CASE REPORT

Presentation of papillary fibroelastoma of the aortic valve with atypical symptoms

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SUMMARY

Papillary fibroelastomas are rare benign tumours arising from the cardiac endothelium. The rationale behind the treatment of fibroelastomas is to eliminate the risk of embolic events and the associated morbidity and mortality. These tumours present predominantly in males during the seventh decade of life and most commonly affect the aortic valve. We report a case of aortic valve papillary fibroelastoma with an atypical clinical presentation, treated with valve-sparing surgical excision. Our case highlights the management of this rare but serious condition and emphasises the need to develop clear guidelines regarding the treatment of asymptomatic patients with a papillary fibroelastoma.

CASE PRESENTATION

A 69-year-old ex-smoker male patient was referred to the Cardiology clinic with a 2-year history of progressive shortness of breath on exertion, but reported neither chest pain, syncope nor orthopnoea. His medical history consisted of hypertension, hypercholesterolaemia, severe chronic obstructive pulmonary disease (COPD), marked thoracic scoliosis and deep vein thrombosis in 2008, for which he is treated with lifelong warfarin. There was no significant family history. On examination, he was in sinus rhythm with regular pulse at 68 bpm and high blood pressure at 168/89 mm Hg. Cardiovascular, respiratory and abdominal examinations were otherwise unremarkable, except for severe scoliosis of the lower thoracic vertebrae (T8–T12).

BACKGROUND

Papillary fibroelastomas are benign cardiac neoplasms which are nevertheless considered for surgical management as they carry a risk of life-threatening and life-altering complications (table 1). Symptomatic patients diagnosed with a fibroelastoma often present with syncope from embolic events or angina. We report a case where a patient presenting with atypical symptoms due to a papillary fibroelastoma attached to the aortic valve underwent surgical excision of the cardiac mass with a good clinical outcome. This is an area in which clear guidelines for asymptomatic patients are lacking and would be of benefit in assisting clinicians to balance the risks of future embolic events with the risks of a surgical intervention.

INVESTIGATIONS

Transthoracic echocardiogram scan (TTE) showed a pedunculated mobile mass measuring 11 mm × 9.5 mm attached to the left coronary cusp (LCC) of the aortic valve on the aortic side. TTE also revealed a preserved biventricular systolic function as well as no significant valvular disease. The aortic root was mildly dilated at 3.7 cm. This was considered as an incidental finding as there was no evidence of aortic stenosis or incompetence, suggesting that the cardiac mass was unlikely to be the cause of the dyspnoea. The patient was subsequently referred for lung function tests and a CT-coronary angiogram (CTCA). Lung function tests revealed a significantly reduced FEV1 and FVC, suggesting a restrictive pattern, likely due to his severe thoracic scoliosis. CTCA also showed multiple calcified plaques with 70% stenosis of the left anterior descending (LAD) artery. Coronal CT images (figure 1) showed a lobulated mass attached to LCC. An ECG was done to complete the cardiac investigations, showing a sinus rhythm.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for the cardiac mass found on TTE included an atrial myxoma, papillary fibroelastoma, infective endocarditis with vegetations on the aortic valve and thrombus formation. Although classification of the cardiac mass found can only be found via surgical excision and histology, the echocardiographic features of this mobile mass pedicled by a stalk to the valvular surface made a papillary fibroelastoma the most likely diagnosis, where other possible diagnoses were less probable in the absence of concomitant symptoms.

Table 1 Main life-threatening complications that patients with a papillary fibroelastoma are at a higher risk of suffering

Life-threatening complications in patients with undiagnosed papillary fibroelastoma

Complication	Cause	Attributable symptoms
Myocardial infarction	Embolisation of tumour fragments into the coronary vasculature	Angina Shortness of breath
Ischaemic stroke	Embolisation of tumour fragments into the cerebral vasculature	Syncope
Ventricular fibrillation	Embolisation of tumour fragments into the coronary vasculature, causing subsequent ischaemia of myocardium	Shortness of breath Syncope

The mechanism for each of the complication involves the embolisation of tumour fragments into the systemic vasculature.^{8,9}

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Figure 1 CT scan showing a lobulated mass attached to the left coronary cusp of the aortic valve (arrow). Measurements were 10.9 mm × 0.95 mm.

