



Atypical presentation of a ventricular papillary fibroelastoma

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Papillary fibroelastoma is the most common valvular cardiac tumor.^[1] The most common non-valvular site these tumors present is the left ventricle,^[2] where they usually manifest as ischemic strokes or myocardial infarction.^[3] Although these tumors also have a risk of embolization into the peripheral system,^[2] reports of such events are exceedingly rare.^[4]

A 77 year old female presented to the hospital for concerns of a non-healing ulcer of the right great toe. Significant findings on physical exam included a grade 3/6 systolic ejection murmur on the 5th intercostal space at mid-clavicular level on the left, which was non-radiating and of blowing quality. Her right lower extremity was noted to have diminished pulses on the popliteal and dorsalis pedis arteries. She was noted to have a partial amputation of the right great toe along with a non-healing surgical wound. Due to concern for osteomyelitis and possible surgical intervention, the patient underwent preoperative cardiac evaluation. A transthoracic echocardiogram revealed an ejection fraction of 75% and a defined, medium-sized, mobile mass within the left ventricle, measuring 18 mm x 14 mm (Figure 1).

Further evaluation with a trans-esophageal echocardiogram confirmed the presence of a pedunculated mobile mass attached to the septal wall of the left ventricle. A diagnostic coronary angiogram revealed non-obstructive coronary disease. The patient underwent successful ventriculotomy and resection of the intra-cardiac tumor, which on gross specimen was noted as a gelatinous, blue-colored mass (Figure 2). Pathologic evaluation confirmed the diagnosis of papillary fibroelastoma. The patient underwent a lower extremity angiogram, which was significant for total occlusion of the anterior tibial, distal posterior tibial and proximal peroneal arteries. In the setting of peripheral arterial

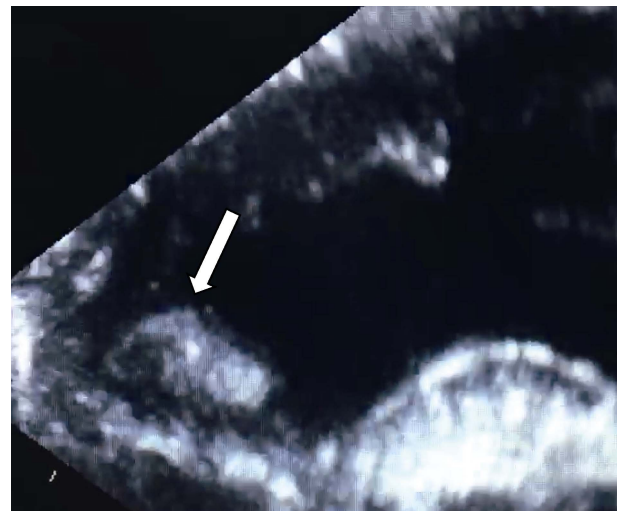


Figure 1. Still image of transthoracic echocardiogram depicting the papillary fibroelastoma (arrow) attached to the septal wall of the left ventricle.



Figure 2. Gross specimen depicting the resected papillary fibroelastoma. Note the gelatinous appearance of the specimen.

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occlusions and the finding of a papillary fibroelastoma, we concluded that her peripheral arterial occlusion was secondary to embolic thrombi originating from the intracardiac mass.

While cardiac myxomas are the most common primary cardiac tumors in all age groups, papillary fibroelastomas are the most common valvular tumors. The incidence rate depends on the valves being involved, with the aortic valve being the culprit in 44% of cases, followed by the mitral valve at 35%.^[3] While these tumors are well known to involve the cardiac valves, the incidence of left ventricular involvement is estimated to be 9%.^[3] Despite the fact that peripheral embolization is a potential consequence of papillary fibroelastomas, to date there is only one reported case of limb ischemia resulting from embolization of a papillary fibroelastoma.^[4] These tumors, due to their structure and propensity to form thrombi, are very friable and have a predilection for embolism. We postulate the notion that left ventricular papillary fibroelastomas, albeit rare in incidence, do in fact have a significant risk of causing peripheral artery disease and subsequent limb ischemia. Tumor mobility, as appreciated on echocardiogram, has been identified as an independent predictor of death and embolization in patients with papillary fibroelastomas.^[2] The management of these cardiac tumors remains controversial, with a current recommendation for surgical intervention on patients who are good surgical candidates, have asymptomatic left sided cardiac involvement, and those that are symptomatic or have experienced symptoms related to it.^[5] Due to symptomatic peripheral artery disease, surgical intervention was warranted for our patient.

4 Conclusions

Our case illustrates the importance of a complete cardiovascular evaluation in patients presenting with peripheral artery disease. Papillary fibroelastomas, albeit relatively rare in occurrence, are now more readily detectable due to advances in cardiac imaging. It is important to maintain a high index of suspicion for papillary fibroelastoma as an etiology of peripheral artery disease, as early identification and treatment not only provides improved quality of life for the patient, but also reduces morbidity and mortality associated with these tumors.

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