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CASE REPORT
Cardiovascular imaging

An unusual case of recurrence of papillary fibroelastoma: a case report

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Background Papillary fibroelastomas (PFEs) are uncommon primary cardiac tumours and their recurrence after resection is rare.

Case summary A 32-year-old woman, who was evaluated for recurrent embolic stroke, was found to have lesions on mitral leaflets on echocardiography. The mitral leaflets were otherwise normal with no clinical or laboratory evidence of infective endocarditis. Transthoracic, as well as transesophageal echocardiography, revealed masses on mitral leaflets, which remained almost the same over 3 years. A tentative diagnosis of PFE on the mitral valve was made. She had undergone intra-cardiac excision of mitral valve mass along with ring annuloplasty in February 2020. In November 2021, during her regular follow-up visit, transthoracic echocardiography revealed the recurrence of the masses on mitral leaflets with severe mitral regurgitation. She underwent mitral valve replacement and excision of tumour masses. Histopathological examination confirmed the diagnosis of recurrent PFE.

Discussion Recurrence of PFEs is rare. We report a rare case of histologically proven recurrence of PFE of the mitral valve. This highlights the importance of routine post-operative echocardiographic follow-up in patients with PFE.

Keywords Papillary fibroelastoma • Tumour • Mitral valve • Echocardiogram • Case report

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 6.8 Cardiac tumours

Learning points

- Papillary fibroelastomas (PFEs) are rare cardiac tumours.
- Regular follow-up with echocardiography and high index of suspicion is prudent in the diagnosis of recurrence of PFE.
- Long-term cardiologic surveillance is essential.

Introduction

Primary cardiac tumours are rare.[1] Papillary fibroelastoma (PFE) is the third most common primary tumour, although likely underdiagnosed. The presentation varies from primarily asymptomatic patients to multiple embolic events. The tumour usually involves the cardiac valves and is now being recognized more frequently with the aid of transthoracic (TTE) and transesophageal echocardiography (TEE). These tumours, although benign, do not exhibit local infiltration on histopathologic examination and carry a low likelihood of recurrence.[2]

We report an unusual case of histologically proven case of recurrent PFE of the mitral valve.

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Timeline

Time	Event
2016	Admitted with first stroke; vegetation/mass on mitral valve leaflet and treated as infective endocarditis.
2019	Admitted with recurrent stroke; same mass on mitral valve; diagnosed as papillary fibroelastoma (PFE) of mitral valve by echocardiography.
February 2020	Intra-cardiac excision of mitral valve mass along with mitral ring annuloplasty done.
November 2021	Recurrence of mitral valve mass; with severe mitral regurgitation.
December 2021	Mitral valve replacement done; histopathology confirms PFE of mitral valve.
January 2022	Came for follow-up and is doing Well.

Case presentation

A 32-year-old Asian woman, who was on treatment for hypothyroidism (Tablet Thyronorm 50 mg once daily) and primary infertility (tablet medroxyprogesterone acetate 10 mg twice daily) presented to our hospital in the year 2016 with left-sided hemiparesis and systemic hypertension. Magnetic resonance imaging (MRI) of the brain revealed an acute ischaemic stroke involving the right posterior cerebral artery (PCA) territory (*Figure 1A*). Clinical examination was normal except for a grade 2/6 short systolic murmur at the mitral area. The electrocardiogram showed normal sinus rhythm with non-specific T waves inversions in leads V5 and V6 (*Figure-2*). The inflammatory markers were unremarkable.

Holter monitoring was normal. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) showed a vegetation-like mass of size 13 × 0.5 mm attached to P2 of the mitral valve leaflet. Even though the blood cultures were negative for infective endocarditis, she was empirically treated with intravenous antibiotics (Ceftriaxone 2 g intravenously (IV) once daily (OD) for 4 weeks and gentamycin 150 mg IV OD for 2 weeks). Although she had a complete neurological recovery, the vegetation remained the same. The patient declined to undergo surgery, and so was discharged. As per the neurologist's advice, she was continued on antiplatelets.

