Atrial Papillary Fibroelastoma: A Stranger in a Strange Place

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ABSTRACT

Papillary fibroelastoma (PFE) is the most common primary tumor of cardiac valves and predominantly located on the left side. Its origin from non-valvular endocardium is extremely rare. We describe a case of an 81-year-old Caucasian male who presented with a mobile right atrial mass at the junction of right atrial wall and superior vena cava (SVC). Initially it was thought to be a thrombus and the patient was treated with anti-coagulation therapy without any change in size of the mass. Surgical excision was performed to establish the diagnosis and histopathology confirmed the diagnosis of PFE. In conclusion, this case is unique due to location of the tumor and its attachment with superior vena cava. Physicians should consider this unusual location of PFE in the differential diagnoses of an intra-atrial mass.

Key words: Atrial, cardiac tumors, papillary fibroelastoma

INTRODUCTION

Papillary fibroelastoma (PFE) is the third-most common benign cardiac tumor followed by atrial myxoma and lipoma. Predomnately, it is located on the valvular endocardium of the left heart. Its origin from the right atrium is rarely seen where it is often misdiagnosed as atrial myxoma or thrombus. Here we report one such case of PFE at the junction of right atrial wall and superior vena cava (SVC) which was initially treated with anticoagulation due to suspicion of thrombus. This case is unique due to location of the tumor and its attachment with superior vena cava which has not been reported previously according to the best of our knowledge.

CASE REPORT

An 81-year-old Caucasian male with a history of hypertension, hyperlipidemia, coronary artery bypass graft, aortic valve replacement, atrial fibrillation, ischemic stroke, and patent foramen ovale presented to our cardiology clinic for a follow-up. Transthoracic echocardiography (TTE) suggested the presence of an 11 mm mobile, pedunculated mass at the superior portion of the right atrium. This was confirmed by transesophageal echocardiogram (TEE), which showed a 12 mm mobile mass at the junction of caudal portion of superior vena cava (SVC) and right atrial appendage [Figure 1]. A right atrial thrombus was considered and the patient was started on anticoagulation with coumadin. A follow-up TEE was obtained 2 months later and there was no definitive change in size of the mass. Surgical excision of the mass was considered for establishing a diagnosis. Preoperative cardiac catheterization showed triple vessel disease. The patient underwent coronary artery bypass grafting and shaved excision of the right atrial mass from base of the stalk with the aid of intra operative TEE. Histopathology of the mass was consistent with PFE [Figure 2]. His follow-up TEE after 3 months was negative for a recurrence.

DISCUSSION

PFE is the most common primary tumor of cardiac valves and most frequently located on the left side. More than 80% of these tumors are located on the valvular endocardium and most commonly involve either the aortic or mitral valve. Most non-valvular PFEs are confined to the left ventricle. Its origin from the right...
atrium is extremely rare. We describe a rare case of PFE located in the right atrium.

Estimated incidence of right atrial PFE is 1.8% to 2.3% and non-valvular right atrial PFE is even rarer. Hakemi, et al., reviewed the reported cases of non-valvular right atrial PFE and most of these cases were diagnosed incidentally during routine cardiac work-up due to unrelated symptoms. Right atrial PFEs generally remain asymptomatic until they grow larger in size. These are benign tumors but they can be fatal due to their propensity to embolize. Paradoxical emboli can occur in the setting of a right to left shunt.

TTE is the initial test of choice and has sensitivity of 88.9% and specificity of 87.8%. TEE is more sensitive than TTE if tumor size is less than 20 mm. Other diagnostic modalities are computed tomography and gadolinium enhanced magnetic resonance imaging. Histologically, PFE’s have characteristic gross appearance described as “sea anemone” like branching tumors with multiple papillary fronds arising from central stalk once placed under water. Under microscopy each frond shows an avascular core of elastin and collagen fibers lined by a flat endocardium.

All symptomatic patients with a right atrial PFE should undergo surgical resection. Surgery is recommended even in asymptomatic patients if the tumor size is more than 10 mm because these tumors have a tendency to embolize over a period of time.

Anticoagulation may be considered if the presence of a thrombus is suspected. Although rare, PFE should be considered in the differential diagnosis of an intra-atrial mass. Our case highlights the unusual location of this benign tumor which clinicians need to be aware of.

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**REFERENCES**


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