Case 190: Papillary Fibroelastoma of the Pulmonary Valve

A 64-year-old male nonsmoker with a history of hypertension and morbus Ménière (Ménière disease) who had undergone a previous appendectomy presented with fatigue and bradycardia. Laboratory tests did not reveal any relevant abnormalities. At physical examination, normal breath sounds and no heart murmurs were heard. Chest radiography demonstrated a normal cardiac silhouette and clear lung fields. Electrocardiography showed sinus rhythm with a normal axis. The indication for transthoracic echocardiography was evaluation of bradycardia of unknown origin. Because of the suggestion of a structure closely related to the pulmonary valve, transesophageal echocardiography was performed. For further tissue characterization, cardiac magnetic resonance (MR) imaging also was performed, and the resultant images are shown.

Imaging Findings
Transesophageal echocardiography revealed a 1-cm mobile round mass attached to the posterior leaflet of the pulmonary valve (Fig 1). During systole, the entire mass prolapsed into the main pulmonary trunk. There was also an accompanying mild (grade 1 of 4) pulmonary regurgitation. Most of the lesion was hyperechoic, but a few echolucencies were suspected within the mass. In addition, a three-dimensional transesophageal echocardiographic examination was performed (Fig 2; Movie 1 [online]). These images show even better the prolapse during systole in the right ventricular outflow tract and the slightly irregular contour of the small mass.

For further evaluation of the tissue characteristics of the mass, cardiac MR imaging was performed. Figures 3 and 4 show a 1.0 × 0.7 cm soft-tissue mass attached to the ventricular side of the pulmonary valve. The T1-weighted images show a mass with homogeneous signal intensity that is mostly isointense to that of cardiac muscle. T2-weighted images were not obtained. After gadolinium pentetic acid administration, the mass showed strong enhancement. At cine imaging, the mass oscillated with the motion of the pulmonary valve (Movie 2 [online]). Both left and right ventricular functions were normal at visual inspection. The left ventricle ejection fraction was 66%. No phase-contrast flow-mapping sequences were performed to evaluate pulmonary regurgitation at MR imaging.

Discussion
The differential diagnosis of a mass closely related to the pulmonary valve includes papillary fibroelastoma, valvular myxoma, nonbacterial thrombotic endocarditis, and infective vegetation in endocarditis.
Primary neoplasms of the heart are rare and have a reported prevalence in autopsy series that varies from 0.002% to 0.3% (1). The majority of primary cardiac tumors are benign. Patients with primary cardiac tumors can present with a wide range of symptoms; the clinical presentation is influenced by tumor location, size, growth rate, embolism tendency, and degree of invasiveness.

Three out of four valvular tumors are papillary fibroelastomas (2). Papillary fibroelastomas are the second most common form of cardiac tumor, and 55% of patients with this tumor are male. The highest prevalence is in octogenarians. A valvular location is seen in 86% of cases. The tumor is located at the aortic valve in 44% of cases, followed by the mitral valve (35% of cases), tricuspid valve (15% of cases), and pulmonary valve (8% of cases). A nonvalvular location is seen in 16% of cases (2). There are only a few descriptions of fibroelastomas at MR imaging (1,3–5). A fibroelastoma appears as an isointense mass, often with a stalk on T1-weighted images. Signal intensity is hyperintense on T2-weighted images (3). On late-enhancement images, the lesion demonstrates homogeneous high signal intensity (4). On cine images, it appears as a hypointense mobile mass and is often mobile and oscillating with every heartbeat (5).

Myxomas are the most common benign cardiac tumors, comprising 50% of all cardiac tumors (6). The majority of myxomas are attached to the fossa ovalis of the interatrial septum, but they can also be attached to the walls of the cardiac chambers or valve leaflets (7,8). Myxomas mostly manifest in adulthood, between the fourth and seventh decade. A female predominance has been reported (9). Myxomas with an irregular surface have the tendency to induce surface thrombi formation and embolism (10). At MR imaging, myxomas demonstrate heterogeneous signal intensity because of the underlying tissue heterogeneity. Myxomatous tissue is hypointense relative to the myocardium on T1-weighted images (1). On T2-weighted images, it is hypointense because of the high water content. The fibrous tissue is hypointense on T2- and T1-weighted images (1). After administration of gadolinium pentetic acid, myxomas typically show a heterogeneous enhancement pattern; however, they occasionally show a homogeneous enhancement pattern. On late-enhancement images, myxomas demonstrate heterogeneous high signal intensity. During cine imaging, mobility characteristics and possible valve prolapse can be evaluated (1,5).

