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CASE REPORT
Cardiac imaging

An atypical cause of myocardial infarction: case report of an obstructing papillary fibroelastoma of the aortic valve

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Background Papillary fibroelastomas are rare primary cardiac tumours with a prevalence of 0.01% at autopsy. They are histologically benign tumours but have been demonstrated through case series to confer an increased risk of thrombo-embolism resulting in: transient ischaemic attack, stroke, myocardial infarction, and pulmonary and systemic embolization.

Case summary A 54-year-old woman presented with central chest pain radiating to her left arm. At presentation there was a significant troponin rise; initial high-sensitivity troponin-I (hsTn-I) 660 pg/mL increased to 3340 pg/mL at 6 h. Coronary angiogram did not reveal any obstructing coronary artery disease. Echocardiography revealed a rounded, mobile mass on the left coronary cusp of the aortic valve suspicious for papillary fibroelastoma. The patient underwent shave excision of the lesion. Intra-operatively it was noted that the mass intermittently sat within the ostium of the left main resulting in its occlusion. Histology confirmed a papillary fibroelastoma.

Discussion Primary cardiac tumours are rare but can cause life-threatening complications such as stroke, myocardial infarction, and cardiac arrest. In the literature, the mechanism of these complications is mainly attributed to thrombo-embolism. This case demonstrates the utility of echocardiogram in investigating and diagnosing a rare cause of myocardial infarction and highlights an unusual mechanism, that is tumour causing obstruction of the coronary ostium.

Keywords Papillary fibroelastoma • MINOCA • Case report • Myocardial infarction with non-obstructive coronary arteries

Learning points

- Papillary fibroelastoma are benign tumours but can cause a number of complications, for example stroke, transient ischaemic attack, myocardial infarction, and cardiac arrest.
- Papillary fibroelastoma can cause complications due to obstruction of the coronary ostia.
- Papillary fibroelastoma can be readily identified with transthoracic echocardiography.
- There is no consensus on the management of these tumours, however, with advanced surgical techniques the majority of cases can be managed with valve-sparing surgery.

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Introduction

Papillary fibroelastomas are benign primary cardiac tumours. They are typically diagnosed as an incidental finding but can also result in thrombo-embolism.[1] These tumours are rare and have a varied clinical course and as a result there is no clear consensus on management. This report describes a case of papillary fibroelastoma presenting as a non-ST elevation myocardial infarction due to obstruction of the coronary ostium.

Timeline

Admission	Presented with typical cardiac chest pain
	High-sensitivity troponin-I rise from 660 to 3340pg/mL
Day 1	Coronary angiogram did not show evidence of obstructive coronary artery disease
Day 2	Further investigation into cause for chest pain and elevated troponin
	Transthoracic echocardiogram found an aortic valve mass suspicious for papillary fibroelastoma
Two weeks post-presentation	Re-admitted for shave excision of aortic valve lesion
Four weeks post-presentation	Returned for outpatient review
	Asymptomatic
	No complications post-operatively
	Histology consistent with papillary fibroelastoma

Case presentation

A 54-year-old female presented to the Emergency Department with sudden, severe central chest pain while cleaning. The pain then persisted at rest. The pain radiated to her left arm with associated diaphoresis. She described three similar episodes occurring in the past year which were not as severe. Her background history included dyslipidaemia, hypothyroidism, and hypertension. Her physical examination was normal.

The patient's initial electrocardiogram was recorded while the patient was pain free and showed minor ST depression in V5 and V6 (*Figure 1*). An initial hsTn-I of 660 pg/mL (reference range <15.6 pg/mL) increased to 3340 pg/mL in a 6-h period. A coronary angiogram revealed non-obstructing coronary artery disease.

A transthoracic echocardiogram (TTE) and subsequent transoesophageal echocardiogram performed while the patient was pain free revealed normal left ventricular systolic function with no regional wall motion abnormalities. However, there was a rounded, echodense 8 × 8 mm mobile mass attached to the left coronary cusp of aortic valve. There was no aortic regurgitation (*Figure 2*). A computed tomography of the thorax, abdomen, and pelvis revealed no other masses or evidence of emboli.

The patient underwent a shave excision of the lesion with preservation of the valve. Intra-operatively it was noted that when the valve leaflet was opened the lesion was obstructing the ostia of the left main (*Figure 3*).

Histology revealed branching papillary fronds with central avascular collagen and variable elastic tissue consistent with papillary fibroelastoma (*Figure 4*).

The patient recovered well post-operatively without any complications and will be followed yearly with TTE to monitor for recurrence.

