Papillary fibroelastoma originating from the left ventricular outflow tract

Irem Turkmen[a], Serkan Kahraman[a], Ersin Kadirogullari[b], Salih Guler[b], Lokman Yalcin[b], Nermin Gunduz[c], Gamze Babur Guler[a]

[a] *University of Health Sciences Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey*
[b] *Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center, University of Health Sciences, Department of Cardiovascular Surgery, Training and Research Hospital, Istanbul, Turkey*
[c] *Department of Pathology, University of Health Sciences Kanuni Sultan Süleyman Training and Research Hospital, Istanbul, Turkey*

ARTICLE INFO

Article history:
Submitted: 16. 1. 2023
Accepted: 8. 5. 2023
Available online: 9. 10. 2023

Klíčová slova:
Embolie
Papilární fibroelastom
Výtokový trakt levé komory

Keywords:
Embolism
Left ventricular outflow tract
Papillary fibroelastoma

SOUHRN

Papilární fibroelastomy jsou vzácné benigní srdeční nádory. V této kazuistice popisujeme případ asymptomatické 64leté ženy s velkým papilárním fibroelastomem ve výtokovém traktu levé komory, který byl odstraněn chirurgicky. Naším cílem je zdůraznit řešení atypického a klinicky asymptomatického papilárního fibroelastomu.

ABSTRACT

Papillary fibroelastomas are rare benign tumors of cardiac origin. In this case report, we present an asymptomatic 64-year-old female patient with a large left ventricular outflow tract papillary fibroelastoma treated surgically. We would like to emphasize the management of atypical located and clinically asymptomatic papillary fibroelastoma.

Introduction

Primary cardiac tumors are usually benign, relatively uncommon masses compared to metastatic tumors of heart.[1] Papillary fibroelastomas, which are benign avascular endocardial papillomas, are one of the most common benign cardiac tumors that arise predominantly from the cardiac valves.[2] Despite predominance of male gender, there are no other significant risk factors or pathophysiologic mechanism proven.[1] Clinical manifestations of tumors depend on their size and location such as originating from the endocardium, heart valves, or myocardium.[2] While the majority of patients is asymptomatic on presentation, the clinical presentation of papillary fibroelastoma varies from asymptomatic to sudden cardiac death because of severe embolic complications such as stroke or myocardial infarction.[3] In this present case, we describe an asymptomatic 64-year-female patient with a big left ventricular outflow tract fibroelastoma treated successfully with surgical approach.

Case report

A 64-year-female patient presented with progressive dyspnea. The patient had a history of hypertension and rheumatoid arthritis. She had no history of stroke or myocardial ischemia. There were no remarkable abnormalities on physical examination. The ECG showed normal sinus rhythm at admission and the laboratory investigations were unremarkable. Two-dimensional transthoracic echocardiography demonstrated normal left ventricular systolic function. The left ventricular internal dimensions (LVED: 45 mm, LVES: 33 mm) were in normal range with an ejection fraction of 58% and the left atrial anteroposterior diameter was 37 mm. A transthoracic echocardiogram also revealed a mobile hypoechogenic mass measuring 15 × 11 mm attached to interventricular septum in the left ventricular outflow tract (LVOT). Then transesophageal echocardiography (TEE) was performed for further evaluation. TEE confirmed a hypoechoic, gelatinous, amorphous mobile mass measuring 1.68 × 0.825 cm was attached to

Address: Irem Turkmen, MD, University of Health Sciences Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istasyon, Turgut Ozal Bulvari No:11, 34303 Kuçukcekmece, Istanbul, Turkey, e-mail: iremturkmen1@gmail.com
DOI: 10.33678/cor.2023.042

Please cite this article as: Turkmen I, Kahraman S, Kadirogullari E, et al. Papillary fibroelastoma originating from the left ventricular outflow tract. Cor Vasa 2023;65:787–789.

