

Valve-Sparing Removal of Aortic Fibroelastoma through Ministernotomy

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Cardiac papillary fibroelastoma is a rare benign tumor which involves the heart valves and may cause thromboembolism or mechanical interference with valvular function. The case is presented of an asymptomatic, 22-year-old man in whom a fibroelastoma was localized on the ventricular aspect of the right coronary leaflet of the aortic valve. The tumor was identified during transthoracic echocardiography performed to monitor a mitral valve prolapse that had occurred at the age of 16 years. The patient

underwent surgical intervention by means of a minimally invasive thoracotomy. The postoperative course was uneventful, and the patient was discharged on postoperative day 5. Due to the high incidence of embolism, the tumor must be surgically removed immediately a diagnosis is confirmed. A minimal surgical approach appears to be safe and allows good exposure of the lesion.

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Cardiac papillary fibroelastoma is a rare benign tumor that involves the heart valves and may cause thromboembolism (1) or mechanical interference with valvular function. The incidence of this lesion has been estimated at approximately 10% of all primary cardiac tumors (2), and it is the second most frequent after myxoma. Edwards et al. (3) reported that is in fact the most common cardiac valve tumor, but because it is frequently asymptomatic the diagnosis may be missed. The tumor is usually attached to the valve leaflets, most often to the aortic valve, but less frequently to the tricuspid, mitral, and pulmonary valves (4).

Case report

An asymptomatic 22-year-old man was diagnosed with a fibroelastoma localized on the ventricular aspect of the right coronary leaflet of the aortic valve. The patient had undergone echocardiography (Fig. 1) to monitor a mitral valve prolapse that had occurred at the age of 16 years. The diagnosis was subsequently confirmed by transesophageal echocardiography (TEE) (Fig. 2) and the patient was referred for surgical removal of the tumor.

Surgery was performed via minimal access using a 'j' incision (Fig. 3). The 9 cm-long skin incision was made on the midline, from 3 cm below the sternal notch to the second intercostal space level. Subcutaneous tissue was undermined up to the sternal notch and down into the right third intercostal space. Sternal division was achieved using a standard saw, starting from the sternal notch and ending directly in the third right intercostal space with a 'j'-shaped cut. After dividing the sternum, a small Finochietto retractor was placed and the upper sternal edges spread: any thymus remnants were divided with electrocautery and the pericardium incised; the application of multiple heavy silk sutures allowed a better exposure of the aorta and right atrium. The patient was connected to the extracorporeal circulation (ECC) by placing an arterial cannula in the distal ascending aorta and a venous cannula in the right atrial appendage. The aorta was encircled and then cross-clamped, after which crystalloid cardioplegia solution was infused into the ascending aorta. The left ventricle was drained via a vent positioned in the right superior pulmonary vein. An 'italic S'-shaped incision was made above the aortic annulus, the aortic edges suspended, and the valve visualized. A 6×9 mm round and gelatinous formation was found adherent to the ventricular aspect of the right coronary cusp. The neoplasm was carefully removed using a small dissector. The gelatinous consistency of the lesion, and its firm adherence to the cusp, made removal difficult, such that multiple 'bites'

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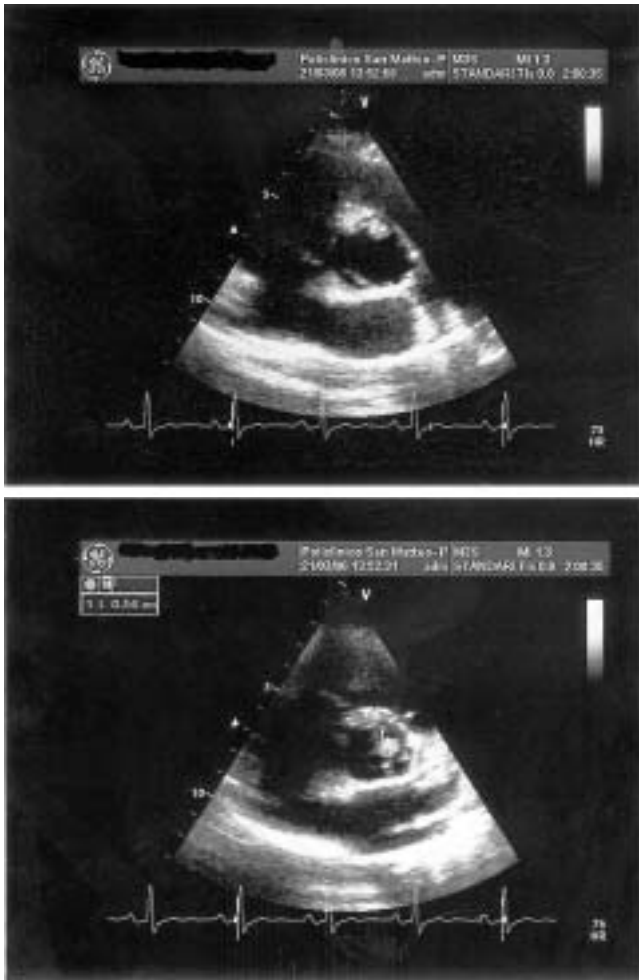


Figure 1: Transthoracic echocardiography showing (in parasternal view) the tumor on the right coronary cusp in diastole (upper) and systole (lower).



Figure 2: Transesophageal echocardiography showing the location of the fibroelastoma (Fib). Ao: Aorta; LA: Left atrium; PA: Pulmonary trunk; RA: Right atrium.

