Incidental finding of a papillary fibroelastoma of the mitral valve chordae

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Cardiac papillary fibroelastoma (CPF) is a rare neoplasm with predilection for heart valves, usually found incidentally on routine echocardiography. Most CPFs are asymptomatic; rarely, they are diagnosed because of cardiac symptoms or after an embolic event. This report describes the case of a 69-year-old woman with the incidental finding of a mass attached to the anterior mitral valve chordae. Owing to the risk of embolic complications, surgery was emergently performed with complete resection of the mass and preservation of the mitral valve. Histological evaluation confirmed the diagnosis of CPF.

Case presentation

A 69-year-old asymptomatic woman was referred to our institution for evaluation of a cardiac mass found on a routine transthoracic echocardiogram (TTE). Eight years earlier, a previous TTE had shown mild aortic regurgitation with an otherwise normal examination. Physical examination, electrocardiogram, chest X-ray, and laboratory data, including full work up for infective endocarditis, were unremarkable except for a mild anaemia. Transoesophageal echocardiography (TEE) revealed a 16 × 20 mm highly mobile mass attached to the anterior mitral valve leaflet chordae at its junction with the anterolateral papillary muscle (Figure 1, see Supplementary Videos S1 and S2 online). Valvular function was preserved without significant mitral regurgitation or stenosis. A patent foramen ovale with a left-to-right shunt was also noted. Considering the significant risk of embolic complications, it was decided to admit the patient to proceed with early surgery. A preoperative coronary angiogram was performed and demonstrated severe stenosis of the right coronary artery.

The patient underwent surgical resection of the mass under cardiopulmonary bypass. The heart was exposed through a median sternotomy. The mitral valve was first exposed via a superior transeptal approach. However, the tumour could not be found with this approach. Then a supra-coronary transverse aortotomy was performed and the mass was visualized adherent to the second-order chordae of the anterior mitral valve leaflet at the junction with the anterior aspect of the anterolateral papillary muscle (see Supplementary material online, Figure S1). On the basis of the macroscopic aspect of the mass suggestive of a benign tumour and its location, complete tumour resection with mitral valve preservation was performed. The tumour was entwined around a single secondary chordae, without involving the mitral leaflets. The patent foramen ovale was closed and a single saphenous venous bypass graft was performed to the distal right coronary artery. Thorough inspection of the mitral valve and left ventricle did not reveal any residual tumour. The patient was weaned from bypass without difficulty. The postoperative TEE confirmed the absence of residual tumour and normal mitral valvular function. Histopathologic examination of the mass confirmed the diagnosis of a cardiac papillary fibroelastoma (CPF). The postoperative course was uneventful and the patient was discharged on day 7 after the surgery.

Discussion

Primary tumours of the heart are rare, with a reported frequency of 0.02% according to large autopsy series.1 Cardiac papillary fibroelastoma are the third most common primary cardiac tumours after myxomas and lipomas. They consist of avascular papillomas covered by a single layer of endothelium. The classic pathologic appearance is of a pedunculated flower-like mass attached to the endocardium. Immersion in water after resection results in a typical sea...
anemone appearance. Cardiac papillary fibroelastomas predominantly affect heart valves, accounting for 75% of all valvular tumours.\(^1,2\) The aortic valve is the most frequently involved followed by the mitral valve. The atrial side is more commonly involved for ativoventricular valves, whereas both sides of the semilunar valves are equally affected. Less frequent sites of involvement include: mitral chordae, right atrial endocardium, endocardial surface of both ventricles including papillary muscles and interventricular septum. The left ventricle is the most common non-valvular site involved.\(^1,3\)

Tumour size varies significantly with reported dimensions between 2 and 70 mm (mean size 9 mm). Male are slightly more affected (55%) with the highest prevalence later in life (mean age at diagnosis 60 years).\(^1,3\) Most CPFs are asymptomatic. When symptomatic, clinical presentation varies depending on site of involvement. Right-sided CPFs are typically asymptomatic, but can rarely cause pulmonary embolism. Left-sided tumours most commonly present with neurologic symptoms from cerebral embolization, with transient ischaemic attack, visual disturbances or stroke.\(^1,4,5\) Other symptoms include angina, heart failure, syncope, or pre-syncope. Alternatively, CPFs can present with life-threatening complications. Acute myocardial infarction, caused by a tumour occluding the coronary ostium or by embolization, may be the presenting symptom. Sudden death in younger individuals has also been described.\(^1,4,5\)

Classically found incidentally at autopsy or surgery, CPFs are now increasingly recognized with the widespread use of echocardiography. Although there are no specific distinctive features, CPF must be differentiated from other intracardiac tumours, vegetations, and thrombi. Noted features include pedunculated finger-like mobile excrescences of various sizes attached to endocardial sites of intracardiac valves by a small stalk.\(^5,6\) With tumour size larger than 2 mm, the sensitivity and specificity of TTE are 88.9% and 87.8%, respectively, with an overall accuracy of 88.4%. With smaller tumours (<2 mm), the overall sensitivity of TTE is 61.9%, compared with 76.6% for TEE.\(^3,6\) Echocardiography also plays an important role in influencing management decisions regarding tumour excision, valve replacement, or repair.\(^3,6\)

The potential for life-threatening complications of CPFs, even asymptomatic ones, generally warrants surgical consideration. Surgical excision is indicated for symptomatic CPFs and large or highly mobile asymptomatic CPFs, as long as there are no major contraindications to surgery. The only independent predictor of death or embolization is tumour mobility.\(^1\) Location of the mass is the most important factor influencing surgical approach and technique.\(^7\) Surgical excision must be complete, and when valvular tissue is involved, the valve should be preserved if possible. Recurrence has not been reported as long as the tumour is completely resected.\(^8\)

This report highlights the rare chordal location of a CPF without valvular involvement. Echocardiography provides a valuable tool by its ability to delineate the exact location of the mass, to assess valvular involvement, and to confirm complete tumour resection. Prompt surgical resection with preservation of valvular integrity is recommended in most cases.

Supplementary data
Supplementary data are available at *European Journal of Echocardiography* online.

References