CASE REPORT

Papillary fibroelastoma diagnosed through multimodality cardiac imaging: a rare tumour in an uncommon location with review of literature

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SUMMARY

We describe the case of a woman presenting with transient ischaemic attack, who was found to have a papillary fibroelastoma arising from the aortic wall, an extremely rare location. We describe the multimodality imaging techniques used in diagnosing this patient and review the most recent literature on evaluation and management of patients with cardiac papillary fibroelastomas.

BACKGROUND

Cardiac papillary fibroelastomas (CPFs) are exceedingly rare primary cardiac tumours. The majority are located on valvular surfaces. While most patients are asymptomatic, their complications include strokes, myocardial infarctions and sudden cardiac death. Their diagnosis and management can be very complicated.

CASE PRESENTATION

A 62-year-old woman with a medical history of diabetes mellitus, hypertension and hyperlipidaemia presented to the emergency room with left arm and leg weakness. The weakness had started the day prior to her presentation to the emergency room, and had persisted until her presentation. Symptoms were associated with a sensation of numbness on the affected side. There was no associated headache, visual or speech disturbances or facial droop. The patient had no prior cardiac history, and denied chest pain, palpitations or shortness of breath associated with this episode.

The symptoms lasted for less than 24 hours and had subsided by the time of her evaluation in the emergency room. Her physical examination showed no focal neurological deficits. Cardiac auscultation revealed a normal S1 and S2 with no murmur or added heart sounds.

INVESTIGATIONS

Her electrolytes and blood counts were all within normal limits, and an ECG performed showed sinus tachycardia with non-specific ST depressions in the inferior and lateral leads. An MRI of the brain with gadolinium contrast showed no evidence of a stroke or cerebral haemorrhage. The patient was diagnosed to have likely had a transient ischaemic attack (TIA). As part of the evaluation for her TIA, a two-dimensional transthoracic echocardiogram (2D-TTE) was ordered. The 2D-TTE showed a left ventricular ejection fraction of 67% with no regional wall motion abnormalities. However, it also noted an independently mobile 0.8 cm mass attached to the aortic valve at the insertion into the aorta, concerning for infective endocarditis or papillary fibroelastoma. A cardiac MRI was performed for better characterisation of the noted mass, which demonstrated the same suspicious mass in the ascending aorta ([figure 1A](#)). Imaging of the aortic leaflets in the closed ([figure 1B](#)) and open ([figure 1C](#)) position showed that the aortic valve leaflets were structurally normal and free of any attached mass. Because of the inconclusiveness of the two imaging modalities, the patient underwent a real-time 3D-transesophageal echocardiogram (RT-3DTEE). This revealed a 1.1 cmx0.6 cm highly mobile, irregular mass attached to the aortic wall adjacent to the commissural area between the left and right coronary cusp of the aortic valve ([figure 2](#), [figure 3](#)). There was trace aortic regurgitation.

TREATMENT

The patient was subsequently admitted to cardiothoracic surgery and underwent surgical resection of the aortic mass. Intraoperatively, the mass was noted to be pedunculated, with gross appearance suggestive of a fibroelastoma, with the location and size matching what was noted on the RT-3DTEE ([figure 4](#)). Histopathological analysis of the mass concluded that the mass was a benign papillary fibroelastoma.

Figure 1 Images from cardiac MRI showing the mass in the ascending aorta (A), and the structurally normal aortic valve leaflets in the closed (B), and open (C) positions.

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Figure 2 Real-time three-dimensional transesophageal echocardiogram: X Plane imaging showing the mass attached to the aortic wall without involving the aortic leaflets.

OUTCOME AND FOLLOW-UP

The patient tolerated the surgical excision well and was discharged in a stable condition. She was seen in clinic for follow-up of 2 months postoperatively. She was asymptomatic and noted to have no evidence of tumour recurrence.

DISCUSSION

We describe a case of a papillary fibroelastoma arising from the aortic wall, which is in an extremely rare location. Primary cardiac tumours are exceedingly rare with a published incidence of <0.1%. CPF are benign pedunculated tumours of the heart and are the second most common primary cardiac tumour after myxomas. Their appearance is often compared with sea anemones, with frond-like arms emanating from a stalked central core.¹ Histologically, they consist of avascular papillary tissue with a surrounding layer of endothelium.² The tumours vary in size from 2 to 70 mm with the mean being of 9 mm. More

Figure 3 Real-time three-dimensional transesophageal echocardiogram showing the mass (Red arrow) attached to the aortic wall adjacent to the commissural area between the left and right coronary cusp of the aortic valve.

