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CASE REPORT

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Papillary fibroelastoma presenting with multi-organ symptoms

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ABSTRACT

Papillary fibroelastomas are a rare cardiac neoplasm typically found on the left side of the heart, and most commonly on the aortic valve, which can present with cardiac or neurologic symptoms.

A 51-year-old woman with no cardiac history presented to a resident clinic with complaints of left-sided facial paresthesias and palpitations for 1 month. Echocardiographic imaging showed a mass on the aortic annulus, concerning for a cardiac tumor. Due to the risk of possible embolization, if the tumor was a myxoma, the patient required intrathoracic surgery. During the intrathoracic procedure the mass was confirmed to be a papillary fibroelastoma and the patient had the mass removed without any complications.

Papillary fibroelastomas are found in less than 1% of the population but can present clinically with a wide variety of symptoms. Patients with this neoplasm are at risk for severe complications, due to embolization, potentially causing cerebrovascular accidents or myocardial infarctions. We present a case of a papillary fibroelastoma producing both cardiac and neurologic symptoms.

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KEYWORDS
Papillary fibroelastoma; cardiac neoplasm; transient ischemic attack

1. Introduction

Papillary fibroelastomas are the second most common benign primary cardiac neoplasms. The majority are asymptomatic, however, they can produce cardiac or neurologic symptoms and even ischemic events. We present a case of a papillary fibroelastoma producing both cardiac and neurologic symptoms in an established patient.

Case:

A 51-year-old woman with no cardiac history presented to a resident clinic with new complaints of palpitations and left-sided facial paresthesias for the prior month. Thus far, MRI of the brain did now show any abnormalities. Due to the patient's history of anxiety, the patient's symptoms were initially considered to be psychogenic. Once anxiolytic therapy failed to relieve the patient's symptoms, further evaluation was pursued.

A transthoracic echocardiogram (TTE) showed an ejection fraction of greater than 75%, normal left ventricular systolic function and no obvious regional wall motion abnormalities. The TTE also revealed an echodensity which appeared to be attached to the interventricular septum. A transesophageal echocardiogram (TEE) was pursued for further evaluation of the echodensity and demonstrated a highly mobile, well circumcised mass measuring 8 mm x 8 mm within the mid-left ventricular outflow tract on the anterior surface approximately 0.8 cm from the aortic annulus, concerning for a cardiac tumor (Figures 1–3). There was no outflow track obstruction.

Due to the risk of recurrence, and the risk of embolization of a possible myxoma, intrathoracic surgery was pursued by cardiothoracic surgery. Pre-operatively, cardiac catheterization was completed with minimal, non-obstructive coronary artery disease.

The patient underwent the intrathoracic surgery and a pedunculated mass was noted in the right coronary sinus adhered to the muscular septum. The mass was extracted from the base and the endocardium was shaved to prevent recurrence. Pathology results, using immonstatin staining, confirmed the diagnosis of papillary fibroelastoma. The patient tolerated the procedure well with no complications.

On 2 week follow up, the patient noted complete resolution of her palpitations and indicated improvement in her left-sided paresthesias. The patient subsequently attended cardiac rehabilitation with no complications following her procedure. Her paresthesias and palpitations were thought to be a result of her papillary fibroelastoma.

2. Discussion

Papillary fibroelastomas are the second most common benign cardiac neoplasm behind only myxomas. Primary cardiac tumors are very rare with a prevalence ranging from 0.0017% to 0.28% [1]. Papillary fibroelastomas

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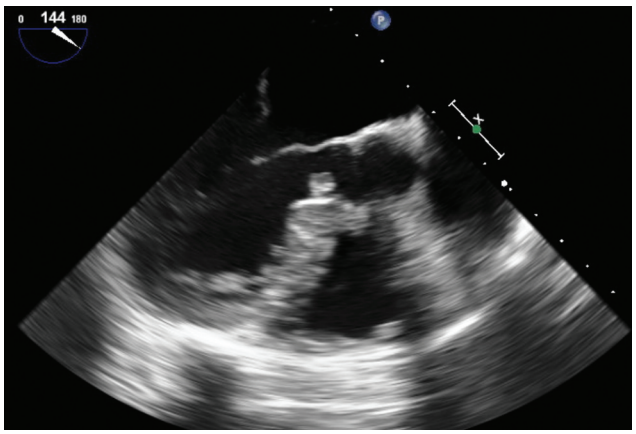

Figure 1. Mid-esophageal long-axis view of the aorta on TEE.

