Multiple Papillary Fibroelastomas

Satya S. Vittala, MBBS; Roger L. Click, MD, PhD; Suresh Challa, MD; Mohammad Q. Najib, MD; Bijoy K. Khandheria, MD; William D. Edwards, MD; Joseph J. Maleszewski, MD; Hari P. Chaliki, MD

A 53-year-old woman who had a history of mitral valve prolapse was referred to our institution after an incidental finding of small masses on the mitral, tricuspid, and pulmonary valves on transthoracic echocardiography. She had no symptoms suggestive of endocarditis with negative blood cultures. Complete blood cell count and erythrocyte sedimentation rate were within normal limits.

The patient underwent transesophageal echocardiography, which showed a pedunculated, papillary mass on the tricuspid valve (Figure 1 and Movie I in the online-only Data Supplement) with mild tricuspid regurgitation. In addition, 2 sessile masses were noted on the pulmonary valve (Figure 2 and Movie I in the online-only Data Supplement) and the posterior mitral valve leaflet (Figure 3 and Movie I in the online-only Data Supplement). She underwent excision of the masses on the tricuspid (Figure 4), mitral (Figure 5), and pulmonary valves, which appeared to be papillary fibroelastomas (PFEs) on gross appearance. Histopathologic examination (Figure 6) of the excised masses showed avascular, endothelial-lined fronds arising from central stalks, diagnostic of PFE. Because of posterior mitral leaflet prolapse with moderately severe mitral regurgitation and moderate aortic regurgitation, the patient also underwent mitral annuloplasty and aortic valve repair. Postoperative recovery was uneventful. The patient had no evidence of recurrence of PFEs at her 1-month follow-up visit.

Figure 1. Transesophageal partial 4-chamber view showing the mobile echocardiographic density on the tricuspid valve (TV) (arrow) consistent with papillary fibroelastoma. RA indicates right atrium; RV, right ventricle.

Figure 2. Transesophageal short-axis view showing mobile density attached to the pulmonary valve (PV) (arrow) with a broad base, consistent with papillary fibroelastoma. Ao indicates aorta; TV, tricuspid valve.

Figure 3. Transesophageal 4-chamber view showing the mobile echocardiographic density on the mitral valve (MV) (arrow), consistent with papillary fibroelastoma. Ao indicates aorta; LA, left atrium; LV, left ventricle.
The majority (77%) of PFEs are seen involving the valvular surfaces of the heart. Most commonly, they involve the aortic valve (36.4%), followed by the mitral valve (29.4%), the tricuspid valve (10.6%), and, last, the pulmonary valve (7%). On echocardiography, they usually are speckled with echolucencies and have a stippled pattern near the edges with shimmer, which correlates with the papillary projections on the tumor surface. Approximately 43.6% of PFEs are mobile, and multiple PFEs are found in only 6.1% to 8.6% of the total presentations in same or different locations of the heart. The right side of the heart rarely is involved, unlike for our patient, in whom both right-sided valves of the heart were involved with no history of cardiac trauma, such as surgery. Although patients are asymptomatic typically, they have a risk of embolization of the tumor fragments, and hence surgical treatment with simple shave excision is the treatment of choice, depending on the size, location, and surgical risk. Patients have an excellent long-term prognosis after surgery.

Disclosures

None.

References


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Figure 4. Surgical view of the tricuspid valve (TV) with sessile mass (arrow), confirmed to be papillary fibroelastoma through histopathologic evaluation.

Figure 5. Surgical view of the mitral valve (MV) showing a pedunculated mass (arrow), confirmed to be papillary fibroelastoma through histopathologic evaluation.

Figure 6. Photomicrographs exhibiting numerous avascular, endocardial-lined fronds from masses resected from the (A) pulmonary valve (Verhoeff-Van Gieson; original magnification, ×250) and (B) tricuspid valve (Verhoeff-Van Gieson; original magnification, ×100).
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