Figure 4 Intraoperative image (A) and gross appearance (B) of the resected papillary fibroelastoma.

than 80% occur on valvular surfaces with the most commonly affected valve being the aortic valve.^{3 4} The remaining 20% occur on extravalvular surfaces of the heart with locations including, in order of decreasing frequency, left ventricle, left atrium, left atrial appendage, atrial septum, right atrium, right atrial appendage, eustachian valve and right ventricle.⁴ However, CPF arising from the wall of the ascending aorta, as seen in our patient, are exceedingly rare with only nine such cases previously being reported in literature.^{5–13} This location was not reported in the two largest reviews published, evaluating and characterising a total of 887 cases of CPF.^{3 4}

Risk factors for its development are unknown and most are diagnosed incidentally with an average age of detection of 60 years. Since the majority of CPF are located on the left side of the heart (>95%), systemic embolisation is common. In patients who are symptomatic, the most common presentations are TIAs, strokes, angina, myocardial infarctions, peripheral embolisms and sudden cardiac death.⁴

The imaging modality most commonly used for diagnosis is transthoracic echocardiography (TTE) or transesophageal echocardiography (TEE). Other imaging modalities used include cardiac MRI and chest CT. TTE by itself has excellent diagnostic sensitivity and specificity for CPF (88.8% and 88.7%, respectively).³ However, when a smaller CPF (<0.2 cm) is suspected, TEE has greater sensitivity (76.6%) than TTE (61.9%).³ Echocardiography usually demonstrates a small, mobile, pedunculated or sessile valvular or endocardial mass, which on many occasions flutters or prolapses into the cardiac chambers during systole or diastole. They may appear speckled with echolucencies and a stippled pattern near the edges, which correlates with the papillary projections on the surface of the tumour.⁴ Multiple case reports have detailed using 3D echocardiography for identifying papillary fibroelastomas.^{14–23} RT-3DTEE has been particularly useful in identifying the attachment point of the tumour, its relation to surrounding structures, discerning whether resection would impede valve function and evaluating the size and mobility of the tumour with regard to risk of embolisation.^{24 25} When compared with 2D echocardiography, 3D echocardiography has been shown to be better at estimating the diameter of intracardiac masses.²⁶ Given the minimal amount of additional time and risk required to obtain RT-3DTEE images, it has been recommended that 3D imaging be used in therapeutic decision-making, particularly choosing optimal surgical access prior to the removal of intracardiac masses.²⁷

MRI has been considered an inferior modality for imaging suspected cases of CPF because they are often too small and are attached to moving objects resulting in motion artefacts.²⁸ However, they can on occasion be seen on MRI and can in fact help in diagnosing cases of atypical locations such as ours.²⁹ A

few such cases identified by MRI have been reported in literature.^{7 30–33} On cardiac MRI, CPF appear as masses attached to the affected valve leaflet or endocardial surface and display differential enhancement with gadolinium contrast. MRI can also demonstrate turbulent blood flow caused by a CPF.⁴

There are currently no guidelines for the treatment of CPF. Inpatients who have embolic events such as our patient, surgery is strongly recommended.^{3 4} After surgical resection, tumour recurrences have not been reported. However, in asymptomatic patients in whom the diagnosis is made incidentally, opinion is varied with regard to therapy. Gowda *et al* had noted that tumour mobility was the only independent predictor of CPF-related death or nonfatal embolisation and as such recommended that asymptomatic patients should be treated if the tumour is mobile.⁴ In addition, Sun *et al* recommended that patients undergoing cardiac surgery for other lesions should also be considered for surgical resection.³ However, other authors have suggested that surgery should be performed even in patients who are asymptomatic with non-mobile tumours citing the high risk of embolic events.^{34–38} Lifelong anticoagulation can be offered to patients who are symptomatic, but are poor surgical candidates.