After three years, she presented to us with recurrent stroke (right-sided hemiparesis). Brain MRI examination revealed an acute ischaemic stroke involving the left PCA territory, left parietal cortex and left superior frontal gyrus (*Figure 1B*). Blood investigations for young stroke including autoimmune and connective tissue disorders were negative. There was no family history of premature coronary artery disease and/or stroke. Detailed TTE (*Figure 3A*) and TEE (*Figure 3B*) (see *Supplementary material online, Video 1*) showed masses over the anterior mitral leaflet (AML) and posterior mitral leaflet of the mitral valve. She was again treated empirically with IV antibiotics (ceftriaxone 2 g IV OD for 4 weeks and Gentamycin 150 mg IV OD for 2 weeks) even though blood cultures were negative. Although her recovery from stroke was complete, mitral valve lesions remained the same. This made us suspect papillary fibroelastoma (PFE) of the mitral valve. An intra-cardiac excision of mitral valve mass with ring annuloplasty including the surrounding endocardium was done in February 2020. The post-operative period was uneventful. She was discharged on the seventh post-operative day and she was on regular follow-up. During her regular

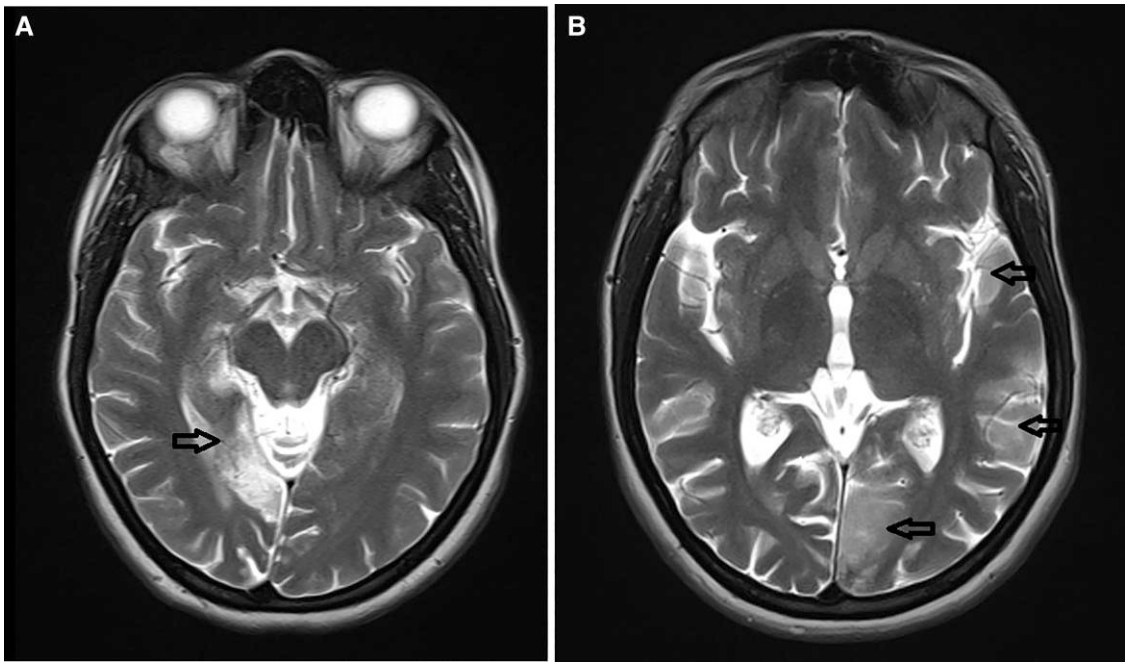

Figure 1 (*A*) MRI brain—T_2-weighted image showing hyperintense signals suggestive of acute infarct in the right posterior cerebral artery territory. (*B*) MRI brain—T_2-weighted image showing hyperintense signals suggestive of acute infarcts involving the left posterior cerebral artery territory, left parietal cortex and left superior frontal gyrus.

