CASE REPORT

Cardiac papillary fibroelastoma presenting as acute stroke

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Dr Atif Saleem Abbasi,
atifabbasi655@hotmail.com**SUMMARY**

We present a case of a young woman who was initially diagnosed with acute stroke with no obvious risk factors. Preliminary investigation with transthoracic echocardiography and subsequent advanced imaging with transoesophageal echocardiography suggested the diagnosis of a benign cardiac tumour on the anterior leaflet of mitral valve. The patient underwent urgent surgical resection. Histology confirmed the diagnosis of cardiac papillary fibroelastoma. She made complete clinical recovery with no recurrence of symptoms.

BACKGROUND

This case illustrates the importance of diagnosing primary cardiac tumours in a young patient presenting with stroke like symptoms with no obvious risk factors. One should always look for secondary cause of stroke in such patients and a part of investigations should include transthoracic and transoesophageal echocardiography. Although primary cardiac tumours are rare, diagnosis and treatment at an early stage may prevent serious complications such as lethal arrhythmias, systemic emboli, heart failure and sudden death.

CASE PRESENTATION

A 42-year-old, previously healthy woman with a history of depression was brought in by ambulance in the early morning with right facial numbness and right upper limb weakness. Collateral history elicited from the patient's partner revealed that the patient developed a sudden onset of right facial droop and slurred speech lasting approximately 10 min. These symptoms had completely resolved upon presentation. On further questioning, the patient described sporadic headaches and palpitations over the previous year. She was taking oral coamoxiclav for her upper respiratory chest infection prescribed by her general practitioner 1 week before. She is a music teacher at a local school and lives with her long-term partner. She is an active smoker of 10–15 pack-years. Her father had a history of atrial fibrillation and died of a cerebrovascular accident in his late 80s. On physical examination, the patient's vital signs were stable. She was alert, oriented with full Glasgow Coma Scale. There was no focal neurological deficit on neurological examination and her cardiovascular examination was normal with no murmurs, added sounds or carotid bruit.

INVESTIGATIONS

Full blood count, urea, electrolytes and liver function tests were all within normal limits.

High-sensitivity troponin T assays were normal. ECG showed normal sinus rhythm.

A negative thrombophilia screen (protein C, protein S deficiency, activated protein C resistance and factor V Leiden mutation).

CT of the brain showed an area of altered, low attenuation in the left parieto-occipital region. Further imaging was required on the advice of the attending radiologist.

MRI of the brain showed a small focus of restricted effusion in the left posterior parietal lobe in the region of postcentral gyrus consistent with a small recent left-hemispheric infarct. There was no evidence of intracranial haemorrhage.

Transthoracic echocardiogram showed a spherical mass attached to the anterior mitral valve leaflet of approximately 0.8 cm in diameter (figure 1 and video 1). The mitral valve opened well with no evidence of stenosis, regurgitation or left ventricular outflow tract obstruction. Left ventricular function was normal with an ejection fraction estimated by Simpson's rule of approximately 55%. A bubble study was negative which ruled out patent foramen ovale. A transoesophageal echocardiogram was performed to further evaluate the mass and to rule out further mechanisms of thromboembolism such as patent foramen ovale. It confirmed the findings of transthoracic echocardiography (figure 2).

DIFFERENTIAL DIAGNOSIS

A wide range of differential diagnosis was included, such as

- ▶ Congenital thrombophilia;
- ▶ Patent foramen ovale;
- ▶ Antiphospholipid syndrome.

Figure 1 Transthoracic echocardiogram of four-chamber apical view showing a spherical mass attached to the anterior mitral valve leaflet of approximately 0.8 cm in diameter.

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Video 1 Mass attached to anterior mitral valve leaflet.

Her thrombophilia screen and bubble studies were negative, a preliminary diagnosis of a cardiac tumour was made on the basis of echocardiography.

TREATMENT

The patient was given antiplatelet therapy (aspirin 75 mg), a lipid-lowering agent (atorvastatin 40 mg) and an intravenous infusion of unfractionated heparin to mitigate the risk of further thromboembolic events. The patient was promptly referred for cardiothoracic surgical review. The decision was made to surgically excise the mass.

During her admission, attention was then directed to the appropriate management of the cardiac mass whether or not her symptoms might be attributable to a cardioembolic event. Given the patient's age, size and mobility of the cardiac mass, it was recommended to proceed with surgical removal of the tumour.

The patient underwent an elective surgical resection under cardiopulmonary bypass. Intraoperatively, the anterior mitral leaflet mass was identified as a 5×3×7 mm (figures 3 and 4) mushroom-shaped mass on the body of the anterior mitral leaflet with fronds—very friable. No additional masses or valvular abnormalities were noted. The lesion was broad-based which excised leaving circular defect in the anterior mitral leaflet. The anterior leaflet was repaired with an autologous pericardium patch. The surgical procedure was completed by ring annuloplasty. Intraoperative transoesophageal echocardiography confirmed a competent valve repair. A histopathological examination of the tissue revealed a benign papillary fibroelastoma. The patient had an uneventful recovery postoperatively. She remained on anticoagulation therapy with

Figure 2 Transoesophageal echocardiography: parasternal long axis view showing mushroom-shaped friable mass attached to the anterior leaflet of mitral valve.

Figure 3 An intraoperative image showing the anterior mitral leaflet mass.

warfarin for 3 months without any further neurological sequelae. Postoperative echocardiography showed satisfactory mitral valve function with no regurgitation or stenosis.

