Multimodality Imaging in the Diagnosis of Coexisting Left Atrial Myxoma and Aortic Valve Papillary Fibroelastoma

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We present the case of a 77-year-old Cambodian woman with a 3-day history of dyspnea on exertion. Physical examination revealed a murmur of mitral regurgitation and decompensated heart failure. The ECG was remarkable for atrial fibrillation with rapid ventricular response. Transthoracic echocardiography showed severe left atrial dilation, a large (5×8 cm) mobile mass attached to the anterior left atrial wall, and several small, mobile aortic masses on the noncoronary cusp of the aortic valve (Figure 1 and online-only Data Supplement Movie I).

Three-dimensional transesophageal echocardiography showed a large multilobular mass being displaced by the central jet of moderate to severe mitral regurgitation during systole and prolapse of the mass through the mitral valve during diastole (Figures 2 and 3 and online-only Data Supplement Movies II and III). The atrial mass was attached with a stalk to the anterior atrial wall and appeared lobulated and deformable, consistent with the appearance of a myxoma (online-only Data Supplement Movie IV). The aortic valve masses had multiple frondlike projections, which had an appearance like a sea anemone, with stippling at the edges that arose from the noncoronary aortic valve leaflet (online-
only Data Supplement Movies IV and V). Preoperative coronary computed tomography angiography was performed to rule out coronary disease, which revealed mild coronary artery disease and (not shown) again demonstrated the classic appearance of both benign tumors as seen on echocardiography (Figures 4 and 5).

The combination of information gained by multimodality imaging allowed for optimal detection and evaluation of likely tumor origins and subsequent planning of the surgical approach and double-tumor excision. The patient underwent right atriotomy to excise the left atrial mass with clear margins (Figure 6) and transverse aortotomy to excise the aortic leaflet masses. The differences in surgical approaches emphasize the importance of being able to differentiate the tumors preoperatively.

Pathological analysis revealed that the left atrial mass was consistent with an atrial myxoma, whereas the aortic valve masses were consistent with papillary fibroelastoma (PFE). The myxoma had hyperchromatic, multinucleated myxoid cells (Figure 7), embedded in light pink extracellular matrix, whereas the PFE showed pink hyalinized stroma encircled by endothelial cells (Figure 8).

Myxomas are primary cardiac adult neoplasms predominantly found in the left atrium. They are almost always single and located near the fossa ovalis. Approximately 10% of myxomas are familial, as part of the autosomal dominant Carney complex, whereas the cause of sporadic cases remains unclear. Multilobulated or pedunculated myxomas are often sufficiently mobile and may prolapse into the mitral valve during diastole, causing potential structural damage to the valve. Surgical excision is recommended because of potential embolization.1 PFEs are rare lesions that can be asymptomatic or lead to cardioembolic stroke and death.2 Although the pathogenesis of PFEs is still under investigation, it is thought that they develop in areas of injury or stress. Surgical excision in symptomatic patients with lesions on the left side of the heart is considered necessary and effective, although asymptomatic lesions on the right side of the heart may simply be observed.2

A review of literature for possible links between the lesions yielded only 4 prior reports to date of coexisting cardiac myxoma and PFE. One case reported coexisting cardiac myxoma and PFE each on the anterior leaflet of the mitral valve.3 Another report suggested that myxoma may serve as a nodal lesion for
Interestingly, myxomatous foci have been noted within PFEs, and granular elastotic material has been observed in atrial myxomas, a finding typically consistent with PFEs. Thus, a histogenetic origin potentially underlies the presence of these 2 cardiac tumors and may serve as an additional pathophysiological explanation when they are found in the same patient.

The integration of newer cardiac imaging techniques proved critical in the management of our patient. A diagnosis was made noninvasively without the need for invasive and potentially risky biopsies. Both 3-dimensional transesophageal echocardiography and coronary computed tomography angiography are newer noninvasive imaging modalities that serve as alternatives for the evaluation and diagnosis of cardiac tumors. In the present case, possible malignant extension from one tumor to the other was suspected on clinical grounds; however, the imaging modalities used demonstrated no invasion of tissue planes, thereby rendering the possibility of a malignancy much less likely. We anticipate that the preferred approach for cardiac tumor detection and for decisions regarding management and surgical planning will be through the integration of noninvasive cardiac imaging techniques.

Disclosures
Dr Vorobiof is on the Speakers’ Bureau for Lantheus Medical Imaging. The other authors report no conflicts.

References
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