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Case Report

Uncommon Location of a Papillary Fbroelastoma: Case Report

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ABSTRACT

An elderly man, with a history of diabetes and hypertension presented to our hospital complaining of attack of syncope and palpitations. Echocardiogram revealed the presence of a pedunculated mass attached to the interventricular septum. Sternotomy was performed and ascending aorta was opened transversely, aortic valve leafets were retracted, and a tumour was resected. The postoperative course was uneventful; the patient was discharged after 1 week from the operation. This case demonstrates atypical location for fibroelastoma on the interventricular septum, thus underpinning the need for proper assessment of all patients with a history of systemic embolization to rule out any unusual intracardiac causes.

RÉSUMÉ

Un homme âgé ayant des antécédents de diabète et d'hypertension s'est présenté à notre hôpital pour des épisodes de syncope et des palpitations. L'échocardiographie a mis en évidence une masse pédiculée fixée au septum interventriculaire. Une sternotomie a été pratiquée; l'aorte ascendante a été ouverte transversalement, les feuillets de la valve aortique ont été rétractés, et une tumeur a été retirée. La période postopératoire s'est déroulée sans incident; le patient a obtenu son congé une semaine après l'intervention chirurgicale. Ce cas montre l'emplacement atypique d'un fibroélastome sur le septum interventriculaire, confirmant qu'il faut procéder à une évaluation appropriée de tous les patients ayant des antécédents d'embolisation générale afin d'exclure toutes les causes intracardiaques inhabituelles.

Cardiac papillary fibroelastoma is a rare benign cardiac tumour, 90% of which is attached to the cardiac valves.[1] Symptoms, if present, are due to flow obstruction or peripheral embolization. We present an unusual case where a papillary fibroelastoma was found attached to the interventricular septum (IVS) and was treated surgically.

Case Presentation

A 76-year-old man with a history of diabetes and hypertension presented to our institution with a first attack of syncope and palpitations. Electrocardiogram showed normal sinus rhythm. Transthoracic echocardiography revealed a mobile mass attached to the ventricular septum approximately 1 cm from the aortic valve and trace to mild central aortic regurgitation (Fig. 1). Left ventricular systolic function was normal. Coronary angiography showed 80% left anterior descending artery stenosis. The patient was referred for cardiac surgery.

Preoperative transesophageal echocardiogram revealed the presence of a pedunculated mass attached to the IVS fopping in and out of the left ventricular outfow tract (Fig. 2; Videos 1 and 2, view video online).

A median sternotomy was performed. Standard cardiopulmonary bypass was established with monocaval cannulation. The left anterior descending territory was revascularized by the left internal mammary artery. The ascending aorta was opened transversely, aortic valve leafets were retracted, and a gelatinous looking tumour was resected with its pedicle, with a safety margin 0.5 cm from the surrounding IVS (Supplemental Fig. S1A). Histopathologic examination demonstrated the presence of multiple papillary with individual fronds consisting of a core of hyalinized hypocellular stroma rich in elastic fibres lined by hyperplastic endocardial cells (Supplemental Fig. S1B). This appearance is typical of a papillary froelastoma (PFE).

The postoperative course was uneventful; the patient was discharged on postoperative day 7.

Discussion

PFE is the third most common primary cardiac tumour, after myxoma and fbroma. It is histologically benign and avascular, and almost 90% of the previously reported PFEs

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Ethics Statement: The research was approved by Institutional Research Borad in Prince Sultan Cardiac Center - AlHassa and it adehrent to good clinical guidelines.

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See page 123 for disclosure information.

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Novel Teaching Points

- Fibroelastoma commonly presents in left heart valves, but it presents in atypical locations such as IVS, as in our case.
- It is a rare benign tumour that has potential risk for systemic emboli.
- The most appropriate and sensitive diagnosis is made by transesophageal echocardiography.
- Surgery remains the sole option for treatment to prevent the complications.

were located on valves, particularly the aortic valve (29%). The mitral valve was the second location of involvement (25%), followed by tricuspid (17%) and pulmonic valvular (13%). Scarcely 10% of the fibroelastoma cases developed from the left ventricular endocardium,[1] as was seen in our patient. Early diagnosis avoids complications such as pulmonary or paradoxical embolism into the systemic circulation.

PFEs are frequently single tumours, rarely multiple, relating to the same or separate valves or both the left and the right cavities.[2] Most cases are detected in asymptomatic patients as incidental findings during cardiac imaging.

Symptomatic patients are commonly found to have transient ischaemic attacks, cerebrovascular accident, myocardial infarction, heart failure, arrhythmias, pulmonary embolism, blindness, peripheral embolism, and sudden death. The most frequent clinical manifestations are caused by embolism to the cerebral, systemic, or coronary arterial circulations.[3]

Embolism usually happens from either the tumour itself or the clot situated within the tumour fronds. Because PFEs more frequently develop from the higher pressure ventricular surface of valves, they have a higher risk of thromboembolism (34%) compared with tumours originating from the lesser pressure atrial chambers such as myxomas (24%).[4]

The cause of fibroelastoma is ambiguous, in addition to classic tumours; some researchers have suggested that they may occur as a result of viral infection, or mechanical trauma to valve leaflets.[5]

The diagnosis is generally by 2-dimensional or transesophageal echocardiography. Recently, 3-dimensional echocardiography, magnetic resonance imaging, and multislice spiral computed tomography have been used to distinguish it from other tumours.[6]

Echocardiography often reveals a tiny, mobile, pedunculated, or sessile valvular or endocardial mass, which occasionally flutters or prolapses into the cardiac chambers during systole or diastole. Echodensity of the central collagen core robustly underpins the diagnosis and permits discrimination from other intracardiac tumours, vegetations, or mural thrombi.[4]

Management of PFE depends on its clinical manifestations. Patients who are asymptomatic, with a small tumour which is sessile, fixed, and has no evidence of encroachment on the coronary ostia, normally need regular follow-ups with consecutive imaging studies and prompt anticoagulation. Surgical intervention is indicated only if the tumour increases in size or if the tumour is mobile or pedunculated.[7] Post-surgical anticoagulant therapy is not unanimously recommended.[7] Surgical excision of PFE has a low operative risk and provides outstanding short- and long-term outcomes.

