

CASE REPORT

Aortic valve fibroelastoma: a rare cause of stroke

Vivek Kumar, Parita Soni, Arsalan Hashmi, Manfred Moskovits

Department of Internal Medicine, Maimonides Medical Center, Brooklyn, New York, USA

Correspondence to
Dr Parita Soni,
sonipss14@gmail.com

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SUMMARY

The prevalence of primary cardiac tumours varies from 0.02% to 0.45%. Cardiac papillary fibroelastoma (CPF) is a rare tumour diagnosed incidentally on imaging. The clinical manifestations result from thromboembolisation and include transient ischaemic attack, stroke and sudden cardiac death. We present a patient aged 57 years with CPF arising from the aortic valve. The patient presented with right hemiparesis due to acute stroke. He received tissue plasminogen activator with complete resolution of neurological symptoms. Echocardiography revealed a broad-based, gelatinous, non-mobile mass on the left aortic cusp. The tumour was excised sparing the aortic valve. The patient recovered rapidly without any complications. The histopathological examination confirmed the diagnosis of CPF. A review of the literature suggests that CPF is a rare but treatable cause of stroke. The course is not clear and there are no tumours or patient-related characteristics which could predict the risk of thromboembolisation. Surgical treatment is definite and is relatively safe.

BACKGROUND

Primary cardiac tumours are rare with a prevalence of 0.02% in an autopsy and 0.45% in an open heart surgery study.¹ Cardiac papillary fibroelastoma (CPF) is the most prevalent valvular and overall second most common cardiac tumour after myxoma.² They affect the aortic valve more often and are recognised more frequently now due to the availability of higher resolution imaging techniques.³ CPFs are histologically benign and frequently asymptomatic. Only occasionally, they manifest with symptoms of transient ischaemic attacks, stroke, heart failure, myocardial infarction, cardiac arrest and sudden cardiac death.⁴ Herein, we present a rare case of a patient with fibroelastoma of the aortic valve presenting with stroke.

CASE PRESENTATION

An Asian man aged 57 years with a medical history of hypertension (on losartan), presented to the emergency room with sudden onset of weakness in his right upper and lower extremities for 2.5 hours duration. He denied any facial droop, visual disturbances or slurred speech. On presentation, his vital signs were stable with blood pressure of 146/87 mm Hg. Physical examination revealed weakness in right upper (3/5 power) and lower extremities (power 4/5). Sensory and cranial nerve examination was unremarkable. Examination of the left side was normal. Cardiac examination revealed normal heart sounds without any murmurs. Laboratory values, including complete blood count, HBA1C, kidney,

liver and lipid panels, were inconspicuous. ECG and chest X-ray were also normal. Non-contrast CT of the head and CT angiogram (CTA) head were negative for any acute intracranial pathology. The patient was eligible for treatment with tissue plasminogen activator and was transferred to the intensive care unit. His symptoms improved significantly post-therapy. A follow-up non-contrast MRI was obtained on the next day which did not reveal any area of restricted diffusion.

Duplex examination was negative for any significant stenosis or thrombosis of carotid and vertebral arteries. Transthoracic echocardiogram was performed as part of stroke work-up, which revealed a mass on the aortic valve suspicious for vegetation. For better evaluation, transoesophageal echocardiogram (TEE) was performed which showed a non-mobile mass on the aortic valve measuring 9×6 mm, gelatinous in consistency with broad-based attachment to the edge of the left coronary cusp (video 1 and figure 1). Cardiothoracic surgery team was consulted and the patient underwent excision of the tumour with sparing of the aortic valve (figure 2). Histopathological assessment of the resected specimen confirmed the diagnosis of fibroelastoma (figure 3).

Postoperative TEE showed no residual tumour and confirmed the integrity of the valve with only trace regurgitation. The patient recovered rapidly without any complications with absolutely normal neurological examination.

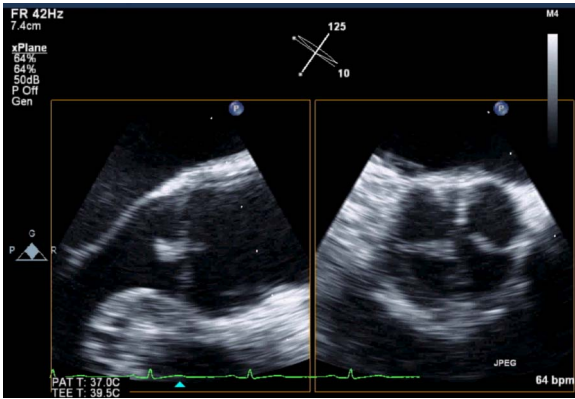
DISCUSSION

Primary tumours of the heart are rare, they are usually benign and they occur mostly in adults. Papillary fibroelastoma accounts for <10% of all cardiac tumours, but they are most common valvular tumours.⁵ Previous studies have shown that more than two-thirds of papillary fibroelastomas are found incidentally preoperatively or intraoperatively. They are solitary tumours mostly <20 mm.² Most papillary fibroelastomas are located in the left heart, which increases the risk of systemic embolism.⁶ The natural history and course of this tumour is not known.⁷ The symptoms are mostly due to thromboembolisation originating from the tumour itself or from thrombus formation on the tumour surface.⁸ The definite treatment is valve-sparing excision of tumour with low recurrence rate.⁹ Surgical resection is indicated in all symptomatic patients and should be offered to incidentally diagnosed patients during evaluation of cardiac surgery for other reasons.² In other patients, the treatment is debatable for incidentally diagnosed tumours and depends on their size, location, mobility and associated comorbidities. In such patients, surgery is



