pagepress

Finger ischemia in a young lady: an unusual presentation of papillary fibroelastoma with intraventricular location

Matteo Pernigo,[1] Elisabetta Dinatolo,[1] Marco Cirillo,[2] Zean Mhagna,[2] Alida Filippini,[3] Fabiana Cozza,[1] Marco Berti,[1] Roberto Bazzani,[3] Tony Sabatini,[3] Claudio Cuccia,[1] Giovanni Troise[2]

[1]Department of Cardiology, Poliambulanza Foundation Hospital, Brescia; [2]Department of Cardiac Surgery, Poliambulanza Foundation Hospital, Brescia; [3]Department of Internal Medicine, Poliambulanza Foundation Hospital, Brescia, Italy

Correspondence: Matteo Pernigo, Department of Cardiology, Poliambulanza Foundation Hospital, Via Bissolati 57, 25124 Brescia, Italy.
Tel.: +39 3383372351.
E-mail: matteo.pernigo@gmail.com

Key words: intracardiac mass, echocardiography, cardiac imaging, open heart surgery.

Contributions: all authors took part in the management of the case and approved the manuscript; MP, drafted the paper; ED, MB, FC, performed and reported the echocardiographic and imaging studies; AF, RB, TS, CC, aroused the suspicion of embolic lesion and took part in the clinical management of the patient; MC, ZM, GT, provided the surgical treatment.

Conflicts of interest: the authors declare no potential conflict of interest.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient included in this study.

Patient consent for publication: the patient gave her informed consent to the publication of material regarding the case. All data in the manuscript and on the figures are anonymized.

Funding: none.

Availability of data and materials: data are available from the corresponding author.

Received: 24 April 2023.
Accepted: 18 May 2023.
Early view: 24 May 2023.

Licensee PAGEPress, Italy
Monaldi Archives for Chest Disease 2024; 94:2615
doi: 10.4081/monaldi.2023.2615

Abstract

An otherwise healthy 32-year-old woman suffered from finger ischemia. An echocardiogram and computed tomography scan revealed a mobile mass in the left ventricle that was attached to the anterior papillary muscle and did not involve the valve leaflets. The tumor was resected, and histopathology confirmed it to be a papillary fibroelastoma. Our case emphasizes the significance of a comprehensive diagnostic work-up for a peripheral ischemic lesion. This resulted in the discovery of an unusual intra-ventricular origin for a commonly benign tumor.

Introduction

Papillary fibroelastomas (PFE) are the most common benign primary tumors of the heart (11.5%) after myxomas and lipomas. They are usually located on heart valves, most commonly the aortic valve. They originate from the endocardium; however, they can be rarely discovered in different cardiac structures [1]. Most cases are limited in dimension and asymptomatic; they are usually discovered incidentally, commonly in men in the 5th decade of life [2]. We present a rare case of fibroelastoma with an atypical intraventricular location, diagnosed in a young lady who suffered from finger ischemia due to peripheral embolization from the tumor.

Case Report

In December 2022, a 32-year-old woman presented to the emergency department of our institution for pain and acrocyanosis of the 3rd and 4th fingers of the right hand, which appeared abruptly 10 hours earlier. She experienced shoulder pain the previous day, lasting for a few hours. Her previous medical history was unremarkable. She did not take any medication, and she was a light smoker. The physical examination was otherwise normal; she was apyretic. Blood samples, including blood count, platelets, coagulation tests, liver and kidney function, and inflammatory markers, were normal. Pain and cyanosis of the fingers showed progressive improvement within 6 hours of observation. The Raynaud phenomenon was suspected, and the patient was initially discharged. The rheumatologist visited the patient the day after; the pain had disappeared and the skin color of the fingers had turned a slight red. The specialist prescribed further blood tests: anti-nuclear factor and anti-neutrophil cytoplasmic antibodies, anti-b-2-glycoprotein antibodies, and measurement of complement C3 and C4 factors, which resulted in normal results. The lesion was judged atypical for Raynaud, and an echocardio-

gram was requested to rule out significant cardiovascular disease. The exam showed normal left and right ventricular dimensions, systolic function, and the absence of significant valvular disease. A highly mobile, iso-echoic mass was noted inside the left ventricle, with a major diameter of 8 mm; it was attached through a short stalk to the anterolateral papillary muscle, not involving the mitral valve leaflets, and it had a frond-like appearance. Findings were consistent with a PFE (Figures 1 and 2; *Supplementary Videos 1-3*).

