CASE REPORTS

Two-Dimensional Echocardiographic Detection and Diagnostic Features of Tricuspid Papillary Fibroelastoma

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Cardiac papillary fibroelastomas are rare and benign primary tumors in the cardiac valves or occasionally the mural endocardium. Before 1977, these tumors were diagnosed exclusively at postmortem examination. Over the last few years, a handful of cases have been diagnosed in vivo by echocardiography. In this report, we describe the first tricuspid valve papillary fibroelastoma detected by echocardiography in an adult. Clinical and echocardiographic features are discussed.

Cardiac papillary fibroelastomas are rare tumors found in the cardiac valves or occasionally, the mural endocardium (1–3). They are small, generally less than 1 cm in diameter and are biologically and histologically benign (2,3). Autopsy series (2,3) have demonstrated these tumors in both children and adults. In children, they are most commonly found on the tricuspid valve and are rarely, if ever, associated with symptoms (2–4). In adults, they are usually found on the aortic valve (1–3,5) or in the left ventricle (6) and have been associated with embolic phenomena, chest pain or sudden death (1–3,6). In this report, we describe the first tricuspid valve papillary fibroelastoma to be detected by echocardiography in an adult.

Case Report

The patient is a 64 year old woman with a history of coronary artery disease who underwent coronary bypass surgery 7 years ago. She was admitted to our hospital with the chief complaint of chest pain and recurrent near syncopal episodes. These episodes occurred unpredictably and were unassociated with her episodes of angina. Initial cardiac and neurologic examination failed to reveal a definitive diagnosis for her symptoms.

Physical examination revealed a thin, pale patient appearing older than her stated age. Cardiovascular examination revealed a fourth heart sound and a low frequency heart sound heard about 100 ms after the second heart sound (Fig. 1). The latter sound was most audible in the fourth right intercostal space at the sternal border. There were no murmurs. The electrocardiogram showed normal sinus rhythm with nonspecific ST-T wave changes.

M-mode echocardiogram revealed a mass of echoes appearing beneath the tricuspid valve leaflets in systole and behind the anterior leaflet of this valve in diastole (Fig. 1).

Real time two-dimensional echocardiogram (Hofrel System 201A/514) showed a discrete homogeneous rounded mass (1.5 x 1.5 cm) adherent to the atrial surface of the tricuspid valve (Fig. 2). The mass prolapsed deeply into the right ventricular cavity during diastole (Fig. 2A). In systole, it was located at the point of coaptation of the tricuspid valve leaflets and protruded into the right atrium (Fig. 2B).

Coronary arteriography demonstrated complete occlusion of all three existing grafts with significant stenosis in the native vessels distal to graft insertions.

The patient underwent coronary artery bypass surgery. The right atrium was explored and a 1.5 x 1.5 cm mass was easily peeled from the anterior leaflet of the tricuspid valve without damage to the valve or its supporting structures. Grossly, the mass was a papillary, gray, translucent structure. Microscopically, it had papillary fronds consisting of a central core of dense connective tissue surrounded by a layer of loose connective tissue and covered by a hyperplastic layer of endothelial cells (Fig. 3). These pathologic findings are typical of a papillary fibroelastoma.

The patient’s postoperative recovery was slow; however, no major complications were noted. She was discharged from the hospital with no further complaints of angina or episodes of presyncope.

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Figure 1. M-mode echocardiographic scan of the tricuspid valve (TV). A mass of echoes on the valve is seen in diastole in A and in systole and diastole in B. On a simultaneous phonocardiogram obtained from the apex (B), a protodiastolic low frequency sound is recorded 120 ms after S2 coincident with the maximal opening of the tricuspid valve. This is the characteristic phonocardiographic finding of a "tumor plop" (tp).

Discussion

In this patient, the discovery of an intracardiac mass was an incidental finding made during a routine echocardiographic examination (Fig. 1). The finding of an intracardiac mass was unanticipated, although retrospectively a "tumor plop" could be heard on cardiac auscultation and was recorded by phonocardiography (Fig. 1).

Echocardiographic findings. The findings on M-mode echocardiography were characteristic of a "mass," but not specific enough to make a definitive diagnosis. The mass of echoes was seen behind the echoes produced by the anterior leaflet of the tricuspid valve in diastole and resembled the atrial myxoma described by Wolfe et al. (7). However, unlike the latter case (7) and most subsequent reports (8,9), this mass was also seen behind the coaptation point of the tricuspid valve in systole. This suggested attachment to the tricuspid valve and encouraged consideration of valvular vegetation (10,11) or thrombus (12,13) in the differential diagnosis.

The two-dimensional echocardiographic examination confirmed and expanded the M-mode findings. On two-dimensional examination, the mass was shown to be homogeneous, rounded and well demarcated (Fig. 2). It moved with the anterior leaflet of the tricuspid valve and appeared to be adherent to its atrial surface. The echogenic properties were clearly consistent with tumor; however, its location on the valve was quite unusual. We speculated that this might represent a myxoma (8,14–16) with an unusual attachment to the tricuspid valve. The pathologic diagnosis

Figure 2. Apical four chamber echocardiographic views during diastole (A) and systole (B). A large, well demarcated, homogeneous mass (m) is seen attached to and moving freely with the anterior leaflet of the tricuspid valve. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.
Figure 3. Histologic sections of the resected tumor. A. Elastic stain at 10 × magnification demonstrating the three salient histologic features of the cardiac papillary fibroelastoma: 1) a dense central core of collagen and elastic fibers, 2) a peripheral myxomatous layer with deposits of acid mucopolysaccharides, and 3) overlying hyperplastic endothelial cells. B. Hematoxylin-eosin stain at 4 × magnification demonstrating the characteristic papillary fronds of a cardiac papillary fibroelastoma.

of papillary fibroelastoma provided another, although equally unusual, explanation for this finding.

Incidence. Papillary fibroelastomas are considered rare, but in fact, comprise 7 to 8% of all cardiac tumors (3). In contrast to myxomas, these tumors are smaller (2,3) and most commonly arise from the cardiac valves rather than the mural endocardium (1-3). Left and right-sided papillary fibroelastomas occur with equal frequency (1-6).

Clinical significance. The clinical significance of papillary fibroelastoma is quite uncertain. To our knowledge, there have been no reports of patients with clinical symptoms clearly shown to stem from this tumor. Previous authors (5) have speculated that this may be due to the tumor's small size. There have been reports of chest pain (3), embolic phenomena or sudden death (1-3,6) in patients with this tumor, but a cause/effect relation has not been established. In our case, the intermittent near syncopal episodes could not be correlated with any electrical or mechanical phenomenon. The possibility of intermittent right ventricular outflow obstruction or small pulmonary emboli originating from the tumor was not ruled out.

In conclusion, this case demonstrates the ability of two-dimensional echocardiography to detect the presence, location and character of intracardiac masses. Information obtained from the two-dimensional echocardiogram led to an appropriate differential diagnosis. In addition, echocardiographic information was adequate for planning the surgical approach without catheterization. The clinical significance of the tumor is uncertain; however, it is clear that cardiac papillary fibroelastoma must be included in the differential diagnosis of an intracardiac mass particularly when the mass is associated with cardiac valves.

References