

Aortic Valve Papillary Fibroelastoma

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Papillary fibroelastomas are benign and rare tumors usually involving the cardiac valves. The diagnosis of this condition has gradually increased over the past few years. This is attributed to the increasing number of echocardiographic and transesophageal procedures that are currently performed. The case is presented of a woman diagnosed with an aortic valve

Papillary fibroelastomas (PFEs) are rare and benign heart tumors which, prior to 1981, were usually diagnosed at autopsy (1). Recently, there has been a surge in the number of echocardiographic and transesophageal procedures being performed worldwide, and this has led to increased recognition of asymptomatic intracardiac masses and also to potential sources of coronary and cerebral embolization.

Here, the case is reported of a woman who was completely asymptomatic. A routine physical examination prompted transthoracic echocardiography (TTE) to be conducted, followed by transesophageal echocardiography (TEE) which revealed a mobile mass on the aortic valve.

Case report

A 55-year-old woman was found to have a mass attached to the aortic valve on routine echocardiography performed after a physical examination. TEE revealed a 1.2 × 1.0 cm circular mass (Fig. 1) attached to the left coronary cusp. The patient denied chest pain, shortness of breath or dizziness. She had no previous history of stroke or myocardial infarction, but was known to have hyperlipidemia and a history of hypertension. On physical examination the lungs were clear. Auscultation of the heart revealed normal heart sounds. There were no gallops or rubs, nor any systolic

mass, the sessile nature of which in the aortic root was considered a potential source of cerebral and coronary embolism. The mass was surgically removed, with no major complications being encountered perioperatively.

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or diastolic murmurs. The neurological examination was within normal limits, with no focal deficits.

Transthoracic echocardiography (TTE) revealed a mobile mass attached to the left coronary cusp and present on the aortic surface close to the left coronary ostium. Subsequent TEE corroborated the findings of TTE. The mass measured approximately 1.0 × 1.5 cm in diameter, appeared to originate from the aortic surface of the left aortic cusp, and was sessile in nature.

At surgery, the mass was found to be attached to the aortic valve (Fig. 2). The attachment to the valve precluded resection of the mass only. The pathology and anatomy were consistent with PFE (Fig. 3). The patient received a mechanical prosthetic valve (20 mm Medtronic-Hall), and her postoperative course was uneventful.

Discussion

PFE is the third most common tumor of the heart after myxomas and lipomas, and accounts for 7-8% of all cases (2,3). The lesions are small, ranging from 0.2 to 5 cm in size, and are commonly found on the cardiac valves. PFEs tend to have a predilection for the left side of the heart. The mean age of patients at diagnosis is 60 years, but a few cases have been documented in infants as young as six months (4).

The tumors can arise from any endocardial surface, but between 83 and 90% arise from the cardiac valves (5). When the tumors are on the aortic valve, they tend to occur on the ventricular side (2,5), but when they involve the semi-lunar valves they may occur on either side (2). Left-sided PFEs are usually smaller as they are diagnosed relatively earlier than their right-sided

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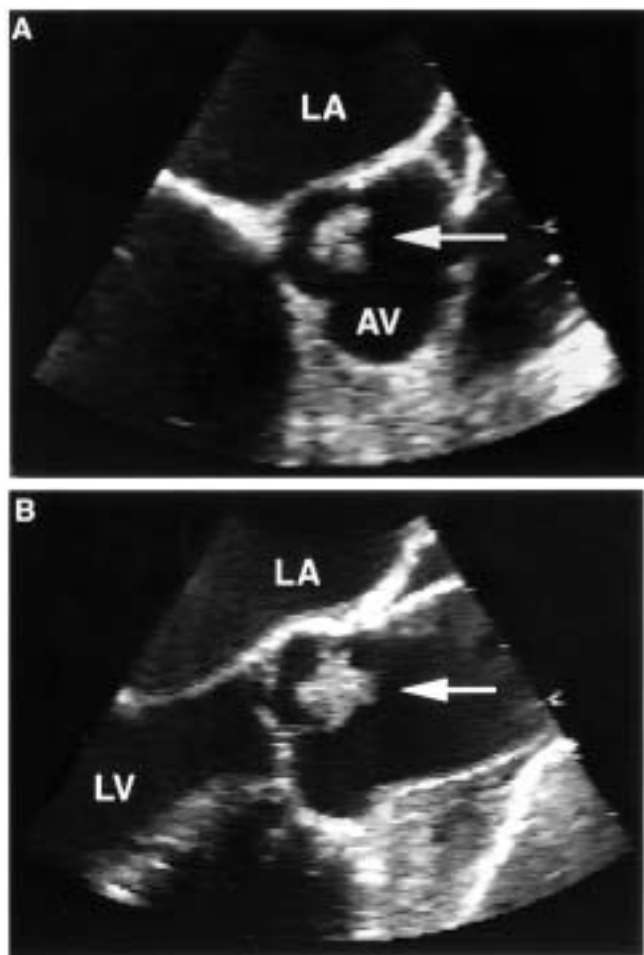


