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IMAGE

Multiple Cardiac Papillary Fibroelastomas — Are They Really Rare or Underdiagnosed?

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Introduction

Cardiac papillary fibroelastoma (PFE) is a rare tumour involving mainly the valvular surfaces of the heart [1,2]. Even more rare is multifocal localisation of these tumours as well as their residence inside the cardiac chambers [3,6]. Herein, we report the case report of a 67-year-old woman with multiple PFEs of which only three were diagnosed preoperatively. Eight other tumours were discovered intraoperatively on the mitral valve chordal apparatus. This case highlights the importance of performing attentive inspection of cardiac chambers during the excision of fibroelastomas, as a failure to address multiple lesions—although their existence is rare—might expose the patient to dangers of future embolisation or reoperation.

Case Presentation

A 67-year-old asymptomatic woman with a history of non-obstructive asymmetric hypertrophic cardiomyopathy was referred to our centre because of the increasing size of multiple heart tumours detected on sequential echocardiograms. The patient was also known for paroxysmal atrial fibrillation and had benefited from the placement of a defibrillator after ablation of atrioventricular node. Finally, she was on tri-therapy for human immunodeficiency virus infection.

During echocardiographic follow-up, the patient had developed several cardiac tumours. On the latest transthoracic echocardiogram (TTE) three tumours were identified: a large one, developed on the left coronary aortic valve leaflet (13 × 18 mm), a smaller one, on the non-coronary leaflet (13 × 6 mm), and a third one, attached to the anterior subvalvular apparatus of the mitral valve (11 × 5 mm) (Figure 1, Video 1, 2). The patient had no clinical events suggestive of systemic embolisation and cerebral magnetic resonance imaging was normal.

Although asymptomatic, the patient was scheduled for surgery on the basis of the size of the aortic valve tumours (>10 mm). Preoperative transoesophageal echocardiogram (TOE) confirmed three cardiac tumours. After transverse aortotomy, we found a large gelatinous tumour filling the left coronary sigmoid valve leaflet and a smaller tumour attached to the non-coronary sigmoid leaflet. The right coronary sigmoid leaflet was free from lesions. After resection of the native aortic valve, we inspected the left ventricular outflow tract and the mitral subvalvular apparatus through the aortic orifice. We found eight additional tumours of small size (2–3 mm in diameter) adhering to the mitral valve chordae (7) or the endocardium of the left ventricular outflow tract (1) (Figure 2). The native aortic valve was then replaced

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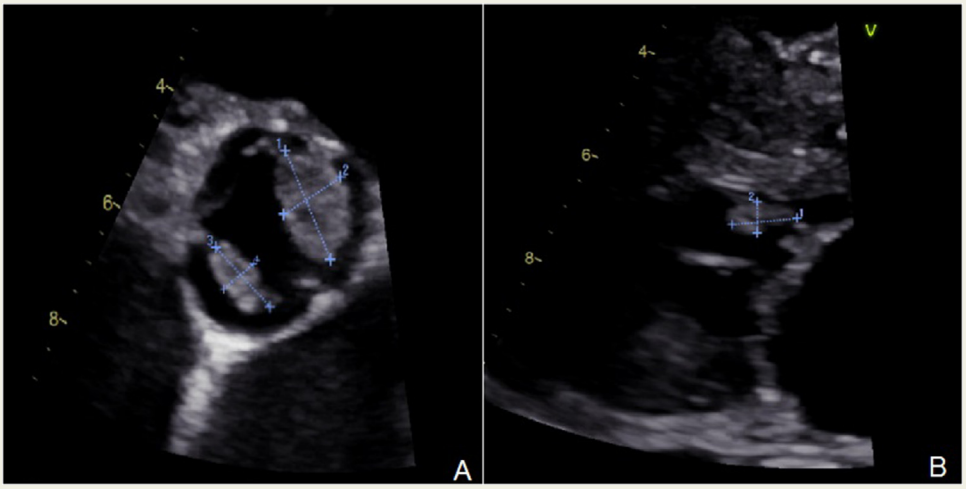

Figure 1 Video 1 A: Transoesophageal echocardiography (short-axis views) prior to surgery showing presence of large tumour on the left coronary sigmoid of the aortic valve and a smaller one, on the non-coronary leaflet. B: Same transoesophageal echocardiographic views (Video 2) showing density at the level of the anterior subvalvular apparatus of the mitral valve.

using a stented pericardial bioprosthesis (St Jude, Trifecta, size 23, St. Jude Medical, Inc., St Paul, MN, USA). The immediate postoperative course was uneventful.

