An Unusual Cardiac Fibroelastoma Case

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A 12-year-old boy presented with paroxysmal chest pain for 2 months, and sudden onset of syncope lasted ≈2 minutes. On transthoracic echocardiogram, the parasternal long-axis view showed an echo-dense mass attached to the left ventricular anterolateral papillary muscle, extending to the anterior mitral valve chordae, the left ventricular outflow tract, and the aortic root (Figure 1A and Movie I in the online-only Data Supplement). The apical 5-chamber view demonstrated that the mass in the left ventricular outflow tract extended into the aortic root through the aortic valves (Figure 1B). The apical 4-chamber view illustrated that the mass was on the mitral valvular chordae and papillary muscle (Figure 1C and 1D and Movie II in the online-only Data Supplement). The series of parasternal long-axis views showed that the mass in the aortic root was close to the orifice of the right coronary artery (RCA) and swung into the RCA during the cardiac cycle (Figure 2 and Movie I in the online-only Data Supplement). Left ventricular function was normal, with ejection fraction of 61%.

The cardiac computer tomographic coronal view showed that a large mass with a long tail extended from the left mid-ventricular area via the left ventricular outflow tract into the ascending aorta through the aortic valve (Figure 3A, black arrows). The computer tomographic cardiac long-axis view clearly illustrated that the mass on the papillary muscle extended to the mitral valvular chordae, appeared in the aortic root, and invaded the right Valsalva sinus (Figure 3B). The computer tomographic short-axis view showed that the mass was on the mitral valvular chordae and the anterolateral papillary muscle (Figure 3C).

During the operation, the surgeon found 1 large mass with a frond-like appearance that resembled a sea anemone on the papillary muscle with a long tail, which extended to the anterior mitral valvular chordae, the left ventricular outflow tract, and the aortic root through the aortic valve (Figures 4 and 5). Part of the mass in the aortic root invaded the RCA. The histopathological diagnosis of the mass was left ventricular cardiac papillary fibroelastoma (CPF).

CPF is a rare primary cardiac neoplasm, particularly in teenagers.1 Although CPF is histologically a benign tumor, it may result in life-threatening complications such as stroke, myocardial infarction, pulmonary embolism, and sudden death, although the frequency with which that occurs is not well established.2,3

The teenage boy in our case presented with angina and syncope, which may be life threatening and are indications for surgery. Figure 2 and Movie I in the online-only Data Supplement show that the mass in the aortic root was close to the orifice of the RCA and swung into the RCA during the cardiac cycle; the boy’s symptoms could be attributable to temporary obstruction of RCA by the mass in the aortic root. The report with the largest number of pathologically confirmed CPF cases included 168 patients.4 Echocardiographically, this entity has been described as a small, well-delineated, pedunculated mass with a predilection for valvular endocardium.1 However, CPF that presents as a large tumor that extends into multiple structures in the heart, as in this case, is very rare. This irregular tumor was well demarcated and homogeneous on echocardiography; these characteristics could be differentiated from the rich blood vessels of malignant tumors. Decisions regarding the primary surgical excision of CPF depend on the size, location, mobility, and potential of the tumor or the strength of the association of the tumor with symptoms, as in our case.

Disclosures

None.

References


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Figure 1. **A**, The parasternal long-axis view showed an echo-dense mass attached to the left ventricular anterior papillary muscle (closed arrows) that extended to the anterior mitral valvular chordate (circular arrows), left ventricular outflow tract, and aortic root (open arrows). **B**, The apical 5-chamber view demonstrated that the mass in the left ventricular outflow tract extended into the aortic root through the aortic valve. **C** and **D**, The apical 4-chamber view illustrated that the mass was on the papillary muscle and extended to the mitral chordae.

Figure 2. The series of parasternal long-axis views (**A** through **D**) illustrated that the mass (diamond arrows) in the aortic root was close to the orifice of the right coronary artery (open arrows) and swung into the right coronary artery during the cardiac cycle. AO indicates aorta; LA, left atrium; LV, left ventricle; and RV, right ventricle.

Figure 3. **A**, Cardiac computed tomographic coronal view showed that a large mass with a long tail extended from the left midventricular area via the left ventricular (LV) outflow tract into the ascending aorta (AO) through the aortic valve. **B**, The computed tomographic cardiac long-axis view clearly illustrated that the mass was on the papillary muscle, extended to the mitral valvular chordae, appeared in the aortic root, and invaded the right Valsalva sinus (star). **C**, The computed tomographic short-axis view showed that the mass was on the papillary muscle and mitral valvular chordae. LA indicates left atrium; and RV, right ventricle.
Figure 4. Appearance of the cardiac mass (arrow) during operation.

Figure 5. Gross appearance of the mass. Part 1 was on the papillary muscle, part 2 was on the mitral chordae, part 3 was in the left ventricular outflow tract, and part 4 was in the aortic root.
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