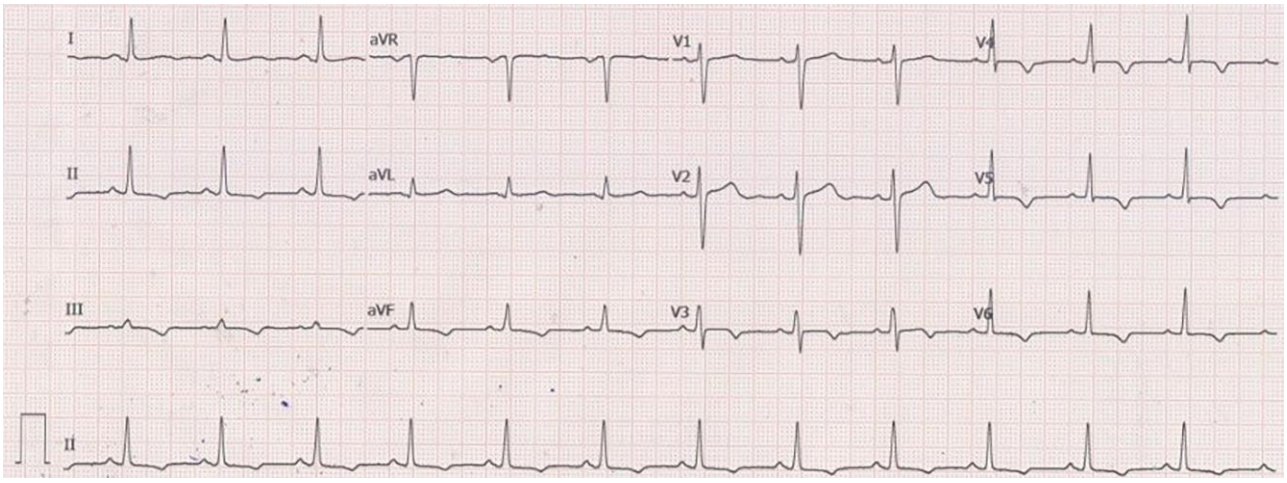

Figure 2 Twelve-lead ECG recorded at 25 mm/s with a gain setting of 10 mm/mV showing normal sinus rhythm with non-specific T waves inversions in leads V5 and V6.