Figure 1: Transesophageal images of circular mass attached to aortic valve. A) Short-axis view (~20-30°); B) 120° view showing the aortic valve. The arrow points to mass. AV: Aortic valve; LA: Left atrium; LV: Left ventricle.

counterparts. Although these tumors can cause strokes and angina, those on the right side are well tolerated until they become large enough to block the passage of blood.

Transient ischemic attack/stroke is the most common manifestation of PFE. Other complications include angina, retinal artery emboli, conduction disturbances and valvular dysfunction (2,5-8). Paraplegia, although very rare, has also been described (9). Right-sided PFEs can rarely cause pulmonary embolism or outflow tract obstruction (9). Microscopically, the PFEs consist of a stalk with multiple papillary projections; they are avascular structures with a core of dense collagen and an outer layer of acid mucopolysaccharide (5).

Echocardiography is the preferred method for the diagnosis of PFEs. However, lesions smaller than 0.2 cm are less likely to be diagnosed by echocardiography

because of the current resolution capacity of the transducer (5). Echocardiographic features include a stalk rising out of the endocardium, high mobility, and refractile appearance. On TEE, 'stippled edge and shimmer' of the peripheral edge of the tumor is typically seen (10), and the appearance of the tumor has sometimes been compared to that of a sea anemone (1,5,8). When TEE is either contraindicated or technically not possible, multislice computed tomography scanning may be used which, because of its high spatial resolution, may provide an alternative means of diagnosis (11).

The management of these tumors is controversial. Aggressive surgical treatment is recommended for symptomatic PFE (6,12,13), whereas in asymptomatic PFEs resection is sometimes recommended, especially if the lesions are present on the left side (4,6-8,14). Others recommend anticoagulation or observation for the left-sided lesions (2,3). Asymptomatic right-sided lesions are usually not resected. Valve-sparing surgery can usually be accomplished in most patients. Most PFEs diagnosed prospectively present on normal-looking valves (10,12). Echocardiographic follow up has been recommended because of the risk of recurrence of these lesions, though most patients on follow up have not demonstrated evidence of recurrent tumor after surgical resection (4-6,10,14).

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Figure 2: Surgical excision of the mass attached to the aortic valve.

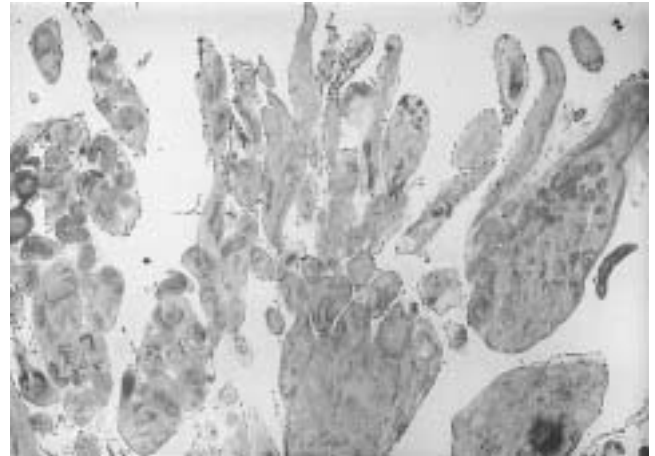


Figure 3: Microscopic findings of fibroelastoma excised from the aortic valve.

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