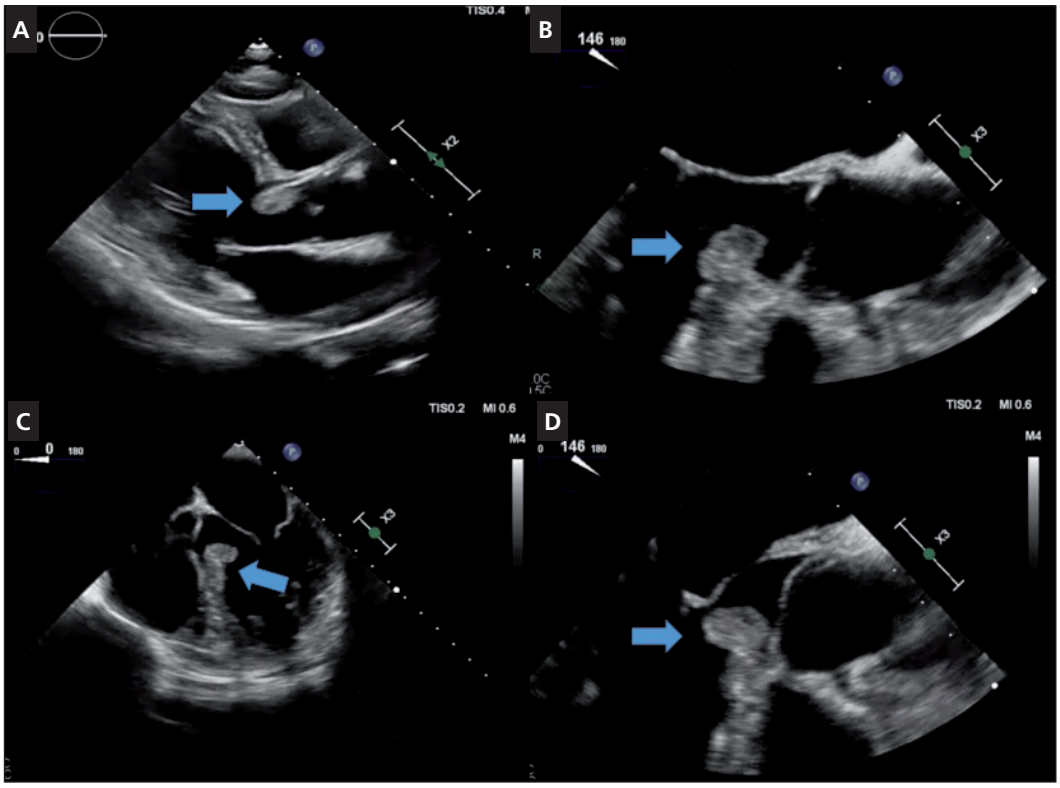

Fig. 1 – (A) Parasternal long axis view of the mass in LVOT (arrowhead). (B) Zoom LVOT view of the mass on TEE (arrowhead). (C) Apical four chamber mid septum view of the mass (arrowhead). (D) Zoom LVOT view of the mass on TEE (arrowhead). LVOT – left ventricular outflow tract; TEE – transesophageal echocardiography.

the septal wall of the LVOT at 7 mm proximal from aortic valve (Fig. 1, video 1). Aortic valve is intact and there was no aortic insufficiency. Based on all examinations, although fibroelastoma is preliminary diagnosis because of pedunculated, hypoechoic, and well-circumscribed TTE image of the mass, also myxoma should be in differential diagnosis. Cranial MRI was planned for the patient before the surgical evaluation and no evidence of any ischemic foci was reported. Although there was no cardioembolic event, heart team decided to perform surgery to prevent further complications. In histological analysis of surgical specimens; villous structures, lined with endothelial cells, containing hyalinized myxoid cores were observed. Positive staining was detected in endothelial cells lining villous structures with CD31 staining, and elastic fibrils in the center of myxoid cores with elastic Van Gieson (EVG). These features were characteristic for cardiac papillary fibroelastoma (Fig. 2). The patient was operated successfully by the surgical team and discharged on the 2nd postoperative day.

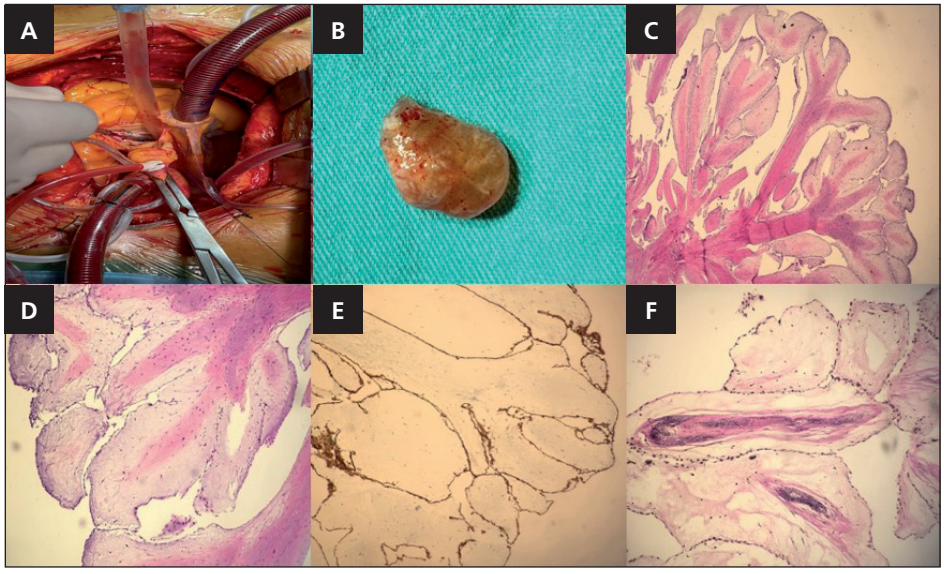

Fig. 2 – (A) Image of the surgical procedure. (B) Specimen of the LVOT mass. (C) Avascular branching papillary structures with central hyalinized (H&E, ×40). (D) Papillary structures with myxoid and hyalinized cores, lined with endothelial cells (H&E, ×100). (E) Positive staining of endothelial cells lining the papillary structures with CD31 (CD31, ×100). (F) Positive staining of papillary cores with elastic Van Gieson (EVG, ×100). LVOT – left ventricular outflow tract.