Histopathologic examination (Figure 3) of the excised masses showed avascular, endothelial-lined fronds arising from central stalks, diagnostic of PFE. The patient had no evidence of recurrence of PFEs at her 1-month follow-up visit.

Discussion

Papillary fibroelastomas most commonly occur on cardiac valves (77%), although they may be found on any endocardial surface and are likely underdiagnosed. Mean age at detection is 60 years, although PFE can occur in all age groups.

Multiple PFEs are uncommon [1] and are reported to represent approximately 7% of all PFE cases [2]. However, Tamin et al. described in their study, 21% of cases with multiple PFE (range, 2 to 40) [3]. Our patient was diagnosed preoperatively to have only three tumours, but careful surgical exploration allowed retrieval of eight additional tumours.

Most cases of PFE are probably acquired, but the precise aetiology remains elusive. Grandmougin et al. report the

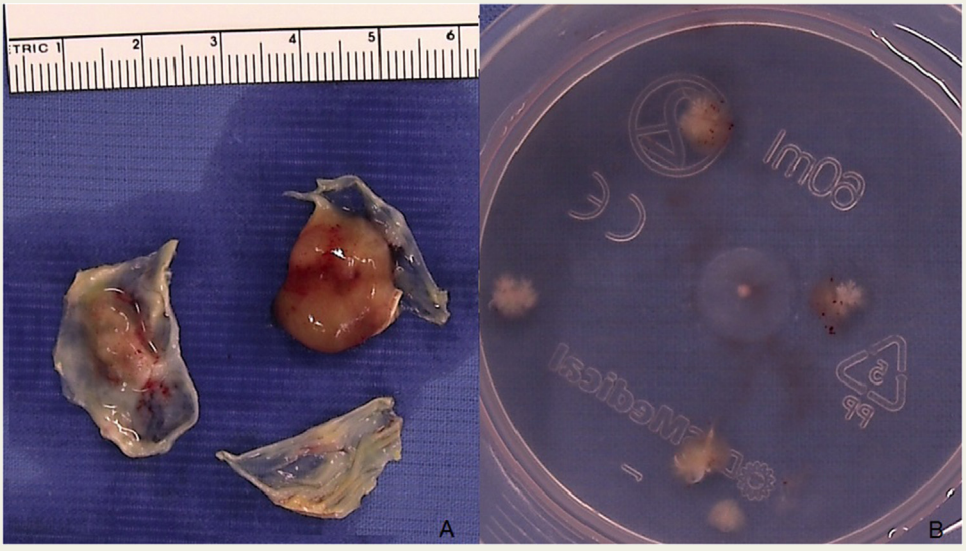

Figure 2 Intraoperative images. A. We found a large gelatinous tumour filling the left coronary sigmoid valve leaflet and a smaller tumour attached to the non-coronary sigmoid leaflet. The right coronary sigmoid leaflet was free from lesions. B. Multiple small tumours found in the subvalvular apparatus of the mitral valve.