Also represented in the differential diagnosis is valvular vegetation, as seen in patients with infectious endocarditis. Patients with (bacterial) valvular vegetation mostly have at least one predisposing factor, such as intravenous drug use, alcoholism, previous right-sided catheterization, or congenital heart disease. The pulmonary valve is least involved
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Most patients present with fever, shortness of breath, or pleuritic chest pain due to small septic infiltrates. At echocardiography, shaggy irregular echoes are seen on or near one or two pulmonary cusps, and motion can be restricted, causing pulmonary regurgitation (11,12). Few cases of vegetation at cardiac MR imaging have been described (11,13). At cardiac MR imaging, functional deficits (regurgitation or stenosis) and low-intensity valvular vegetation are seen on the bright-blood fast gradient-echo images. On spin-echo MR images, the vegetations are not visible (13).

Nonbacterial thrombotic endocarditis (NBTE) is a disease characterized by valvular vegetations, which consist of fibrin and platelet aggregates and which are devoid of inflammation or bacteria (14). Some patients present with a heart murmur or evidence of embolism (15,16). NBTE has been reported in patients with advanced-stage malignancy, hematologic disorders, connective tissue disease, autoimmune systemic lupus erythematoses, AIDS, and hypercoagulable-state disorders (14). Trauma from indwelling pulmonary or central venous catheter, overdose, snakebite, or late effect of radiation therapy has also been reported. In transthoracic and transesophageal echocardiography, the vegetations in patients with NBTE are dense, small (mostly smaller than 1.0 cm), broad based, and irregular in shape (16). No reports on cardiac MR imaging are available.

In this patient, none of the previously mentioned predisposing features for infectious endocarditis or NBTE were present. Thus, because of the patient’s unremarkable cardiovascular history, the most likely diagnosis is papillary fibroelastoma or valvular myxoma. Echocardiography showed a round mobile lesion attached to the posterior leaflet of the pulmonary valve, which enabled us to confirm the presence of a tumor-like lesion instead of infection or debris. Although the pulmonary valve is an infrequent location for both fibroelastoma and valvular myxoma, these entities do occur in this location. No further differentiation between the two entities could be made based on the echocardiographic findings. On the T1-weighted MR images, the lesion was predominantly homogeneous, and it was isointense when compared with cardiac muscle. On the basis of these findings, fibroelastoma is a more likely diagnosis. T2-weighted sequences may show high signal intensity (5). After contrast material administration, there was strong immediate enhancement of the lesion. No evidence of a stalk, which occurs often with a fibroelastoma, could be identified with the various imaging modalities. In this patient, it was not possible to definitely differentiate between a valvular myxoma and a papillary fibroelastoma.

This patient underwent resection of the lesion via median sternotomy, with...
resection of the valvar mass (Fig 5). The mass was easily removed from the valve without damaging the posterior leaflet. No complications occurred, and the patient was discharged from the hospital 5 days after surgery. Postoperative transesophageal echocardiography did not show any pulmonary regurgitation or pericardial effusion. Histopathologic examination of the excised mass revealed multiple frondlike structures comprised of a dense fibroelastin core with surrounding loose connecting tissue. The correct diagnosis is papillary fibroelastoma of the pulmonary valve.

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References


Congratulations to the 104 individuals and five resident groups that submitted the most likely diagnosis (papillary fibroelastoma of the pulmonary valve) for Diagnosis Please, Case 190. The names and locations of the individuals and resident groups, as submitted, are as follows:

Individual responses

Osamu Abe, MD, PhD, Itabashi-ku, Tokyo, Japan
Stephane Aillaud, MD, Aix En Provence, Bouches De Rhone, France
Harry A. Allen III, MD, Virginia Beach, Va
Guiz S. Astacio, MD, Rio de Janeiro, Brazil
Paul D. Bailey, MD, Baltimore, Md
Kenneth F. Baliga, MD, Tokyo, Japan
Ian A. Burgess, MD, North Sydney, New South Wales, Australia
Daniel Castellon, PhD, Vigo, Pontevedra, Spain
Antonio A. Cavalcanti, MD, Sao Paulo, Brazil
Philip M. Cheng, MD, MS, Culver City, Calif
Perry Choi, MD, Richmond Hill, Ontario, Canada
Christopher Chu, MBBS, FRANZC, Sydney, New South Wales, Australia
Marco A. Curu, MD, Highland Park, Tex
Peter de Baets, MD, Damme, Belgium