

Incidental Disclosure of Asymptomatic Coronary Embolic Occlusion Related to Mitral Valve Papillary Fibroelastoma: An Unusual Finding and a Review of the Literature

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Cardiac papillary fibroelastoma (CPF) located on mitral and aortic valves are known to produce systemic embolism mainly represented by strokes, whereas myocardial infarction and sudden death usually result from specific locations around LVOT. Coronary artery embolic occlusion originating from a mitral location has not yet been reported. The case is reported of a 42-year-old man referred for surgical treatment of a mitral valve papillary fibroelastoma disclosed after transitory and completely regressive

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Case report

A 42-year-old male was referred to the authors' institution following the disclosure of a small mobile mass located on the anterior mitral leaflet after a transitory and completely regressive neurological stroke. The

left hemicorporeal deficiency and previous myocardial infarct. Due to the left chamber location, surgery was scheduled and complete removal of the mass achieved. These findings emphasize the potential life-threatening complications of CPF and, independent of risk factors, the need to perform systematic coronary angiography before surgical excision is considered.

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patient had no specific cardiovascular risk factors, and his previous medical history was mainly characterized by alcoholism, without any symptoms of angina. Chest radiography was within normal limits, but the electrocardiogram showed unexpected signs of anterior apical myocardial infarction. The patient's serum troponin level was 4 µg/ml. Cardiac catheterization revealed a moderate apical akinesia related to occlusion of the left anterior descending artery in its distal portion, evoking an embolic mechanism (Fig. 1). No additional coronary lesion was disclosed.



Figure 1: Cardiac catheterization showing a distal embolic occlusion of the left anterior descending artery (arrow).

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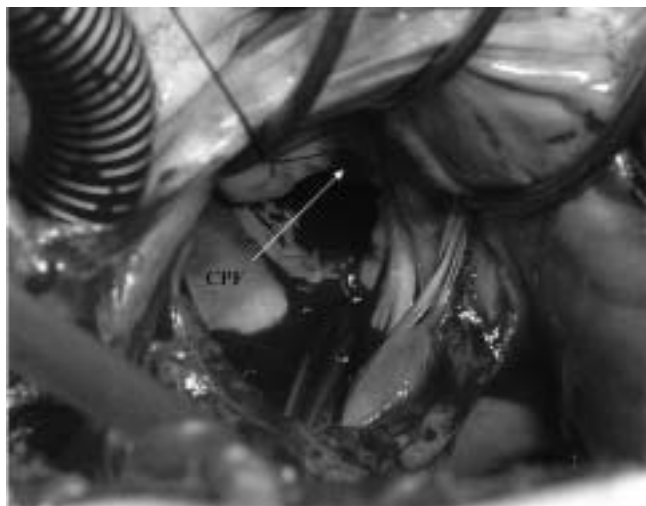


Figure 2: Operative view showing cardiac papillary fibroelastoma (CPF) located on the free edge of the anterior mitral leaflet (arrow).

Transthoracic echocardiography was suggestive of apical akinesia (left ventricular ejection fraction 45%) and disclosed an 8 × 9 mm mobile mass attached to the free edge of the anterior mitral leaflet with a trivial mitral insufficiency (grade 1/4). Transesophageal echocardiography provided interesting structural information by identifying an echolucent center that was strongly suggestive of a papillary fibroelastoma.

Because of a known high incidence of embolic events, the patient was referred to surgery for excision of the tumor. The surgical procedure was carried out through a median sternotomy.

After heparinization (300 U/kg), normothermic cardiopulmonary bypass was instituted by cannulation of the ascending aorta and bicaval venous drainage. The aorta was then clamped and cardioplegic arrest achieved with cold blood cardioplegia delivered in antegrade manner through the ascending aorta. The left atrium was opened in the interatrial groove and, following adequate placement of the self-retaining blades of a Cosgrove retractor, exposure of the mitral valve was completed. The mass was mobile and located on the free edge of the anterior mitral leaflet (Fig. 2).

A cleavage layer was easily found and the mass gently excised, preserving the whole integrity of the leaflet. Macroscopic examination showed an irregular surface partly covered with thrombotic deposits (Fig. 3), strongly corroborating an embolic mechanism.