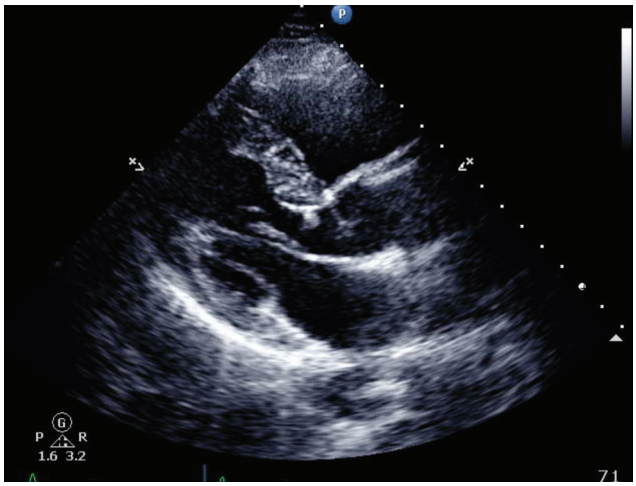

Figure 2. Transgastric long-axis view on TEE.

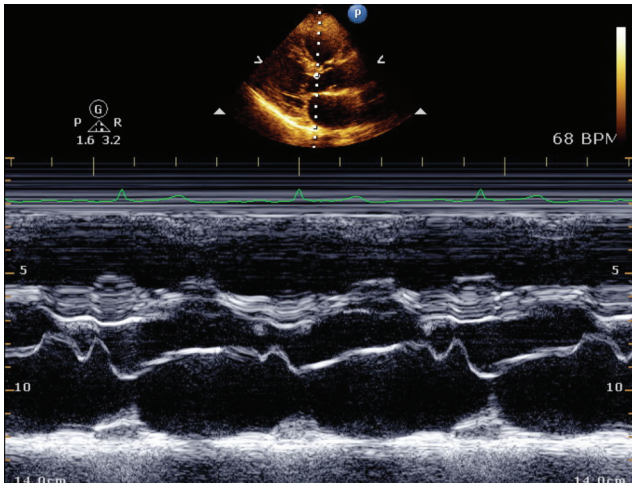

Figure 3. M mode imaging through the left ventricular outflow tract with demonstration of the mass.

account for 7% of all cardiac tumors and are typically found on the left side of the heart, commonly attached aortic valve leaflets [2]. Most patients who have this tumor are asymptomatic but patients can present with various neurologic symptoms if the tumor embolizes. There have also been documented cases where these tumors have embolized into the coronary vessels resulting in ischemic damage to the myocardium.

From a structural standpoint, papillary fibroelastomas appear similar to the chordae tendineae.

Papillary fibroelastomas are avascular in nature and composed of an outer endothelial layer and a dense central core composed of a rim of loose mucopolysaccharide-rich connective tissue, dendritic cells, fibroblasts and smooth muscle cells. The friability of this tissue matrix makes a papillary fibroelastoma a potential risk for embolization resulting in transient ischemic attacks, cerebrovascular accidents or even myocardial infarction [3].

Papillary fibroelastomas are typically diagnosed via echocardiogram appearing as a highly mobile-pedunculated mass attached to the endocardium or valve on echocardiogram. A transthoracic echocardiogram has a sensitivity of 88.9% and a specificity of 87.8% if the mass is greater than 2 mm. If the mass is smaller than 2 mm, the sensitivity is 61.9% and the specificity is 76.6% on transthoracic echocardiogram [4]. One study showed that roughly one-third of papillary fibroelastomas were only seen on transesophageal echocardiograms as opposed to TTE [4]. Transesophageal echocardiogram provides a more clear and detailed image of the masses size, origin and can give a better estimation of the masses potential obstructive nature. Traditionally, CT scan may not detect papillary fibroelastomas due to the fast-moving nature of the mass. However, with the new innovations of multi-slice scanners, CT scans have been able to pick up the quick-moving masses. A benefit of echocardiogram compared to other imaging modality, like a CAT scan, is the live motion aspect of the imaging. Other static images do not show the motion of the mass as in an echocardiogram.

Surgical excision is recommended in all patients with symptomatic papillary fibroelastomas. Currently, there are no clear guidelines for asymptomatic papillary fibroelastomas. One publication recommends following guidelines associated with infective endocarditis, stating that vegetations with a length greater than 10 mm are associated with new embolisms [1]. This publication recommends surgical excision for papillary fibroelastomas greater than 10 mm and for masses smaller than 10 mm serial echocardiographic evaluation rather than surgery [1]. One study followed more than 500 cases of presumed fibroelastoma spanning a 16 year period at the Mayo Clinic, in which patients were stratified based on treatment with surgery compared to conservative management. This study showed the surgical group's risk of stroke was 2% at year 1 and 8% percent at 5 years. In the group treated with conservative management the risk of stroke was 6% at 1 year and 13% at 5 years [5].

3. Conclusion

Although cardiac neoplasms are uncommon, papillary fibroelastomas should be considered for patients with cardiac symptoms or neurologic symptoms that can be attributed to a transient ischemic attack. In these patients, surgical removal of the mass is recommended for resolution of the symptoms and for prevention of future embolization. The guidelines are unclear on patients who are not experiencing symptoms. Further research needs to be conducted on the recurrence rates of these masses once surgical excision has been performed.

Disclosure statement

No potential conflict of interest was reported by the authors.

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