Doppler examination excluded pathology of the ascending aorta and right subclavian artery and demonstrated patency of the arteries of the right arm and right hand. The case was compatible with peripheral embolization of the cardiac mass. The patient was admitted to our department for further work-up. Thrombophilia screening was negative, as well as blood cultures. A cardiac computed tomography scan confirmed the presence of a mass, suggestive of fibroelastoma, on the anterolateral papillary muscle without valvular involvement (Figure 3); the exam ruled out significant coronary disease. The patient underwent cardiac surgery 2 days later. With a median sternotomy and cardiopulmonary bypass, the left ventricle was reached through an aortotomy. A multilobular yellowish mass was seen attached to the anterolateral papillary muscle and was completely resected (Figure 3); chordae tendineae were preserved. Macroscopic examination (Figure 4) and histological findings confirmed the diagnosis of PFE. The postoperative course was unremarkable.

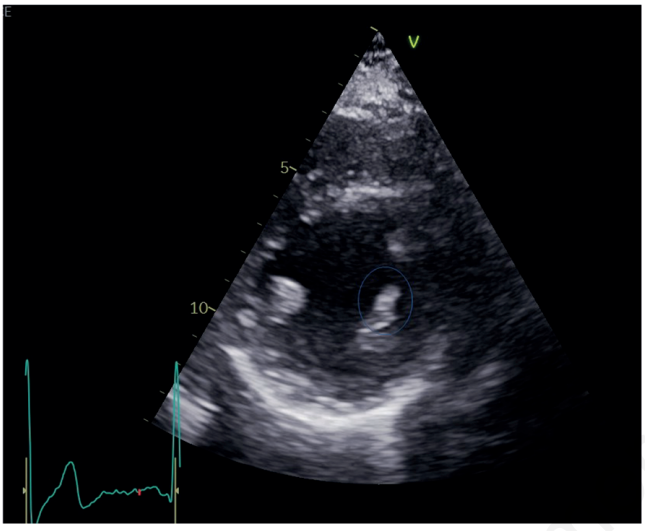

Figure 1. Transthoracic echo short axis view showing the papillary fibroelastoma on the anterior-lateral papillary muscle.

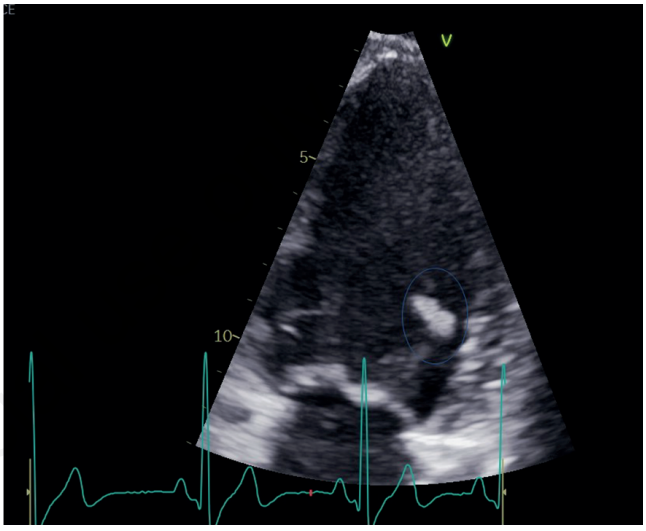

Figure 2. The papillary fibroelastoma shown in apical 2-chamber view; the mass did not involve mitral leaflets.

