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Removal of Unusual Multiple Aortic Valve Fibroelastoma Using Right Mini-Thoracotomy

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Abstract

Papillary Fibroelastomas are rare, primary cardiac tumors. They are typically diagnosed as an incidental finding but can also present as thrombo-embolic events. We present the case of 78-years- old man who presented to emergency room with a cerebrovascular event (CVE). Transesophageal echocardiography (TEE) revealed the presence of a mass on the aortic valve. Intra-operatively, two masses were found: one was highly mobile attached to left ventricular (LV) side of the left coronary cusp, another small size mass attached to the non-coronary cusp, and there was suspicion of a mass attached to the mitral valve. The two masses were surgically excised using a right mini-thoracotomy, and histopathological examination confirmed the diagnosis.”

Keywords: Aortic valve, Fibroelastoma, Right mini- thoracotomy

1. Introduction

Primary cardiac tumors are rare entities. Approximately 90% of primary cardiac tumors are benign and only 10% are malignant [1]. Cardiac valve tumors such as cardiac papillary fibroelastoma (CPF) are the second most common benign cardiac tumor and six times less than atrial myxoma, the most common benign cardiac tumor [1, 2]. Aortic valve fibroelastoma mass when present can be silent without any symptom or can lead to devastating complications including systemic embolization, myocardial infarction and death [3]. Aortic valve fibroelastoma when present can be attached to one or more of the aortic valve leaflets. When cerebralvascular events (CVE) occurred, transesophageal echocardiogram (TEE) is imperative to determine the emboli source and presence of any cardiac mass. In this

article we are presenting a case of multiple aortic fibroelastoma in a patient with CVE diagnosed to have multiple masses attached to the LV side of both left and non-coronary aortic cusps excised successfully using right mini-thoracotomy open-heart surgery.

2. Case presentation

A 78-year-old male with no medical history presented to the emergency room with first-time weakness of his left side with no history of fever. He was fully conscious and alert with only difficulty in speech. Physical examination was unremarkable. Urgent brain computed tomography (CT) and magnetic resonance (MRI) showed recent right-side occipital infarction with no sign of bleeding (Fig. 1). No masses were seen by TEE in left atrial (LA) appendix or LV cavities with good LV function. However, there was a solitary, well-defined highly

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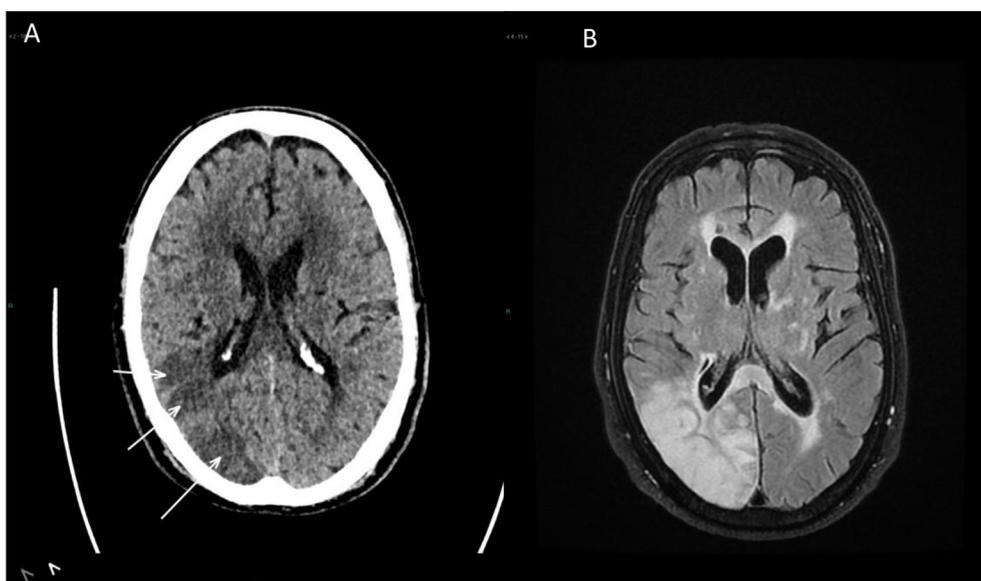