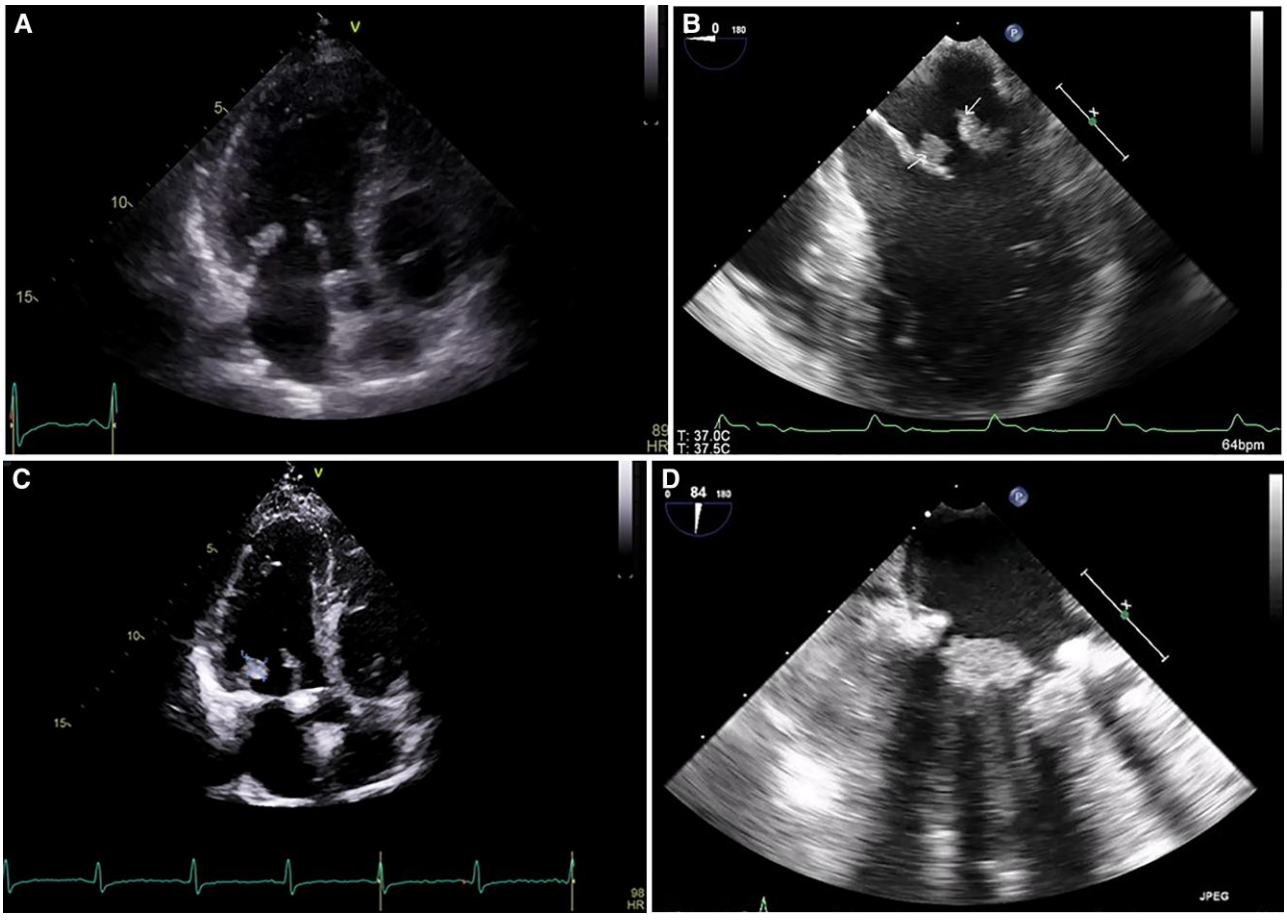

Figure 3 (*A*)TTE-apical four-chamber view showing masses attached to mitral leaflets. (*B*) TEE-midesophageal 2-chamber view showing masses on mitral leaflets. (*C*)TTE-apical four-chamber view showing recurrence of masses on mitral leaflets. (*D*) TEE-midesophageal two-chamber view showing recurrence of masses on mitral leaflets.

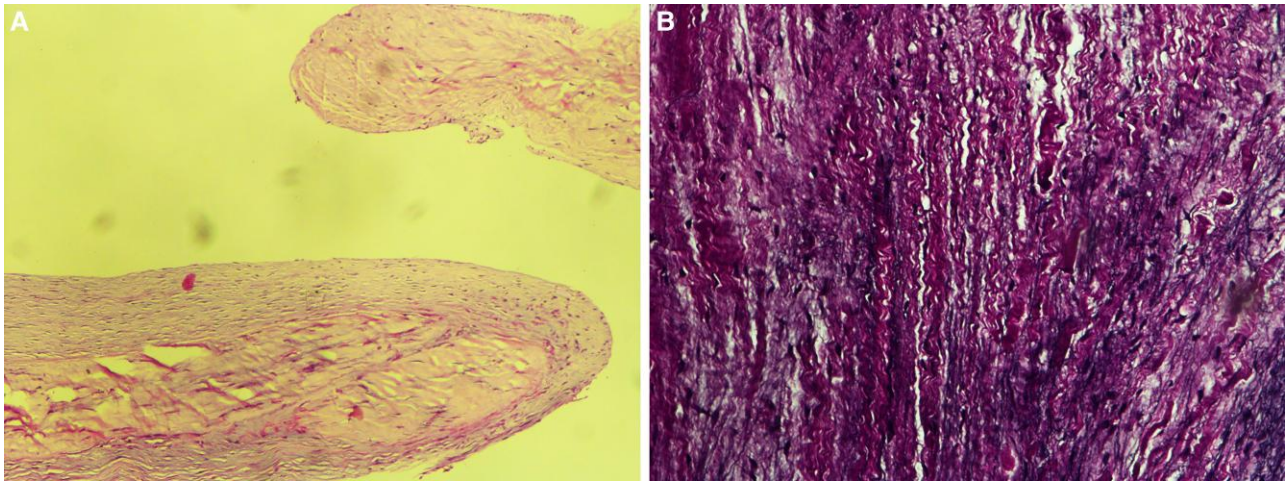

Figure 4 (A) Histological examination of mitral valve tissue. Haematoxylin & eosin staining. Original magnification × 100. Papillary excrescences. (B) Special stain, elastin van Gieson (EVG) staining. Original magnification × 200. Fragmented reticulin fibres.