The postoperative course was uneventful, and the patient was discharged in sinus rhythm after being prescribed low molecular-weight heparin and aspirin.

Histological examination confirmed the diagnosis of papillary fibroelastoma, with specific fronds being identified within three characteristic layers (Fig. 4).

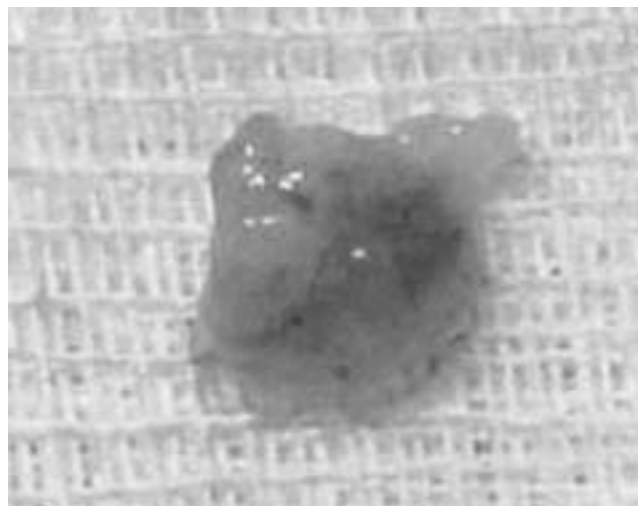


Figure 3: Macroscopic view of the papillary fibroelastoma after excision. The surface is irregular and covered with thrombotic deposits (black area).

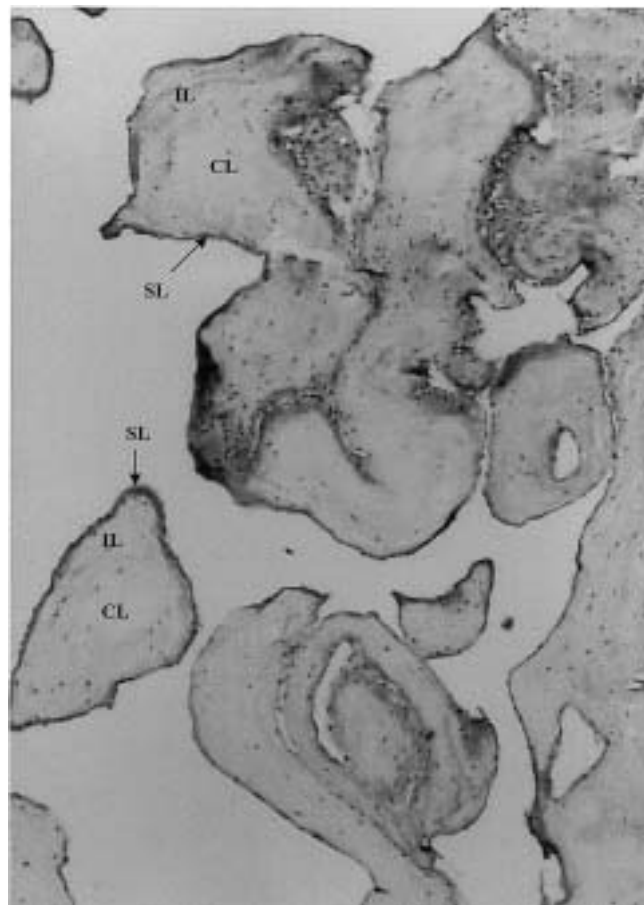


Figure 4: Histological examination showing typical fronds with three characteristic layers. Hematoxylin and eosin staining; original magnification ×20. CL: Central layer; IL: Intermediate layer; SL: Superficial layer.

Discussion

Cardiac papillary fibroelastoma is a rare and benign tumor which accounts for less than 10% of all primary cardiac tumors in adults (1-4). The first cases were described by Yater (5) in 1931, since which time - and despite an underestimated incidence - regular reports involving CPF have been made, with equal predilection between genders (4). CPFs remain exceptional in children, however (6). Papillary fibroelastoma is the third most common benign cardiac tumor after myxoma (30%) and lipoma (10%) (7). Although more than 90% of CPF are solitary, they may on rare occasions present as multiple lesions (8,9). While approximately 90% of the lesions arise from valvular tissue, most commonly from the aortic or mitral valves, other atypical locations have been reported, including the mitral or tricuspid chordae (10,11), papillary muscle (12), left atrial appendage (13), pulmonary vein (14), interatrial septum (15), endocardium (16), pulmonary and tricuspid valves (17,18), and vascular endothelium surrounding the coronary ostium (19). Although, anatomically, the right and left cardiac chambers seem to be involved with equal frequency (3,7,20), surgically reported experiences have noted a larger prevalence (81%) of left-sided CPF, most likely because they are much more frequently symptomatic. The diagnosis of CPF may represent a major challenge as the results of physical examinations are often poor and consequences of embolic events are essential in order to commence cardiovascular investigations.