Fig. 1. Brain CT (A) and MRI image (B) showing right acute-subacute occipito-parietal infarction.

mobile mass measured 0.5×0.7 cm attached to the left-ventricular side of the aortic left coronary cusp causing mild-moderate aortic regurgitation (Fig. 2). All inflammatory blood tests including blood cultures, Procalcitonin level and leukocytosis excluded infective endocarditis. Cardiac CT angiography revealed mild, non-obstructive left coronary lesion with the presence of the aortic valve mass (Fig. 3). The presence of highly mobile mass attached to the aortic valve with recent CVE is an indication for surgical intervention to prevent recurrent systemic embolization. However, the surgery was postponed for 2 months upon neurology advice to prevent any hemorrhagic transformation of the cerebral infarction lesion during open-heart surgery. Intra-operative TEE showed the presence of two aortic valve masses, one large in size attached to the left

coronary cusp and another small size mass attached to non-coronary aortic cusp as well a suspicion of a small mass attached to the P3 of the mitral valve. The surgery was conducted through small right mini-thoracotomy incision (5 cm) using direct aortic and venous cannulation. After opening of the aorta, a large mass was found to be attached to the LV side of left coronary cusp and other small mass attached to the noncoronary cusp (Fig. 4). Both masses were shaved carefully without damaging the aortic leaflets. We elected to open the LA for better assessment of the mitral valve so a small incision in the LA roof was made and careful inspection of the mitral leaflets revealed absence of any mass attached to the mitral leaflets. Post-operative TEE showed no residual masses with good aortic valve function without any regurgitation. The postoperative period

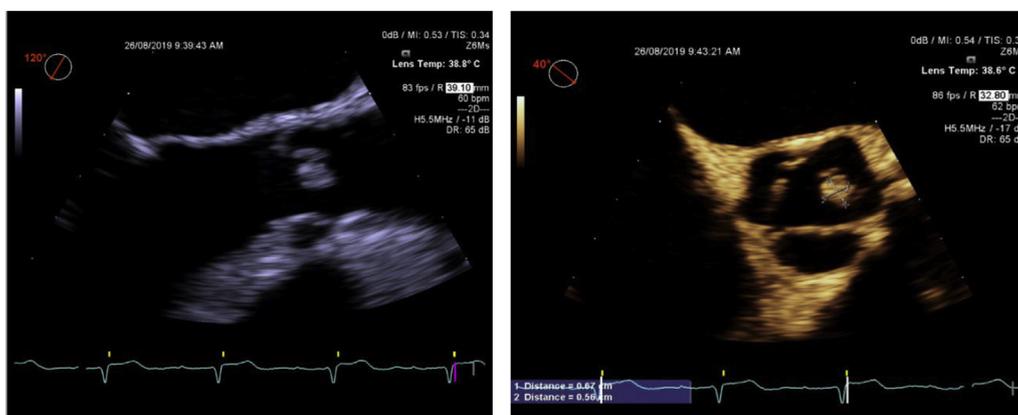


Fig. 2. Transesophageal Echocardiography showed a mass attached to the left ventricle side of the left coronary aortic leaflet.