Recurrent PFE after surgical excision is infrequent and requires long-term transesophageal echocardiogram follow-up studies.

A minimally invasive surgical approach, particularly partial sternotomy, offers the opportunity for rapid recovery; enhanced cosmesis, with the potential for optimal patient satisfaction in the absence of other pathology requiring open cardiac surgery.[8]

Conclusion

Cardiac papillary fibroelastoma is an infrequent cardiac tumour with the preponderance occurring on left-sided cardiac valves.

Urgent removal of the tumour may prevent tumour-related vascular, embolic, or neurologic complications.

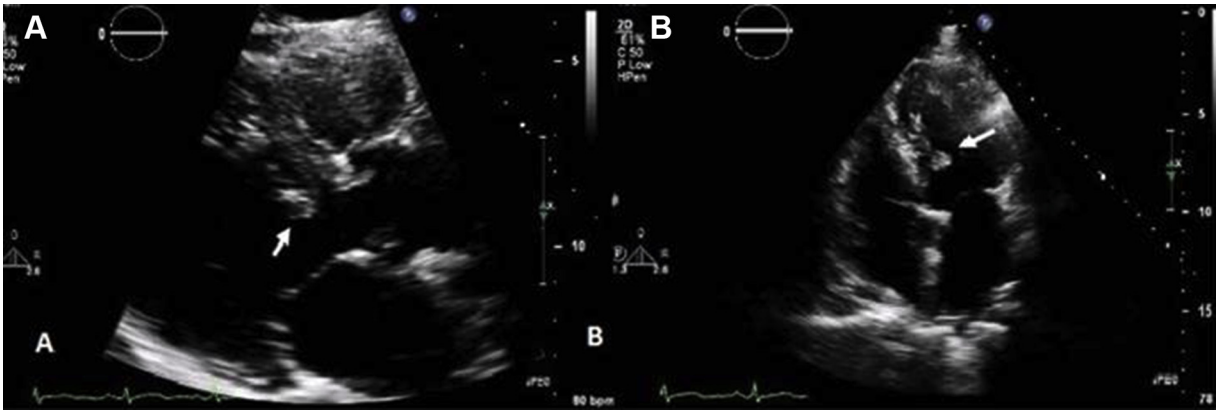

Figure 1. (**A**) Parasternal long-axis view shows a small rounded mass attached to the proximal interventricular septum approximately 1 cm from the aortic valve (**white arrow**). (**B**) Apical 4-chamber view systolic frame demonstrates a rounded mass attached to the basal interventricular septum (**white arrow**).

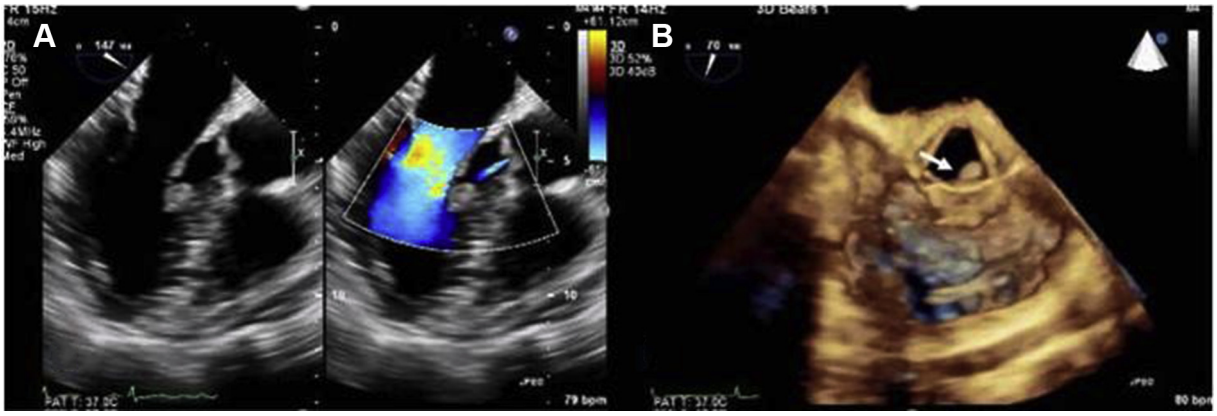

Figure 2. (**A**) Mid-esophageal long-axis view at 147° shows a rounded mass (1 cm × 0.8 cm) attached to the basal interventricular septum approximately 1 cm away from the aortic valve (**white arrow**). There is trace to mild aortic regurgitation not related to the mass. (**B**) Preoperative 3D transesophageal picture at 70° shows the aortic valve in systole seen from aortic prospective; during this systolic frame, the mass is clearly seen (**white arrow**) below the aortic valve. In real time, the motion of the mass is well appreciated.

Treatment with surgical removal tends to be curative and minimizes the risk of recurrence .[8]

Our case report demonstrates the likelihood of atypical location for fibroelastoma on the IVS, thus underpinning the need for proper echocardiographic assessment of all patients with a history of systemic embolization to rule out any unusual intracardiac causes.

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Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit *CJC Open* at https://www.cjcopen.ca/ and at https://doi.org/10.1016/j.cjco.2020.08.013.