In conclusion, although CPF are benign tumours, the consequences of systemic embolisation are severe ranging from strokes to sudden cardiac death from coronary occlusion. While the majority of cases are located on heart valve surfaces and involve the left side of the heart, it is important to keep in mind that they can, on rare occasions, present in atypical locations such as our patient. Our case highlights the value and importance of using multimodality imaging in the accurate diagnosis and patient management when CPF are suspected.

Learning points

- Cardiac papillary fibroelastomas (CPF) are exceedingly rare tumours that occur on valvular surfaces of the heart.
- Most cases are diagnosed through transthoracic echocardiography and require surgical resection.
- CPF can arise from non-valvular surfaces such as the aortic wall.
- In cases presenting with an atypical location, multimodality imaging can be used for diagnoses and management.

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REFERENCES

- 1 Klarich KW, Enriquez-Sarano M, Gura GM, *et al*. Papillary fibroelastoma: echocardiographic characteristics for diagnosis and pathologic correlation. *J Am Coll Cardiol* 1997;30:784–90.
- 2 Burke A. Tumors of the heart and great vessels. *Atlas of tumor pathology* 1996;16:171–9.
- 3 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.
- 4 Gowda RM, Khan IA, Nair CK, *et al*. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. *Am Heart J* 2003;146:404–10.
- 5 Burri H, Vuille C, Sierra J. Papillary fibroelastoma as a cause of cardioembolic stroke. *Heart* 2002;88:216–a–216.
- 6 Czekajski-Chehab E, Tomaszewski A, Wójcik M, *et al*. Papillary fibroelastoma as an accidental finding in a multislice computed tomography of coronary arteries. *Can J Cardiol* 2006;22:1155–7.
- 7 Lembcke A, Meyer R, Kivelitz D, *et al*. Images in cardiovascular medicine. Papillary fibroelastoma of the aortic valve: appearance in 64-slice spiral computed tomography, magnetic resonance imaging, and echocardiography. *Circulation* 2007;115:e3–e6.
- 8 Said SM, Ehlers HC, Hauptmann S, *et al*. Papillary fibroelastoma of the aorta. *Arch Cardiovasc Dis* 2008;101:805–6.
- 9 Yerebakan C, Liebold A, Steinhoff G, *et al*. Papillary fibroelastoma of the aortic wall with partial occlusion of the right coronary ostium. *Ann Thorac Surg* 2009;87:1953–4.
- 10 Jha NK, Khouri M, Murphy DM, *et al*. Papillary fibroelastoma of the aortic valve—a case report and literature review. *J Cardiothorac Surg* 2010;5:84.
- 11 Kim AY, Kim JS, Yoon Y, *et al*. Multidetector computed tomography findings of a papillary fibroelastoma of the aortic valve: a case report. *J Korean Med Sci* 2010;25:809–12.
- 12 Eftekhari H, Islam A, Slawsky M. Aortic valve fibroelastoma presenting with myocardial infarction. *Catheter Cardiovasc Interv* 2011;77:716–9.
- 13 González-Santos JM, Arnáiz-García ME, Vargas-Fajardo MC, *et al*. Aortic wall papillary fibroelastoma. *J Thorac Cardiovasc Surg* 2013;146:e1–e3.
- 14 Singh A, Miller AP, Nanda NC, *et al*. Papillary fibroelastoma of the pulmonary valve: assessment by live/real time three-dimensional transthoracic echocardiography. *Echocardiography* 2006;23:880–3.
- 15 Butz T, Meissner A, Plehn G, *et al*. Papillary fibroelastoma prolapsing into the left ventricular outflow tract: diagnosis using three-dimensional TEE. *Herz* 2010;35:503–5.
- 16 Ahlgren B, Dorosz J, Rohrer A, *et al*. Real time three-dimensional transesophageal echocardiography in the evaluation of two cases of rare mitral valve tumors. *Echocardiography* 2012;29:1011–5.
