mass was

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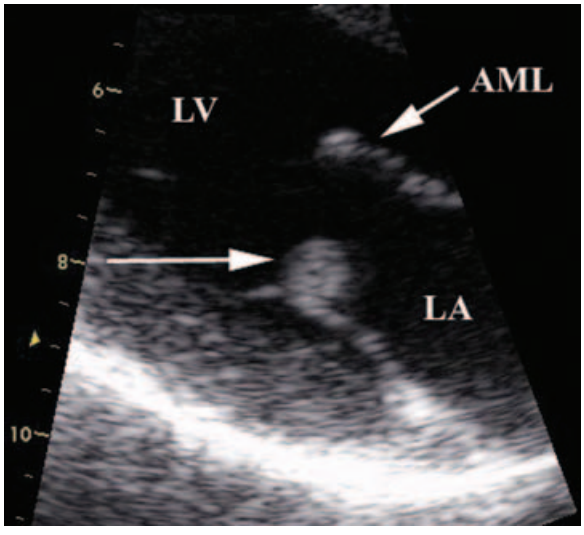

Figure 1. *Diastolic parasternal long-axis image demonstrates a 1-cm rounded mass (horizontal arrow) attached to the tip of the posterior leaflet of the mitral valve. AML = anterior leaflet mitral valve; LA = left atrium; LV = left ventricle.*

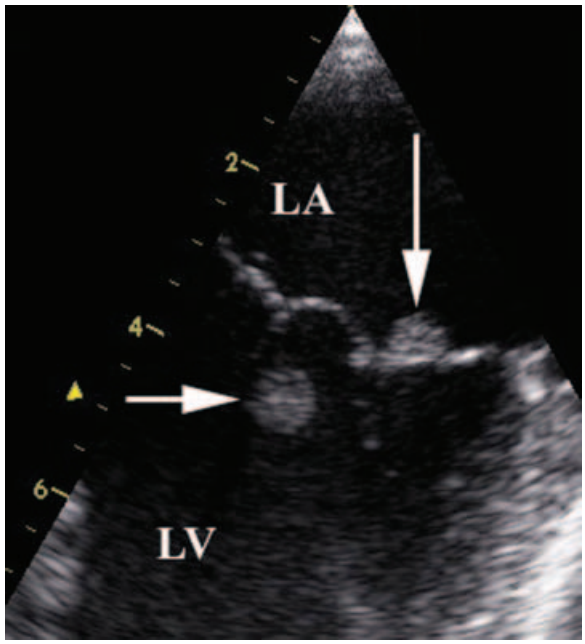

Figure 2. *Systolic TEE frame at 72° in the mid-esophagus demonstrates the mass shown in Figure 1 attached to the posterior mitral leaflet (horizontal arrow), but also another mass attached to the atrial side of the anterior mitral leaflet (vertical arrow). LA = left atrium; LV = left ventricle.*

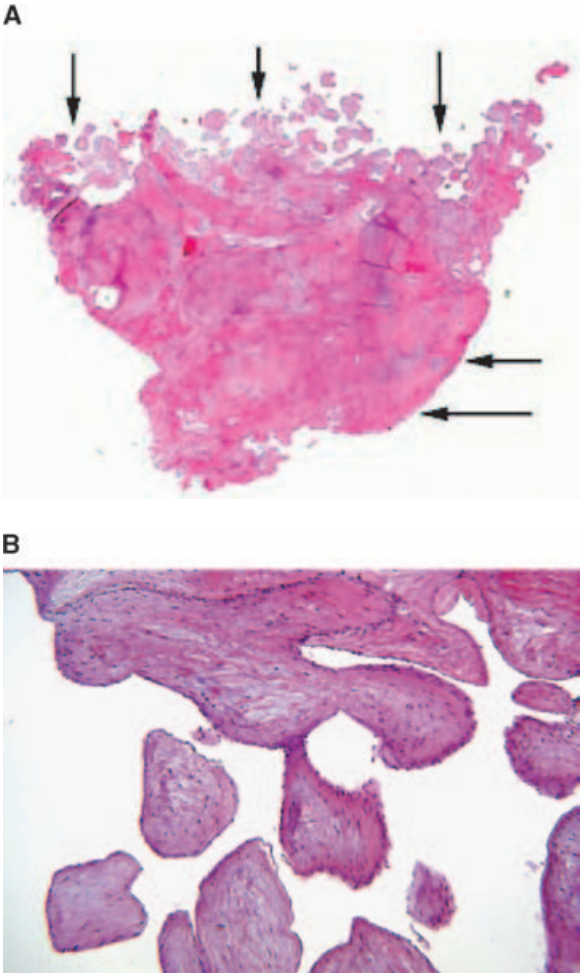

Figure 3. *Hematoxylin-eosin stain of one of the masses removed from the mitral valve at the time of surgery.* ***A.*** *Low power (1×) demonstrates the tumor is composed of a dense base (horizontal arrows) of fibroelastic tissue with papillary fronds (vertical arrows).* ***B.*** *Magnified power (10×) of the papillary fronds composed of fibroelastic tissue covered by endothelium.*