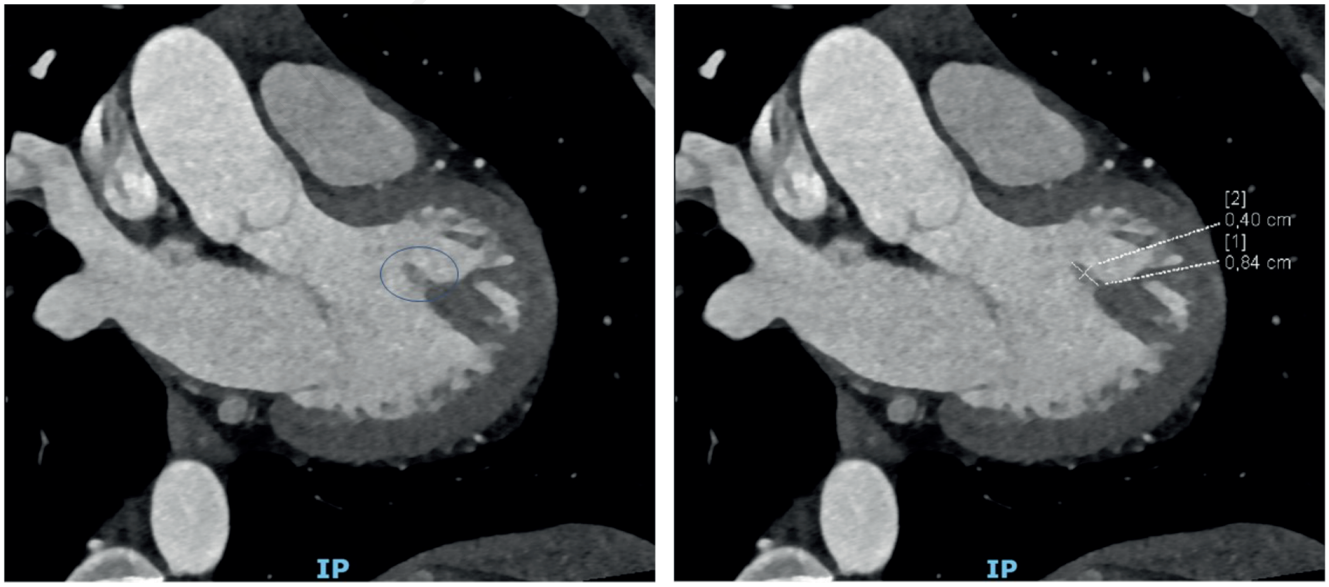

Figure 3. Image from cardiac computed tomography scan showing the mass attached to the anterolateral papillary muscle (left, circle), measuring 0.8×0.4 cm (right).

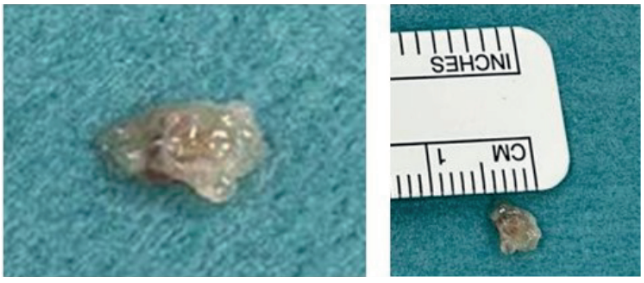

Figure 4. Macroscopic aspect of the papillary fibroelastoma just after surgical resection.

Discussion

Our case has some peculiar aspects. The clinical presentation was very atypical for a PFE; these benign tumors are generally asymptomatic. According to the literature, however, life-threatening complications caused by large or multiple tumors involving the left heart may rarely occur [3]. Angina, acute myocardial infarction, or sudden death, caused by a tumor occluding the coronary ostium or by embolization, may be the presenting symptom [4]. Cerebral embolization, either of fibrin or a tumor fragment, has also been reported; this may present as a transient ischemic attack, visual disturbance, or ischemic stroke [5]. Pulmonary embolization from right-sided PFE may cause respiratory distress [6,7]. To our knowledge, this is the first description in the literature of finger ischemia due to a PFE; it was reasonably caused by the embolization of a tumor fragment.