Discussion

Primary cardiac tumors are rare neoplasms with the prevalence of 0.021–0.019%. The majority of primary cardiac tumors is benign and papillary fibroelastomas are the most common primary cardiac tumors after cardiac myxomas.[1] Papillary fibroelastoma is attached especially to cardiac valves while the aortic valve is affected predominantly (29.4%). Although mitral valve attachment is seen relatively uncommon compared to aortic valve, the studies state that the concurrent valvular disease is represented in 41.7 % of patients.[2] Unlike valvular disease, nonvalvular fibroelastomas are mostly originated from left ventricle.[1] In addition to that, neoplasms occur sporadically and most of them are detected incidentally during routine echocardiographic examination.

Due to the asymptomatic nature of the tumors, their diagnosis was rare in the past. As the increased use of echocardiography in this area, this neoplasm became more noticeable.[2] Moreover, the location, occurrence, and the size of tumor attached to valve or endocardium are crucial to have longstanding valvular heart disease by affecting hemodynamics.[4] Besides asymptomatic presentation, they may result in life-threatening complications such as stroke, myocardial infarction, and systemic embolization. Additionally, most common clinical manifestations that are associated with embolization of tumor fragments are angina, myocardial infarction, sudden cardiac death. Stroke or transient TIA, which is seen as a neurological deficit, has a significant role in reported clinical presentations.[1] Furthermore, because of the predominance of the left heart localization, systemic embolization occurs and the majority of them comprises cerebral arteries.[3] Also, coronary arteries occlusion leads to angina or myocardial infarction and peripheral ischemia linked by mesenteric, renal infarction have been reported. In our case, a large (over 1 cm) nonvalvular papillary fibroelastoma located in LVOT was detected without causing any cardioembolic event. No silent brain ischemia was observed in the cranial MR examination. Surgical removal of the asymptomatic mass was taken as an expert opinion by the heart team due to the risk of coronary embolism and cerebrovascular event.

Despite its benign nature, it causes severe complications due to their location and pedunculated form. Therefore it should be surgically removed as surgical intervention is the definitive treatment.[5] Although cardiac imaging like transthoracic echocardiography and transesophageal echocardiography is crucial to differentiate the neoplasm, histological analysis is necessary for exact diagnosis. If the diameter of the mass exceeds 1 cm, patients should be offered surgical resection, and if the mass is left-sided and endocardial, regardless of its size and accidental nature, the necessity of surgery should be discussed with a multidisciplinary team, considering the risk of embolism.[6]

Conclusion

Cardiac papillary fibroelastoma is one of the rare neoplasms of the heart. Its clinical manifestations vary from asymptomatic to sudden cardiac death. In this case, we aimed to present the management of a large, asymptomatic, atypically located papillary fibroelastoma.

Contributorship

All of the authors contributed to planning, conducting, and reporting of the work. All authors had a full access to all data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Conflict of interest

The authors declare that there are no conflicts of interests.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical statement

Detailed written informed consent was obtained from the patient for the publication of this case report and images.

Data available statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Supplementary material

Supplementary material is available in the online version.

References

1. Lak HM, Kerndt CC, Unai S, Maroo A. Cardiac papillary fibroelastoma originating from the coumadin ridge and review of literature. BMJ Case Rep 2020;13:e235361.
2. Val-Bernal JF, Mayorga M, Garijo MF, et al. Cardiac papillary fibroelastoma: retrospective clinicopathologic study of 17 tumors with resection at a single institution and literature review. Pathol Res Pract 2013;209:208–214.
3. Gowda RM, Khan IA, Nair CK, et al. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J 2003;146:404–410.
4. Kesavan S, Dyall L, Nelson M, et al. Fibroelastomas of the left ventricular outflow tract. JRSM Short Rep 2011;2:3.
5. Coats CJ, Reid J, Wright S, et al. Papillary fibroelastoma arising from left ventricular outflow tract. Eur Heart J Cardiovasc Imaging 2018;19:826.
6. Lyon AR, López-Fernández T, Couch LS, Asteggiano R, et al. 2022 ESC Guidelines on cardio-oncology developed in collaboration with the European Hematology Association (EHA), the European Society for Therapeutic Radiology and Oncology (ESTRO) and the International Cardio-Oncology Society (IC-OS): Developed by the task force on cardio--oncology of the European Society of Cardiology (ESC). Eur Heart J 2022;43;4229–4361.
7. 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. Eur Heart J Suppl 2014(16):B49–B51.