Neurological accidents resulting from embolism arising from the left cavities are the most common symptoms, although unusual clinical presentations have also been reported as they depend on the location of the CPF. Consequently, a right chamber or tricuspid location might have mimicked multiple vegetations in suspected bacterial endocarditis (17,21), cyanosis or pulmonary embolism (22,23). Coronary symptoms such as angina, myocardial infarction, syncope, sudden death or atrioventricular block involve papillary fibroelastomas arising from the LVOT, aortic valve (24) or areas surrounding the coronary ostia. The pathogenic mechanisms of deleterious coronary consequences are then related to either coronary embolism or dynamic ostial obstruction resulting from a tumoral prolapse inside the coronary ostium (19,25). Gowda et al. (3) demonstrated a strong correlation between mitral tumors and transient ischemic attack or stroke. In addition to neurological accidents, aortic locations are currently associated with angina, sudden death and myocardial infarction. A direct correlation between coronary embolism and mitral-located CPF has not yet been reported, as an aortic location and tumor mobility are predictors of CPF-related death or non-fatal embolization (3). The present case emphasizes the high potential of non-specific embolic complications related to mitrally located CPF. Consequently, independently of the tumor

size and taking into consideration the spontaneously pejorative outcome (3), surgery remains strictly mandatory. Integrity of the superficial endothelial layer is essential in order to prevent embolic events, since embolism seems due to the migration of platelet and fibrin aggregates rather than to fragmentation of the tumor (26).

The histogenesis of papillary fibroelastomas continues to be a source of controversy. Several microscopic, immunohistochemical and molecular investigations (26,27) have provided multiple hypotheses, but none of these could definitely clarify whether CPF is an organized thrombus or occurs as the result of hamartomatous, neoplastic or reparative processes. The structural organization of the lesion is well known, but tumoral induction remains elusive and participation of the surface endothelial lining cells with excessive formation of basal membrane material is assessed in the formation of CPF. In a previous study (26), the present authors demonstrated the presence of dendritic cells and remnants of cytomegalovirus in the intermediate tumoral layer, together with a centrifugal mesenchymal cellular migration from the central layer towards the superficial layer, with differentiation steps. These findings suggested that papillary fibroelastoma might be virally induced, thereby evoking the concept of a chronic form of viral endocarditis as an underlying cause of CPF formation.

Repetitive hemodynamic trauma (rheumatic heart disease, hypertrophic cardiomyopathy, atrial septal defect, mitral valve prolapse) and iatrogenic factors (28,29) (thoracic irradiation, open-heart surgery) might be involved in the development of at least 18% of cardiac fibroelastomas. In contrast to sporadic cases, which occur most commonly on valvular tissues, iatrogenic papillary fibroelastomas tend to occur on a variety of non-valvular endocardial surfaces (7), usually close to the predisposing iatrogenic factor - for example, a cardiac chamber associated with the site of surgery.

In conclusion, although CPF is a histologically benign tumor, its strategic location may produce devastating consequences as a result of the high cumulative risk of embolization. Thus, a diagnosis of CPF mandates prompt surgical resection. If both aortic and mitral locations are known to be responsible for neurological accidents, then any deleterious coronary consequences are normally associated with LVOT and aortic valve locations. The present case highlights the ability of mitral fibroelastoma to cause unexpected coronary problems since, to the present authors' knowledge, there have been no reported cases of mitral papillary fibroelastoma that correlate directly with coronary embolic occlusion. Thus, in order to evaluate any unusual deleterious consequences of CPF, coronary angiography should be considered in patients with left-sided cardiac papillary fibroelastoma, independently of risk factors and age.

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