The diagnostic work-up was complex; the patient did not present a heart murmur, signs of bacterial infection, or fever suggestive of endocarditis, so an echocardiogram was scheduled only after ruling out a common cause of acrocyanosis as the Raynaud phenomenon. Once the intra-cardiac mass was discovered, Doppler examination excluded concomitant pathology of peripheral arteries, corroborating the hypothesis of an embolic lesion from the mass itself. The reported symptoms mark the migration of the embolus in the right arterial system up to the terminal arteries of the hand. PFE is usually found on valve surfaces; its intra-ventricular location is very rare [2]. Infrequent sites of involvement are the mitral chordae tendinae, right atrial endocardium, and endocardial surface of both ventricles, including papillary muscles and the interventricular septum [3]. In our patient, transthoracic echocardiography aroused suspicion of a PFE given the location of the mitral subvalvular apparatus and the frond-like appearance. The origin of PFE is not fully understood, but several mechanisms have been suggested: prior damage to the endothelium, hamartomatous origin, and organizing emboli [8]. The histologic features of the tumor consist of multiple papillary villous fronds radiating from a central fibrocollagenous stalk, with each frond showing three zones: a central fibrocollagenous core; a peripheral myxomatous zone; and an outer rim of hyperplastic endothelial cells. This architecture explains the typical tumor echocardiographic aspect, as found in our case: a pedunculated finger-like mobile excrescence attached to the endocardium ('sea anemone'-like appearance). The architecture of papillary fronds distinguishes PFE from cardiac myxoma at the histological examination and imaging modalities [6,8]. Other common differential diagnoses are vegetation, excluded in our case by negative inflammatory markers and blood cultures; thrombus, excluded by atypical location and negative thrombophilia tests. Cardiac computed tomography can provide further information about the exact mass location, extension, and involvement of cardiac structures. Cardiac magnetic resonance imaging is the reference standard technique to examine a suspected cardiac tumor, providing a non-invasive tissue characterization based on a multi-parametric assessment of the chemical micro-environment of the mass [9]. Since diagnosis was established in our case with other modalities, this exam was not performed on our patient as it was not deemed necessary, and we risked postponing cardiac surgery.

Surgical excision is the treatment of choice for PFE at high embolic risk [10]. Complete surgical resection is generally safe and effective since the risk of recurrence is very low [10]. Once the diagnosis was established, our patient was promptly referred for surgery, preventing further embolic events with potential major complications.

Conclusions

Our case highlights the importance of a complete diagnostic work-up of a peripheral ischemic lesion, taking into account the hypothesis of cardiac embolism even in patients without known risk factors. PFE, generally regarded as a benign common tumor, may have an atypical intra-cardiac location, leading to potentially life-threatening complications.

References

1. Tyebally S, Chen D, Bhattacharyya S, et al. Cardiac tumors: JACC CardioOncology state-of-the-art review. JACC CardioOncol 2020;2:293-311.
2. Gowda RM, Khan IA, Nair CK, et al. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003;146:404-10.
3. Sun JP, Asher CR, Yang X, et al. Clinical and echocardiographic characteristics of papillary fibroelastomas: a retrospective and prospective study in 162 patients. Circulation 2001;103:2687-93.
4. Meng F, Sun JP, Chen M, et al. An unusual cardiac fibroelastoma case. Circulation 2014;30:520-2.
5. Barbarossa A, Coraducci F, Cipolletta L, et al. Very mobile left ventricular outflow tract papillary fibroelastoma presenting with multiple ischemic strokes: a case report and brief review of the literature. J Cardiovasc Echogr 2022;32:222-4.
6. Kurian KC, 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.
7. Zhang M, Liu X, Song Z, et al Cardiac papillary fibroelastoma: a retrospect of four cases. J Cardiothorac Surg 2013; 8:65.
8. Klarich KW, Enriquez-Sarano M, Gura GM, et al. Papillary fibroelastoma: echocardio-graphic characteristics for diagnosis and pathologic correlation. J Am Coll Cardiol 1997;30: 784-90.
9. Hoey ETD, Shahid M, Ganeshan A, et al. MRI assessment of cardiac tumours: part 1, multiparametric imaging protocols

and spectrum of appearances of histologically benign lesions. Quant Imaging Med Surg 2014;4:478-88.
10. Yopp AC, Vaynblat M, Cunningham JN Jr, Lazzaro RS. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. J Card Surg 2007; 22:72-3.

Online supplementary material:

Video 1. Echocardiography clip, short axis view of the left ventricle at papillary muscles level, showing the frondlike mobile mass attached to the antero-lateral papillary muscle.

Video 2 and Video 3. Echocardiographic modified apical 3-chamber (2) and apical 2-chamber (3) clips, focused on the mass, showing its attachment and highly mobile appearance.