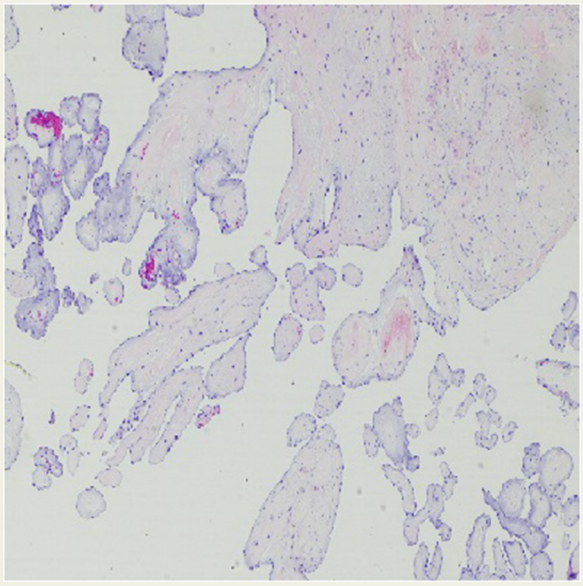

Figure 3 Histologic results of fibroelastoma exhibiting numerous avascular, endocardial-lined fronds from masses resected (H&E x 4).

presence of dendritic cells and remnants of cytomegalovirus in the intermediate layers of the tumour, suggesting a virus induced aetiology [4]. Moreover, Tamin et al. mention the presence of PFE in immunocompromised patients. Our patient was under tri-therapy for HIV infection, which might have contributed to the extensive and multifocal development of PFEs.

Surgical treatment is indicated in cases of systemic embolisation or tumour induced valve dysfunction or in case of large tumours (> 10 mm) [5–7]. In our case, the progressive increase in size of the tumours with a maximal diameter >10 mm prompted surgical intervention. In most of the reported cases, the native valve could be preserved. However, considering the large tumour sizes and the involvement of two aortic valve leaflets, we preferred to replace the valve to ensure complete tumour removal. This attitude is in line with the recent review of Arikan et al.

Conclusion

Cardiac PFE are very rare tumours, and their presence is often found by coincidence in patients followed by echocardiography for an another reason or after a neurologic event. Furthermore, emphasis must be placed on the importance of performing accurate preoperative and intraoperative exploration, as a failure to address multiple lesions—although their existence is rare—might expose the patient to dangers of future embolisation or reoperation.

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Competing interests

The authors declare that they have no competing interests.

Author Contributions

All the physicians were directly in charge of the patient throughout hospitalisation and follow-up. A.N and M.K performed surgical aortic valve replacement and tumors resection. PM performed echocardiography. AN prepared the manuscript draft, which was critically revised by M.K and approved by all authors.

Consent

Informed consent was obtained from this patient for publication of this case history and associated images in line with COPE recommendations.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.hlc.2018.08.020.

References

[1] Hattori R, Oishi C, Iwasaka J, Iwasaka T, Okada T, Johno H, et al. Multiple papillary fibroelastoma with quadricuspid aortic valve. J Thorac Cardiovasc Surg 2009;137:1280–2.
[2] Neuman Y, Luthringer DJ, Kobal S, Miyamoto T, Trento A, Siegel RJ. Multiple aortic valve papillary fibroelastoma: an unusual presentation of a rare tumor. J Am Soc Echocardiogr 2003;16:494–6.
[3] Tamin S, Maleszewski J, Scott C, Edwards W, Bruce C, Oh J, et al. Prognostic and bioepidemiologic implications of papillary fibroelastomas. J Am Coll Cardiol 2015;65:2420–9.
[4] Grandmougin D, Fayad G, Moukassa D, Docoene C, Abolmaali K, Ch Bodart J, et al. Cardiac valve papillary fibroelastomas: clinical, histological and immunohistochemical studies and a physiopathogenic hypothesis. J Heart Valve Dis 2000;9:832–941.
[5] Mathis S, Lamy M, Ciron J, Iljicsov A, Arjmand R, Agius P, et al. Paroxysmal sneezing at the onset of syncopes and transient ischemic attack revealing a papillary cardiac fibroelastoma. Case Rep Neurol Med 2014;2014:734849.
[6] Gowda RM, Khan IA, Nair CK, Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003;146(3):404–10.
[7] Arikan AA, Omay O, Aydın F, Kanko M, Gür S, Derviş E, et al. Aortic valve replacement for papillary fibroelastoma. J Card Surg 2017;32(June (6)):347–54.