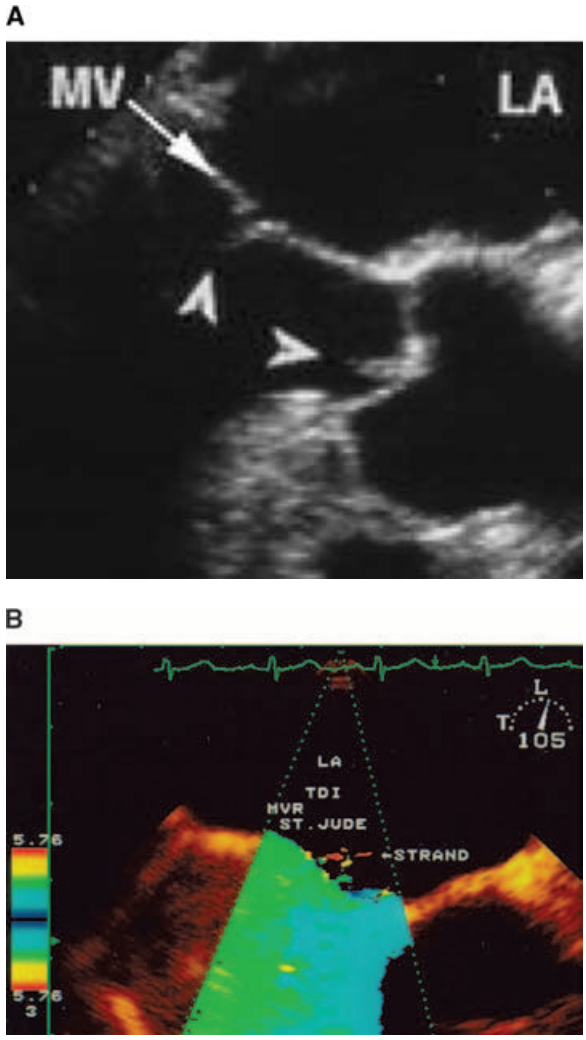

Figure 4. ***A.*** *TEE in the mid-esophagus (157°) of a patient with an aortic valve Lambl's excrescence (horizontal marker).* ***B.*** *Systolic TEE frame (105°) in the mid-esophagus of a patient with a St. Jude mechanical mitral prosthesis. Using tissue Doppler imaging (TDI), a mobile valve strand is noted (arrow—red color strand) in the left atrium (LA). (Fig. 4A modified with permission from Ref. 5).*

also noted by transesophageal echocardiography (TEE), but another smaller immobile mass was also found attached to the atrial surface of the anterior leaflet of the mitral valve (Fig. 2). Mitral valve replacement was then performed. Histopathology revealed both masses to be that of papillary fibroelastoma (Fig. 3).

Papillary fibroelastomas are the most common primary tumors of the cardiac valves. This benign tumor is covered by endothelium surrounding loose connective tissue made up of an acid mucopolysaccharide matrix, smooth muscles, collagen, and elastic fibers.[1] These tumors have been found on all four cardiac valves, but in adults are most often found on the aortic valve (usually ventricular surface) and also the mitral valve (atrial surface). Rarely they are attached to the subvalvular apparatus of the mitral or tricuspid valve, and very rarely to the free wall of a ventricular chamber.[2] In children, the tumor is most often noted on the tricuspid valve.[1]

A papillary fibroelastoma may serve as a nidus for platelet and fibrin aggregation, leading to arterial embolism, but most are found at autopsy as an incidental finding. Some

investigators have recommended anticoagulation once a papillary fibroelastoma is noted by echocardiography. There appear to be no firm data to support or refute surgical removal of a papillary fibroelastoma in an asymptomatic patient.[3]

Echocardiographic characteristics of a papillary fibroelastoma include:

1. most often the tumor is solitary
2. usually <1 cm in diameter, but may become 3–4 cm in size
3. the tumor usually arises from the midportion of valve leaflets (a fibrous strand usually arises from the line of closure)
4. often pedunculated (occasionally sessile) with high-frequency oscillations during the cardiac cycle
5. characteristic "frond-like" appearance[3]

In contrast to papillary fibroelastoma, Lambl's excrescences (fibrous strands) are best identified by TEE as fine thread-like strands arising on the line of closure (contact surface) of heart valves. Most commonly, they occur on the mitral, followed by aortic valve (Fig. 4). These strands have been described on prosthetic valves, and rarely on native tricuspid or pulmonic valves.[4,5] They are acellular strands covered by a single layer of endothelium. Found in 70–80% of adults, they are multiple in >90% of affected hearts.[6] Most often these strands are not associated with cardioembolic events.[7]

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