Discussion

Primary cardiac tumours are rare, with a prevalence at autopsy of 0.01%. Papillary fibroelastoma is the most common valvular tumour of the heart, accounting for 15% of all primary cardiac tumours. It most commonly affects the aortic valve, but is also found in the left ventricular outflow tract, tricuspid valve, and mitral valve. Single or multiple lesions can develop.[2]

Papillary fibroelastomas are benign cardiac tumours. They are avascular and comprised of fibroelastic tissue surrounded by endocardium.[3] Their incidence increases with age and there appears to be an association with previous endocardial damage, for example radiation, rheumatic heart disease, aortic sclerosis, and previous surgery.[3,4] The origin of these lesions is not known. They are believed to be acquired rather than congenital. One widely accepted theory on their origin is the microthrombus theory that is small thrombi combine at a site of endothelial damage and form a tumour.[5]

Papillary fibroelastomas are frequently an incidental finding; 67% of patients who underwent resection of a papillary fibroelastomas in one case series were asymptomatic and the lesion was found incidentally.[1] They can also present with systemic embolization such as: transient ischaemic attack, stroke, myocardial infarction, pulmonary embolism, and limb, mesenteric, and renal ischaemia.[1,6] A review of the literature found four case reports where the authors proposed that a papillary fibroelastoma occluding the coronary ostia was the cause for the patient's presentation. Three of these cases presented with angina and the 4th case presented in cardiac arrest.[1,6–8]

In our case, a diagnosis of acute myocardial infarction was made based on the presence of ischaemic symptoms and a rise in hsTn-I.[9] Cardiac magnetic resonance imaging would have been useful to diagnose a subendocardial myocardial infarction; however, the patient was unable to tolerate this despite sedation due to claustrophobia. The patient presented with a non-ST elevation myocardial infarction in the setting of normal coronary arteries and a fibroelastoma on the aortic valve. We felt the mechanism of myocardial infarction in this case was transient occlusion of the left coronary ostium because the coronary angiogram did not show any evidence of coronary occlusion or coronary artery stenosis and it was noted intra-operatively that the tumour obstructed the left coronary ostium. Given the established thrombo-embolic risk with papillary fibroelastomas another possible aetiology that could be considered in this case is embolization of tumour associated thrombi with subsequent

