STUDENTS' CORNER
LETTER TO THE EDITOR

Papillary Fibroelastoma: A significant lesion in cardiac medicine

Asra Hasan, Aresha Nehal, Umema Abu Turab

Dear Madam, Primary cardiac tumours are rare, with papillary fibroelastoma being the third most common tumour following atrial myxoma and lipoma. It is considered a benign neoplasm with an incidence of less than 0.1%, emanating from the valvular endocardium.[1] However, it is regarded as an asymptomatic tumour and is diagnosed incidentally via routine echocardiography or during autopsy. Despite being a silent tumour, it still holds the risk of severe future complications, which are associated with systemic emboli, stroke, myocardial infarction, and sudden death. Hence, early diagnosis is the principal step that allows prioritising the management options suggesting close monitoring or surgery.[2]

Due to the rarity of this disease, there is still insufficient data that could provide us with the associated etiological risk factors. It arises from the valvular endocardium in most cases with each valve of the heart having a varying percentage of involvement. The aortic valve accounts for 29% of cases, the mitral valve for 25%, the tricuspid valve for 17%, and the pulmonary valve for 13%.[3] Predominantly patients suffering from papillary fibroelastoma have no clinical features but it does have a few manifestations such as syncope, angina, and shortness of breath. The fragility of this tumour implies a probable risk of embolization which could ultimately result in transient ischaemic attacks and myocardial infarction. Additionally, some cases of congestive heart failure, thromboembolism, ventricular fibrillation, and sudden death have been reported. Embolization usually occurs due to the abrupt formation of thrombus frequently on the tumour nidus resulting from platelet aggregation. A highly mobile pedunculated mass attached to a valve or endocardium is a typical incidental finding of papillary fibroelastoma through an echocardiogram.[4] However, new techniques including 3-D echocardiography, magnetic resonance imaging, and multi-slice spiral computed tomography have also been reported to be beneficial for the diagnosis. The tumour almost always appears as a solo lesion yet can also emerge in multiple forms. Treatment strategies comprises of early anticoagulation therapy, and surgical correction implying tumour resection and replacement of an entire diseased valve.[5]

Papillary fibroelastoma is an infrequent diagnosis and prior detection is an appropriate tool to prevent its life-threatening complications. Implementation of proper screening methods for high-risk patients and the elderly is necessary. More research should be conducted on this rare tumour for favourable outcomes. Moreover, the latest and less invasive treatment modalities should be introduced for better disease control.

Disclaimer: None.

Conflict of interest: None.

Funding disclosure: None.

DOI: https://doi.org/10.47391/JPMA.10953

References

1. Devanabanda AR, Lee LS. Papillary Fibroelastoma. Treasure Island (FL): StatPearls Publishing, 2023.
2. Eslami-Varzaneh F, Brun EA, Sears-Rogan P. An unusual case of multiple papillary fibroelastoma, review of literature. Cardiovasc Pathol. 2003; 12:170-3. doi: 10.1016/s1054-8807(03)00033-4.
3. Grinda JM, Couetil JP, Chauvaud S, D'Attellis N, Berrebi A, Fabiani JN, et al. Cardiac valve papillary fibroelastoma: surgical excision for revealed or potential embolization. J Thorac Cardiovasc Surg. 1999; 117:106-10. doi: 10.1016/s0022-5223(99)70474-5.
4. Kaplan JG, Kanwal A, Bahoora J, Berquist J, Hunyadi V, Keirn R. Papillary fibroelastoma presenting with multi-organ symptoms. J Community Hosp Intern Med Perspect. 2020; 10:597-9. doi: 10.1080/20009666.2020.1811067.
5. Jha NK, Khouri M, Murphy DM, Salustri A, Khan JA, Saleh MA, et al. Papillary fibroelastoma of the aortic valve--a case report and literature review. J Cardiothorac Surg. 2010; 5:84. doi: 10.1186/1749-8090-5-84.

Final year MBBS Student, Dow University of Health Sciences.
Correspondence: Asra Hasan. e-mail: asrahasan4061@gmail.com
ORCID ID. 0009-0007-1269-3720

Submission complete: 20-09-2023 **Review began:** 11-12-2023
Acceptance: 18-04-2024 **Review end:** 20-03-2024

Author Contribution:
AH: Conception, literature search, writing and reviewing.
AN & UAT: Literature search writing and reviewing.