The symptomatic presentation of the patient was considered unlikely to be caused by the cardiac mass due to the lack of aortic stenosis or regurgitation. His dyspnoea was thought to be caused either by a combination of the severe thoracic scoliosis and COPD or an atypical presentation of the papillary fibroelastoma recently diagnosed.

TREATMENT

Following discussion at the cardiothoracic multidisciplinary team meeting, surgical excision of the Papillary Fibroelastoma (PFE) was considered the most appropriate treatment method. Risks quoted for this operation included the standard complications associated with a sternotomy and cardiopulmonary bypass; additional risk of aortic cusp injury; requirement for aortic valve replacement or embolisation of the fibroelastoma.

Prior to undergoing the excision of the fibroelastoma, coronary stenting of the proximal LAD artery was achieved to address the significant stenosis of the proximal LAD diagnosed during preoperative CTCA.

Following successful coronary intervention, the patient underwent surgical valve-sparing excision of fibroelastoma via median sternotomy. The mass was removed from the base of the pedicle without involving the cusp itself, as intraoperative findings revealed that the mass was not directly attached to the LCC, allowing complete removal of the lesion without requiring aortic valve repair. Standard cannulation of the ascending aorta (arterial line) with a two-stage venous cannulation (venous line) was established. After administration of standard antegrade cardioplegia via the aortic root, a transverse aortotomy was achieved, allowing the fibroelastoma to be excised from the LCC of the aortic valve.

Box 1 Common echocardiographic findings of fibroelastomas seen on transoesophageal echocardiograph (TOE)³

- ▶ Common echocardiographic characteristics of papillary fibroelastomas.
- ▶ Small size (<1.5 cm).
- ▶ Pedical or stalk attachment to endocardium.
- ▶ Areas of echolucency.
- ▶ Variable mobility.

Figure 2 Intraoperative TOE image showing the papillary fibroelastoma attached to the left coronary cusp of the aortic valve (arrow). TOE, transoesophageal echocardiogram.

Intraoperative transoesophageal echocardiogram (TOE) findings included good left ventricular ejection fraction (>50%) with no evidence aortic incompetence, with an aortic valve maximum pressure gradient (AV maxPG) of 4.39 mm Hg. The patient was weaned off Cardiopulmonary Bypass (CPB) in sinus rhythm.

OUTCOME AND FOLLOW-UP

Postoperative TTE was carried out 5 days following surgical excision of the cardiac mass and showed preserved biventricular function with no obvious residual mass detected. The function of the aortic valve was also preserved, with no evidence of aortic incompetence. Histopathology showed that the excised specimen was a papillary fibroelastoma. As this was a benign neoplasm, no further surgical treatment was indicated, with regular follow-up in clinic. His postoperative course was unremarkable and he was discharged on day 6 postoperatively in stable health on his regular medications, along with a trial of Salbutamol (2 puffs two times per day) and Beclomethasone (2 puffs two times per day) inhalers to improve his breathlessness, likely exacerbated by his lung pathology.

At 2-month follow-up, the patient's shortness of breath had improved significantly, being able to walk for a mile at slow pace, and was otherwise asymptomatic.

Video 1 Intraoperative TOE showing a lobulated mass attached to the LCC of the aortic valve. LCC, left coronary cusp; TOE, transoesophageal echocardiogram.

Figure 3 Algorithm for the management plan of papillary fibroelastomas, depending on the presence of symptoms and location, as proposed by Yee *et al.*⁷

DISCUSSION

Papillary fibroelastomas are rare avascular neoplasms first described in the literature by Cheitlin *et al* in 1975¹ but nonetheless represent the second most common benign neoplasm affecting the cardiac valves, behind atrial myxomas, constituting 10% of all cardiac tumours.² Although they may occur anywhere in the cardiac endothelium, papillary fibroelastomas often develop on the valvular endocardium (84%), with the aortic valve being most commonly affected (44%), mitral valve (35%), tricuspid valve (15%) and pulmonary valve (8%) being involved in that order.¹ Non-valvular fibroelastomas otherwise occur within the heart chamber endocardium (16%).