follow-up visit in November 2021, TTE revealed a mass of size 14 ×11 mm on the P2 of the mitral valve along with severe mitral regurgitation (*Figure 3C*). TEE revealed masses over the mitral leaflets (*Figure 3D*) (see *Supplementary material online, Video 2*). She underwent mitral valve replacement in December 2021. Histopathological examination of the mass revealed papillary excrescences by haematoxylin and eosin staining (*Figure 4A*) along with fragmented reticulin fibres in elastin van geison staining (*Figure 4B*), which confirmed the diagnosis of PFE. Our patient had a complete neurological recovery with a well functioning prosthetic mitral valve. The recurrence of PFE, although rare, can occur. This patient highlights the importance of follow-up echocardiogram in patients after resection of PFE even if the patient is asymptomatic.

Discussion

Our patient who presented with recurrent cardio-embolic stroke was diagnosed to have a recurrence of PFE of the mitral valve. Hence, this is a rare and atypical presentation of the recurrence of PFE of the mitral valve.

PFEs are rare benign cardiac tumours with a prevalence of 0.002% to 0.28%.[2] The average age of diagnosis is 56 years with a primarily male preponderance (58%). While the clinical presentation may vary from clinically asymptomatic to multiple embolic events, cerebrovascular accidents are common. It often arises from the left side of the heart. It has a high propensity towards the aortic valve (44%), the left ventricular outflow tract, and the AML (35%).[1] While aortic valve PFEs present with sudden death and myocardial infarction; systemic tumour embolization is common with mitral valve involvement.[3]

PFEs appear as round, homogenous speckled, mobile, sessile, or pedunculated mass, attached to the endocardium. They are mostly located on the downstream surface of cardiac valves and appear like sea anemone with multiple frond-like projections on echocardiography.[2] These are avascular tumours. It consists of an outer endothelial layer, a dense central core composed of a rim of loose mucopolysaccharide-rich connective tissue, smooth muscle cells, fibroblasts, and dendritic cells.[1] Diagnosis done by imaging is confirmed with histopathological examination and immunohistochemical staining.[4]

Friability of the tissue matrix along with their extreme mobility contribute to the higher embolic potential of these tumours.[4] Dislodgement of the fibrin/platelet thrombi formed on them or papillary frond fragments *per se* leads to thromboembolism.[5] It is formed by minor endothelial damage on valve margins and is the most accepted hypothesis for the development of PFE.[6]

PFEs are screened with TTE. TEE is more sensitive and is of utmost importance for intraoperative guidance.[7] Differential diagnosis includes Lambl's excrescences, myxomas, thrombi, and bacterial vegetation. Cardiac magnetic resonance (CMR) and multi-detector CT are other useful investigations. When imaged with MRI, features are those of a low signal well circumscribed mobile valve nodule on Cine steady-state free precession sequences, often with peri-lesional flow artefact. T_1- and T_2-weighted images reflect their fibroelastic composition with uniform intermediate signal intensity similar to myocardium. Both CT and MR imaging allow evaluation of the extracardiac extent of disease.

Asymptomatic immobile tumours with diameters less than 1 cm are to be closely followed-up. Surgical resection has to be done urgently for all symptomatic patients with mobile pedunculated tumours, increasing in size. Valve-sparing surgery is safe and curative. However, valve replacement should be considered in advanced cardiac valve involvement as in the present case.[8]

Recurrence, although rare can occur.[9] Hence, long-term cardiologic surveillance is essential.

Lead author biography

Dr Vijayaraghavan, Padmasree awardee from The President of India, is the Vice Chairman & Founder Director of KIMSHEALTH and the President of the Society for Continuing Medical Education and Research, Trivandrum, Kerala. He is one of the senior most medical teachers of this country and is widely known in India for establishing the first two-dimensional echocardiography laboratory in India way back in 1980. He graduated from Trivandrum Medical College in 1964 and passed MD in General Medicine in 1969. From Christian Medical College, Vellore, he got Doctorate in Cardiology [D.M (Card)] in 1973.

Dr Vijayaraghavan is the recipient of numerous awards & honours. He received honorary fellowships FRCP Edin, FRCP Lond, FACe, and FAHA.

Supplementary material

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

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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 declare that written consent was acquired from the patient for the submission and publishing of this case report, including images and textual content, in compliance with COPE guidelines.

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