Incidental cardiac papillary fibroelastoma: a management dilemma

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DESCRIPTION

A 78-year-old woman with a history of diastolic heart failure and paroxysmal atrial fibrillation (PAF) presented after a syncope. She was on warfarin therapy for PAF. Her cardiovascular examination was essentially unremarkable. Her orthostatic vital signs were stable and electrocardiogram revealed a normal sinus rhythm. Laboratory examination was unremarkable and her international normalised ratio (INR) was 2.5. Telemetry monitoring was largely unremarkable. Transthoracic echocardiography showed a mobile pedunculated mass (2.2 cm in the largest dimension) on the ventricular aspect of the anterior mitral valve, with a homogeneous speckled pattern and a characteristic stippling along the edges, most likely consistent with an incidental cardiac papillary fibroelastoma (CPFE) (figure 1 and video 1). Transoesophageal echocardiography was also consistent with similar findings of CPFE (video 2). The patient was discharged with a holter monitor with a discharge plan for follow-up with cardiothoracic surgery.

Primary cardiac tumours are rare with an incidence of less than 0.03%.¹ CPFE is the second most common cardiac tumour after myxoma.² More than 70% of CPFE are benign, and only 8% are papillary fibroelastomas, mostly located on the heart valves. CPFE is more commonly seen on the aortic valve closely followed by the mitral valve. It is usually more prevalent in middle-aged women. More than half of the patients are asymptomatic.³ The common complications include transient ischaemic attack, stroke, syncope, acute myocardial infarction and cardiac arrest, due to either embolism or mechanical obstruction.³ It is usually

Figure 1 Transthoracic echocardiogram shows a mobile pedunculated mass (2.2 cm in the largest dimension) on the ventricular aspect of the anterior mitral valve, with a homogeneous speckled pattern and a characteristic stippling along the edges, most likely consistent with an incidental cardiac papillary fibroelastoma.

diagnosed on echocardiography and can be confirmed with biopsy. In typical cases, papillary fibroelastoma phenotypically resembles a sea anemone.

Although CPFE is rare, its associated complications can be serious and life-threatening. When symptomatic, the recommended treatment is surgical excision. The dilemma occurs when it is diagnosed incidentally. There are no clear guidelines on the management of asymptomatic CPFE. Because of their potential for causing cerebral and coronary embolisation, many experts recommend surgical excision even in asymptomatic patients. Thus, it

Video 1 Transthoracic echocardiogram shows a mobile pedunculated mass (2.2 cm in the largest dimension) on the ventricular aspect of the anterior mitral valve, with a homogeneous speckled pattern and a characteristic stippling along the edges, most likely consistent with an incidental cardiac papillary fibroelastoma.

Video 2 Transesophageal echocardiogram is again consistent with findings of cardiac papillary fibroelastoma.

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would be advisable that if a patient is a low-risk surgical candidate, excision should be undertaken, especially if the tumour is left sided, mobile and more than 1 cm in size.⁴

Learning points

- ▶ Cardiac papillary fibroelastoma (CPFE) is the second common primary cardiac tumour with the most common location being aortic valve, followed by mitral valve.
- ▶ Because of the potential risk of embolism and mechanical obstruction, it can lead to serious morbidity or life-threatening situation.
- ▶ Symptomatic CPFE should be managed surgically, and surgical excision should also be strongly considered for asymptomatic patients especially if the tumour is left-sided, mobile and greater than 1 cm in size.

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