These tumours are composed of fronds of collagen and elastin covered by hyperplastic endothelial cells.¹ Macroscopically, such tumours are often described as a 'sea anemone'-like structure,² owing to the multiple frond-like projections which are prone to embolisation.³ The pathological origin of fibroelastomas remains unknown with several theoretical aetiologies including viral infections or repeated trauma to the cardiac valves or congenital cardiac anomalies.⁴ Although the coexistence of congenital heart disease and PFE remains extremely rare, Özkan *et al* observed that certain congenital cardiac anomalies, specifically atrial septal defect and patent ductus arteriosus, may be linked with the development of papillary fibroelastoma.⁵

Clinical presentation of patients with PFE varies greatly and does not necessarily correlate with the size of fibroelastoma.⁴ Gowda *et al* produced a comprehensive analysis of 725 cases of papillary fibroelastomas and found that the majority of patients with PFE were asymptomatic at the time of diagnosis (63%). However, symptomatic patients will typically present acutely with angina (6%–10%) or syncope due to a stroke of transient ischaemic attack (16%).³ Atypical manifestations include

dyspnoea secondary to heart failure (3%) and blindness due to distal embolisation into the retinal vasculature (1%).¹ Gowda *et al* also observed that the mean age of presentation with these neoplasms is in the seventh decade of age, with a slight predominance in males (56%).

Symptomatic papillary fibroelastomas should be differentiated from coronary artery disease and cerebrovascular disease.⁶ Asymptomatic patients will often be found incidentally on TTE findings for other indications and will require surgical excision with histological studies to differentiate a PFE from alternative causes of abnormal growth on valvular tissue, mainly being atrial myxomas,⁶ infective endocarditis with vegetations and thrombus formation.

Although cases of papillary fibroelastoma often present incidentally on TTE findings requested for other indications, the gold standard diagnostic test remains a TOE.³ Common echocardiographic characteristics of papillary fibroelastomas have been summarised in box 1. Although there are no specific diagnostic criteria for PFE, the echocardiographic findings in our patient (figure 2, video 1) are in agreement with those from previously published data.³

While there are no evidence-based guidelines governing the management of PFE, conclusions drawn from published case series clearly support surgical excision in symptomatic patients, mainly to prevent further embolic events rather than treat the original symptoms *per se*.^{3,4} The choice between surgical excision and therapeutic anticoagulation is debated if a patient is asymptomatic. Yee *et al* proposed an algorithm for the assessment and management of PFE (figure 3)⁷ based on the echocardiographic size, location and mobility of the tumour. This study describes that the treatment of choice for patients with left-sided PFE is surgical excision of the cardiac mass. Regular follow-up

with monitoring of tumour size until definitive surgical treatment can be done in patients who have a low-risk of complications with embolisation of the tumour. Considering our patient's medical history and tumour size, surgical excision was the most appropriate treatment option.

In the case series published by Gowda *et al*, the most common surgical treatment strategy was valve-sparing surgical excision (81%), with the rest of the surgical interventions being excision with valve replacement (10%) or excision with valve repair (9%) which may be required due to adhesion formation of the tumour to the heart valve leaflets.¹ Valve-sparing excision offers a good clinical outcome and is the preferred surgical method.¹ However, valve repair or replacement is indicated if there is underlying destruction of the valvular structures.²

Learning points

- Presentation of papillary fibroelastomas is variable and many are identified incidentally in patients who are asymptomatic.
- Untreated fibroelastomas represent an increased risk of life-threatening embolisation, such as myocardial infarction or neurological events.
- No evidence-based guidelines exist to aid decision making in this area. Surgical excision is often recommended in symptomatic patients but there is a lack of consensus regarding the optimal management of patients who are asymptomatic.

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