- 17 Bonnicksen CR, Burkhardt HM, Klarich KW, *et al*. Surgical Resection of Mitral Valve Papillary Fibroelastoma: A Robot-Assisted, Minimally Invasive Approach with Three-Dimensional Transesophageal Echocardiography Imaging. *World J Cardiovasc Surg* 2012;02:10–13.
- 18 Arifi AA, Mohamed AA, Najm HK. Incidental finding of cardiac papillary fibroelastoma of the left ventricular outflow tract, associated with rheumatic mitral and tricuspid stenosis. *European Heart Journal Supplements* 2014;16(suppl B):B49–B51.
- 19 Lin H, Zheng J, O'Beirne P, *et al*. Papillary fibroelastomas of the aortic and tricuspid valves mimicking vegetations in a patient with turner syndrome. *Can J Cardiol* 2014;30:1250.e5–1250.e7.
- 20 Vaskelyte L, Risteski P, Bertog S, *et al*. A case of a fibroelastoma and patent foramen ovale in a patient with prior stroke. *Cardiovasc Diagn Ther* 2014;4:47–9.
- 21 Jain R, Bajwa T, O'Hair D, *et al*. Three-dimensional echocardiography guidance in case of papillary fibroelastoma complicating transaortic valve implantation. *Eur Heart J Cardiovasc Imaging* 2015;16:570.
- 22 Moya-Mur JL, Silva-Guisasaola J, Casas-Rojo E, *et al*. Aortic fibroelastoma: detailed preoperative analysis with 3D echocardiography. *Echocardiography* 2015;32:1213–4.
- 23 Rohani A, Bigdeli L, Nezafati M, *et al*. Three-dimensional echocardiography of a tricuspid valve papillary fibroelastoma. *J Saudi Heart Assoc* 2017;29.
- 24 Zaragoza-Macias E, Chen MA, Gill EA. Real time three-dimensional echocardiography evaluation of intracardiac masses. *Echocardiography* 2012;29:207–19.
- 25 Le Tourneau T, Pouwels S, Gal B, *et al*. Assessment of papillary fibroelastomas with live three-dimensional transthoracic echocardiography. *Echocardiography* 2008;25:489–95.
- 26 Asch FM, Bieganski SP, Panza JA, *et al*. Real-time 3-dimensional echocardiography evaluation of intracardiac masses. *Echocardiography* 2006;23:218–24.
- 27 Müller S, Feuchtnr G, Bonatti J, *et al*. Value of transesophageal 3D echocardiography as an adjunct to conventional 2D imaging in preoperative evaluation of cardiac masses. *Echocardiography* 2008;25:624–31.
- 28 Araoz PA, Mulvagh SL, Tazelaar HD, *et al*. CT and MR imaging of benign primary cardiac neoplasms with echocardiographic correlation. *Radiographics* 2000;20:1303–19.
- 29 Luna A, Ribes R, Caro P, *et al*. Evaluation of cardiac tumors with magnetic resonance imaging. *Eur Radiol* 2005;15:1446–55.
- 30 al-Mohammad A, Pambakian H, Young C. Fibroelastoma: case report and review of the literature. *Heart* 1998;79:301–4.
- 31 Wintersperger BJ, Becker CR, Gulbins H, *et al*. Tumors of the cardiac valves: imaging findings in magnetic resonance imaging, electron beam computed tomography, and echocardiography. *Eur Radiol* 2000;10:443–9.
- 32 Shiraishi J, Tagawa M, Yamada T, *et al*. Papillary fibroelastoma of the aortic valve: evaluation with transesophageal echocardiography and magnetic resonance imaging. *Jpn Heart J* 2003;44:799–803.
- 33 Kurian K, Edwards FH, Jacob B, *et al*. Papillary fibroelastoma presenting as left ventricular mass: Current update and literature review. *Tex Heart Inst J* 2006;33:63–5.
- 34 Colucci V, Alberti A, Bonacina E, *et al*. Papillary fibroelastoma of the mitral valve. A rare cause of embolic events. *Tex Heart Inst J* 1995;22:327.
- 35 Giannesi C, Kubis N, N'Guyen A, *et al*. Cardiac papillary fibroelastoma: a rare cause of ischemic stroke in the young. *Cerebrovasc Dis* 1999;9:45–9.

- 36 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.
- 37 Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. *Ann Thorac Surg* 1995;59:538–41.
- 38 Ni Y, von Segesser LK, Dirsch O, *et al.* Cardiac papillary fibroelastoma. *Thorac Cardiovasc Surg* 1996;44:257–60.

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