OUTCOME AND FOLLOW-UP

The patient was reviewed in the clinic 6 months postsurgery and had no recurrent events.

DISCUSSION

Primary cardiac tumours are rare constituting only around 8% of all cardiac tumours.¹ They are usually discovered incidentally on cardiac MRI, echocardiogram and transoesophageal echocardiography.² They often (85%) originate from the valvular endocardium, most commonly the aortic valve (29%), mitral valve (25%), tricuspid valve (17%) and pulmonary valves (13%) involved in that order.³

They can present with a wide spectrum of clinical conditions including heart failure by obstructing the cardiac output, cardiac arrhythmias, thromboembolic events and neurological symptoms.⁴

Despite the benign nature of this tumour, it carries very high risk of embolic complications including neurological deficit.⁵ The fragile nature and frond-like papillary tissues of the tumour itself is prone to thromboembolism.⁴ Therefore, once diagnosed, urgent surgical management is indicated even in an asymptomatic patient.^{5–9}

Figure 4 A mushroom-shaped mass of 5×3×7 mm.

Of cardiac valve tumours, however, papillary fibroelastomas are the most common.¹

A cardiac papillary fibroelastoma (CPF) is a rare benign tumour of the heart with a tendency towards valvular involvement. Although most patients with these tumours are adults with a mean age of 50 years, but it can occur in any age group.^{3 4}

The tumours are small, ranging in size from 2 to 28 mm (median 8 mm), with 99% being <20 mm. Nearly half of all CPFs are mobile with a stalk, and in about 9% of cases there can be multiple.²

The histological characteristics of the tumour consist of multiple papillary villous fronds radiating from a central fibrocollagenous stalk, with each frond showing three zones: a central core that contains collagen, reticulin and elastin; a peripheral myomatous zone; and an outer rim of hyperplastic endothelial cells. It is this architecture of papillary fronds that distinguishes papillary fibroelastoma from cardiac myxoma.² Echocardiography is the most common imaging modality with the typical characteristics have been described, a definitive echocardiographic diagnosis may be challenging with a differential diagnosis that includes vegetations, thrombi, degenerative valve lesions and other tumours. When possible, transoesophageal echocardiography should be performed in patients to accurately define its size, attachments, number and presence of a stalk.²

Surgical considerations specific to CPF include defining the risk of embolisation in order to consider elective resection and the surgical approach.^{2-4 10}

Prompt excision, preserving the original valvular tissue and function when possible, is indicated for all symptomatic patients as well as for those who are asymptomatic, considering the high risk of embolisation with such tumours. These are benign neoplasms with the propensity for severe symptoms, if systemic embolisation or valve obstruction occurs. The treatment of choice for papillary fibroelastomas is surgical excision, which is safe without causing significant morbidity or mortality.¹¹ However, valve replacement or repair may be required when the tumour has invaded the valve or when there is combined degeneration of the valve.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Madhu Sankar N, Odayan MK, Morris M, *et al.* Cardiac valvular papillary fibroelastoma. *Tex Heart Inst J* 1999;26:298–9.
- 2 Sun JP, Asher CR, Yang XS, *et al.* Clinical and echocardiographic characteristics of papillary fibroelastomas: a retrospective and prospective study in 162 patients. *Circulation* 2001;103:2687–93.
- 3 Edwards FH, Hale D, Cohen A, *et al.* Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127–31.
- 4 McAllister HA, Fenoglio JJ. *Tumors of the cardiovascular system*. In: *Atlas of tumor pathology*, 2nd series, Washington DC: Armed forces Institute of Pathology, vol. 15, 1978:20–5.
- 5 Grinda JM, Couetil JP, Chauvaud S, *et al.* Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. *J Thorac Cardiovasc Surg* 1999;117:106–10.
- 6 Domenech A, Arenaza DP, Rivello HG, *et al.* Surgery for papillary fibroelastoma with uncommon location in left ventricle. *Asian Cardiovasc Thorac Ann* 2010;18:174–6.
- 7 Boodhwani M, Veinot JP, Hendry PJ. Surgical approach to cardiac papillary fibroelastomas. *Can J Cardiol* 2007;23:301–2.
- 8 Darvishian F, Farmer P. Papillary fibroelastoma of the heart: report of two cases and review of the literature. *Ann Clin Lab Sci* 2001;31:291–6.
- 9 Gopaldas RR, Atluri PV, Blaustein AS, *et al.* Papillary fibroelastoma of the aortic valve-operative approaches upon incidental discovery. *Tex Heart Inst J* 2009;36:160–3.
- 10 Howard RA, Aldea GS, Shapira OM, *et al.* Papillary fibroelastoma: increasing recognition of a surgical disease. *Ann Thorac Surg* 1999;68:1881–5.
- 11 Kurian KC, Edwards FH, Jacob B, *et al.* Papillary fibroelastoma presenting as left ventricular mass. *Tex Heart Inst J* 2006;33:63–5.

Learning points

- Benign cardiac tumours should be considered in the differential diagnosis of the aetiology of stroke in young patients with minimal or no risk factors.
- Appropriate use of imaging modalities such as transthoracic and transoesophageal echocardiography should be used for diagnosing cardiac tumours.
- Efficient diagnosis and prompt surgical intervention can reduce morbidity and mortality from embolic stroke.

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