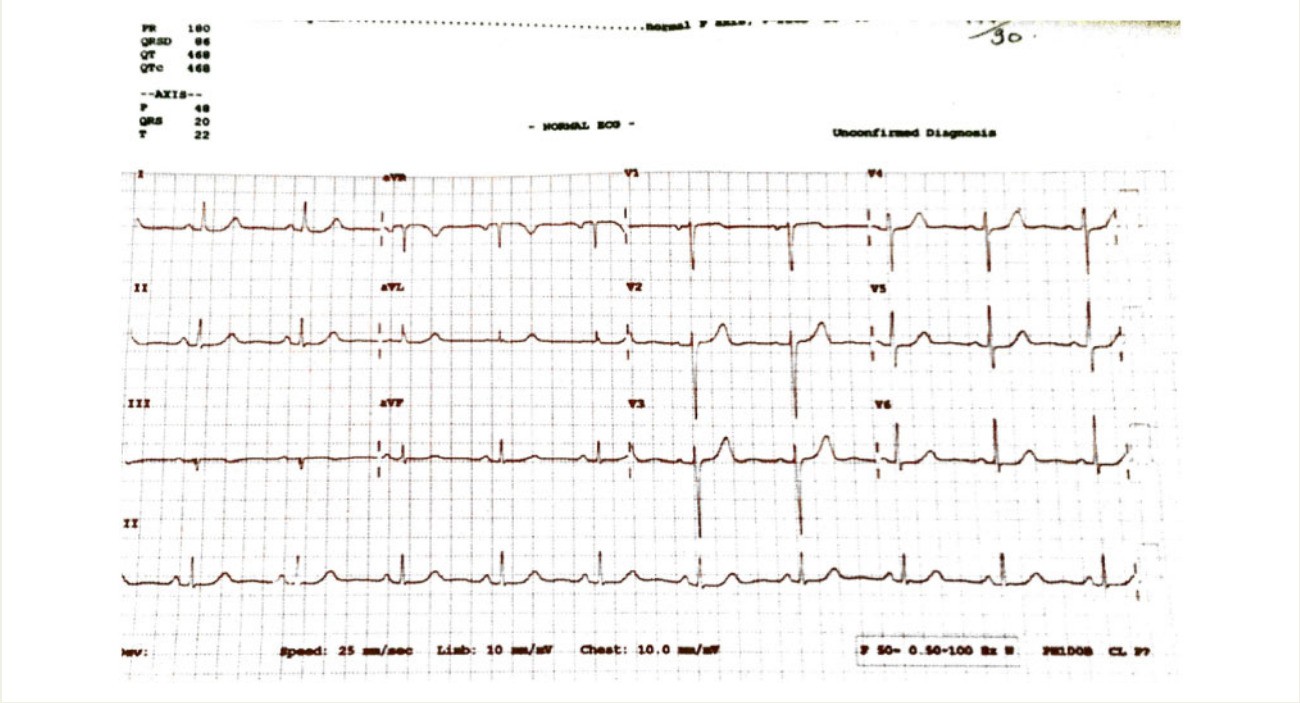

Figure 1 Electrocardiogram at admission showing minor ST depression in V5 and V6.

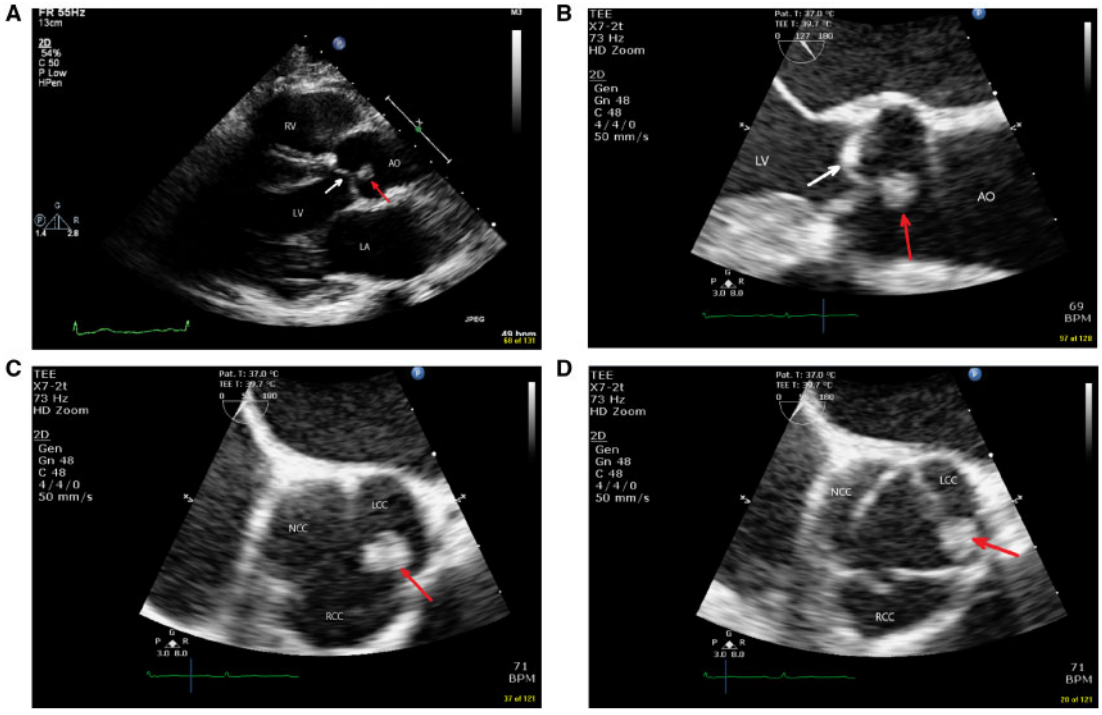

Figure 2 (*A*) Transthoracic echocardiogram parasternal long-axis view; (*B*) transoesophageal echocardiogram mid-oesophageal long-axis view; (*C*) transoesophageal echocardiogram mid-oesophageal short-axis view in diastole; and (*D*) transoesophageal echocardiogram mid-oesophageal short-axis view in systole. Red arrow: papillary fibroelastoma; white arrow: aortic valve. AO, aorta; LA, left atrium; LCC, left coronary cusp of the aortic valve; LV, left ventricle; NCC, non-coronary cusp of the aortic valve; RCC, right coronary cusp of the aortic valve; RV, right ventricle.