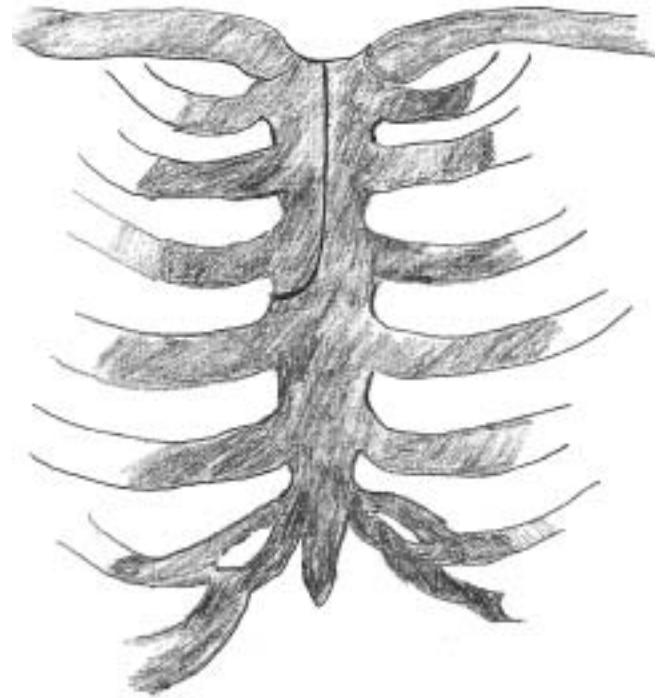


Figure 3: A schematic representation of the surgical approach, showing the 'j' incision.

were necessary for complete removal.

The aortic incision was closed using a 4-0 Prolene suture, and air removed from the cardiac chambers by gentle shaking of the heart, under TEE control. Pacing wires were positioned on the right ventricle. After declamping the aorta, the heart showed an immediate sinus rhythm. The aortic ischemic time was 50 min.

The patient was rapidly weaned from the ECC, the cannulas were removed and the pericardium was closed, leaving a chest tube in the pericardial cavity. TEE showed normal competence of the aortic valve.

Histology of the excised tissue showed multiple branching papillary fronds consisting of fibroelastic tissue surrounded by a layer of connective tissue with mucopolysaccharides and endothelial cells (Fig. 4).

The patient's postoperative course was uneventful, and he was discharged on day 5 after surgery. At a six-month follow up examination, echocardiography demonstrated a normal aortic valve with neither regurgitation nor tumor recurrence.

Discussion

Although cardiac papillary fibroelastomas have been reported in all cardiac chambers, they show a high propensity to localize in left-heart structures, with the aortic valve more often involved than the mitral valve (5). Embolism due to left-heart tumors may cause neurological symptoms, stroke or transient ischemic

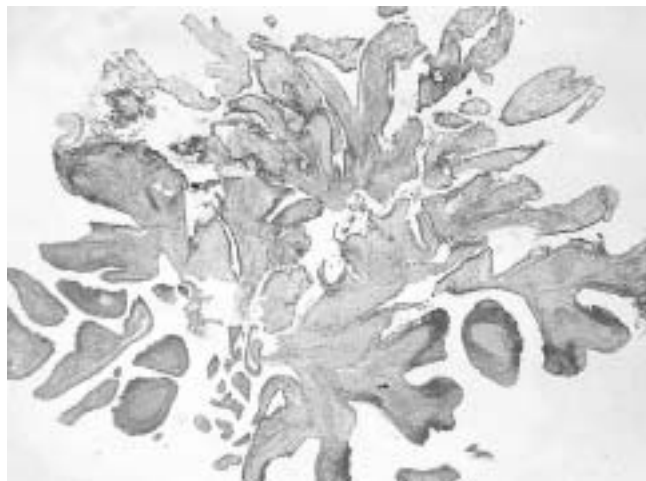


Figure 4: Histology of the excised aortic valve mass, showing the multiple fronds characteristic of fibroelastoma. (Movat pentachrome staining; original magnification, $\times 5$.)

attack, as well as retinal complications. Coronary embolism has also been reported, causing angina, myocardial infarction, or sudden death (6). Although in the present case the tumor was localized to the ventricular surface of the aortic cusp, the potential for fibroelastoma on the coronary cusps to prolapse into the coronary ostia should be always considered. Although a right-heart localization may be less dangerous, a patent foramen ovale may determine paradoxical embolism.

As this tumor is more frequently located in the high-flow and high-pressure areas of the heart, the risk of thromboembolism is greater than for the larger (and more common) benign heart neoplasm, atrial myxoma. Among a series of 88 patients with papillary fibroelastoma, Ngaage et al. (7) found that 53% had findings of embolism, and 43% had aortic valve lesions. In that study the aortic valve was spared in 89% of cases where valve tumors were treated with a simple shave incision; the valve was changed only in the presence of degenerative aortic valve disease. The first surgical resection of cardiac papillary fibroelastoma was performed in 1979 (8), since when all reports have indicated a median sternotomy as being the preferred surgical approach.

The minimal-access procedure for aortic valve surgery - the 'j'-approach - is made using a sternal saw, starting the bone incision in the midline from the sternal notch down to the right third intercostal space (9,10) (Fig. 3). Naturally, the skin incision must be short (but not too short) in order to avoid edge lesions.

Previously, there have been no reports of a minimally invasive approach, in particular a ministernotomy in the third intercostal space for this type of lesion affecting the aortic valve. Due to the high incidence of embolism, the tumor must be surgically removed as soon as a diagnosis is made. It is believed that the minimal surgical approach used in the present patient is safe and allows good exposure, especially if a left vent is placed in a pulmonary vein to drain the left ventricle and perform an accurate shave excision of the tumor. Moreover, this approach is well tolerated and results in a shorter hospital stay, less discomfort, and earlier return to work.

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