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Video 1 Video clip of the transoesophageal echocardiogram showing the tumour attached to the left aortic valve cusp in the parasternal long-axis view (left side) and the parasternal short-axis view (right side).



Figure 1 Long-axis view of the transoesophageal echocardiogram image showing mass on the left aortic valve cusp.

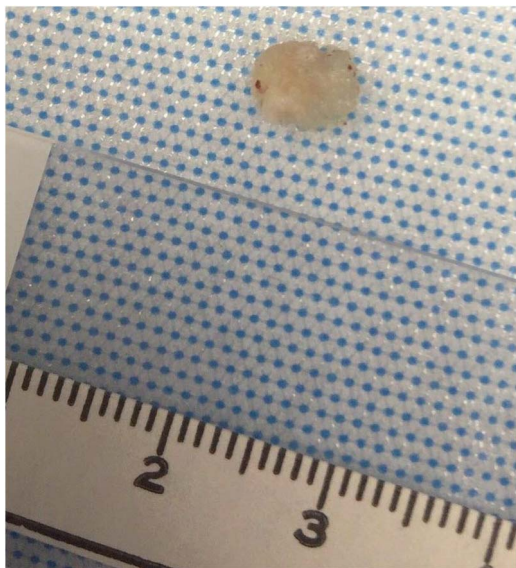


Figure 2 The gross specimen of resected tumour appearing like gelatinous nodule which is different from typical appearance resembling a sea anemone described in the literature.

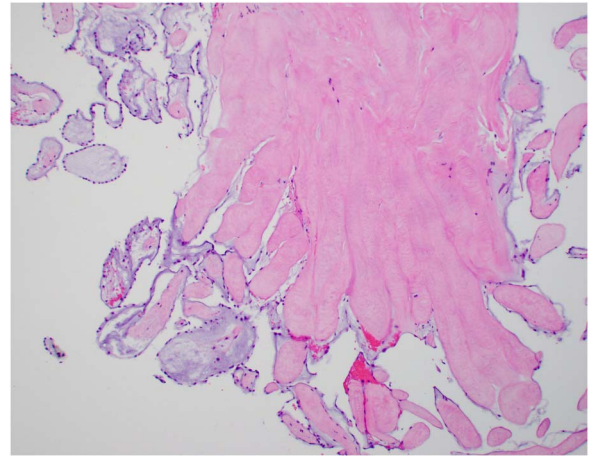


Figure 3 Histological examination under high-power field displaying a mass composed of multiple fronds of acellular collagen and dense elastin covered by flat endothelial cells highly characteristic of papillary fibroelastoma.

reasonable for pedunculated lesions or tumours larger than 1 cm in diameter, mobile lesions on echocardiography or if located adjacent to coronary ostia.¹⁰

Medical management with oral anticoagulation has been advocated for smaller, non-mobile tumours and in cases where valve-sparing surgery is not possible. Of note, this approach was associated with 50% tumour-related mortality in one study and has not been universally accepted.¹¹

Despite being histologically benign, CPF can result in fatal complications such as stroke, acute valvular dysfunction, embolism, ventricular fibrillation and sudden death.⁴ However, cerebrovascular complications have been associated with tumours of diameter as small as 3 mm.¹² Of note, our patient developed stroke, despite the tumour appearing non-mobile on TEE. Previous reports also suggest that echocardiographic appearance may not predict the risk of thromboembolisation.² The excision of papillary fibroelastoma is a relatively safe procedure and is indicated in all symptomatic patients.² Asymptomatic patients should also be carefully assessed for surgery to prevent catastrophic events in future.

Learning points

- ▶ Cardiac papillary fibroelastoma most commonly affects the aortic valve and are an uncommon cause of stroke. Its diagnosis can be established reliably only on histopathology.
- ▶ The natural history is unknown and there are no patients or tumour-related characteristics which could reliably predict the risk of thromboembolisation.
- ▶ The shave excision of tumour is a relatively safe procedure. Therefore, even asymptomatic patients should be actively assessed for surgery to prevent future complications.
- ▶ The medical management involves oral anticoagulation in selected patients to prevent thromboembolisation from tumour surface; however, such approach has been found to increase the tumour-related mortality in some series and is not widely accepted.

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