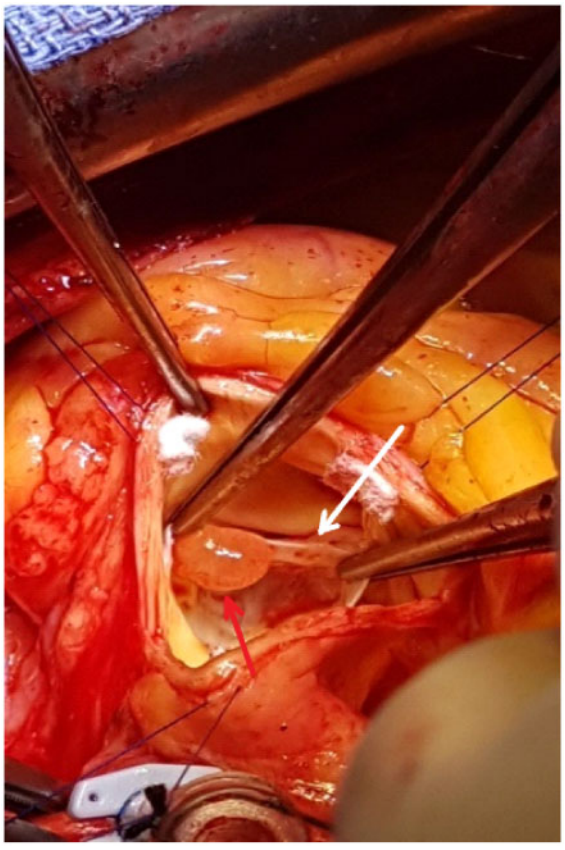

Figure 3 Intra-operative view, a jelly like mass attached to the left coronary cusp of the aortic valve.

reperfusion or embolization of small tumour fragments that could not be detected on coronary angiography.

Echocardiography is a useful tool in differentiating types of cardiac tumour. Transoesophageal echocardiogram has a 76% accuracy in the diagnosis of papillary fibroelastoma. They appear as a pedunculated, mobile, homogeneous, small mass (typically <20 mm) with a characteristic stippling along their edges.[10] Transoesophageal echocardiogram can precisely locate the point of attachment to facilitate effective surgical planning. In the majority of cases, the diagnosis can be made based on typical echocardiographic features and so surgical excision is not always necessary to diagnose the lesion.

The role of anticoagulants and antiplatelets in the management of papillary fibroelastoma remains unclear. The pathophysiology of embolic events in patients with papillary fibroelastoma may be due to embolism of thrombi or of the tumour itself. Thrombi have been reported on the surface of papillary fibroelastoma.[3] One case report describes a patient with a right-sided hemiplegia who underwent thrombectomy and the thrombus histology was consistent with papillary fibroelastoma.[11]

The decision on management of papillary fibroelastoma is difficult as there have been no randomized controlled trials. In the literature the consensus is that if the patient is a good surgical candidate they should undergo surgical excision.[3] Tamin *et al.*[3] found that the risk of stroke in patients with a known papillary fibroelastoma who did not undergo surgical excision was 13% at 5 years, this was higher than age- and sex-matched rates. Gowda *et al.*[12] recommend surgery if the tumour is mobile as they found that tumour mobility was a predictor of mortality and risk of embolism.

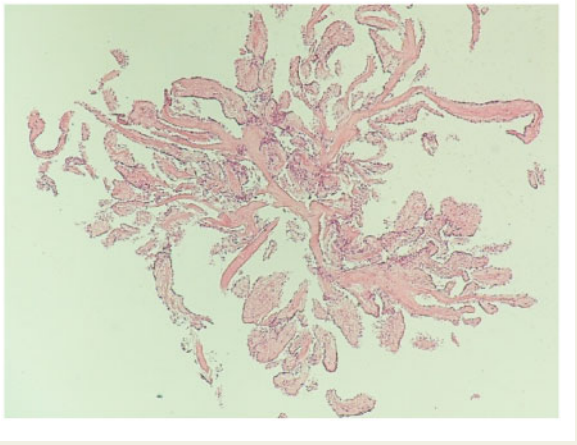

Figure 4 Histology slide, branching papillary fronds with central avascular collagen and variable elastic tissue.

Different surgical techniques for fibroelastoma excision have been reported; shave excision, excision with valve repair, and excision with valve replacement. In one case series, 83% of patients underwent valve-sparing shave excision with no evidence of tumour recurrence at 3 year follow-up.[13]

Although papillary fibroelastoma is a benign tumour it can cause systemic embolization and as shown in this case localized mechanical complications such as transient occlusion of coronary ostia. This case demonstrates the value of transthoracic imaging, which identified a very rare and unusual cause of myocardial infarction.

Lead author biography

Dr Niamh Logan graduated from University College Cork in 2014. She graduated with an honours degree and was awarded the Pearson Medal and John Kelly Prize for academic achievements in the final year examinations. In 2018 she became a member of the Royal College of Physicians, Ireland. She is currently completing her physician training in the Mater Misericordiae University Hospital, Dublin.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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