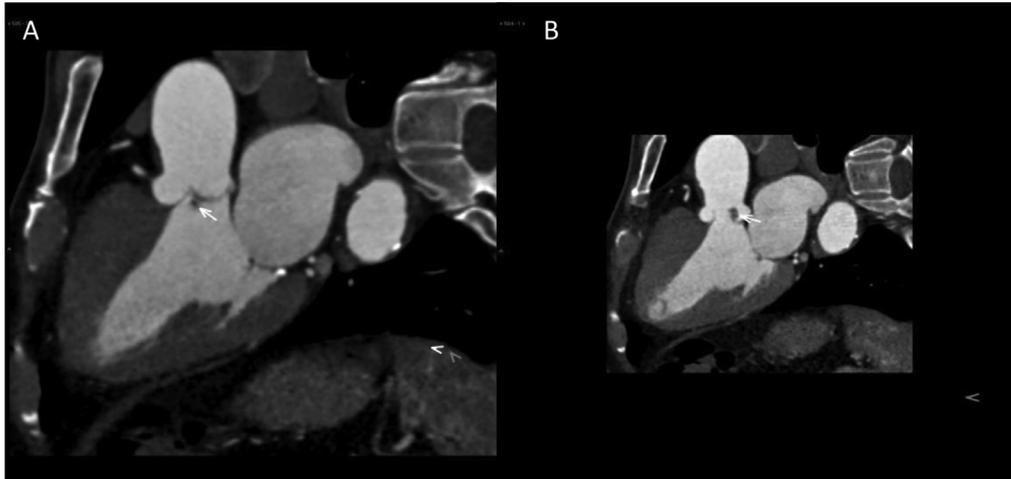


Fig. 3. Cardiac CT revealing mass attached to the left coronary cusp during cardiac (A) Diastole, and (B) in Systole.

went uneventfully, and the patient was discharged home in day 5 postoperatively on Aspirin for 3 months. At the follow up the patient was doing well without any aortic regurgitation. Histology examination of the removed mass confirmed the diagnosis of the papillary aortic valve fibroelastoma.

3. Discussion

Papillary fibroelastoma is a rare benign lesion, consider the second most common primary cardiac tumor and represents 10% of all primary cardiac tumors with a predilection for the aortic valve [1,3]. They are usually small solitary slow-growing lesions which typically attached to the left-ventricular side of the left-sided valves. However, it can be attached to the mitral valve, left atrium or left ventricle walls [4]. CPFs have been documented in patients ranging from 11 months to 86 years of age and are more likely to occur in women than men [5]. The origin of

these lesions is not known and believed to be acquired rather than congenital. One widely accepted theory on their origin is the microthrombus theory that is small thrombi combine at a site of endothelial damage and form a tumor [5]. The clinical presentation of the CPF varies widely from an incidental finding in an asymptomatic individual to cardiac or/and neurological manifestations relating to embolism or obstruction. In the retrospective single-center study, 32% of symptomatic patients presented with neurological events (Transit ischemic attack, syncope, stroke), coronary ischemia (angina, myocardial infarction, cardiac arrest, sudden death) [6]. Our patient presented with cerebral infarction of the right occipital lobe due to CPF fragmentation of the tumor. Echocardiography remains the gold standard in the diagnostic workup of suspected cardiac sources of emboli. Preoperative TEE showed clearly the mass in the aortic valve and its attachment, however, did not show the small mass



Fig. 4. Intraoperative view, a jelly like mass attached to (A) left coronary cusp and (B) non-coronary cusp.

attached to the non-coronary cusp. Also, intra-operative TEE failed to prove the absence of the mitral valve mass. Sun et al., [5] in a single-center retrospective review of 162 pathologically confirmed cases reported the sensitivity and specificity of TTE in the dictation of CPF 0.2 cm to be 88.9% and 87.8% retrospectively. As per our hospital protocol, any patient aged above 60 years and going for mini-thoracotomy cardiac surgery should have cardiac CT and coronary angiography to determine the ascending aorta anatomy and position related to the thoracic wall, as well as to evaluate the patient for the possibility of coronary artery disease. However, due to the location of the mass in our patient, it was felt that the angiography carries a high risk for mass detachment and systemic embolization. Dynamic cardiac CT was an excellent alternative that provides us with useful information regarding the ascending aorta anatomy as well as coronary arteries status and the mass characteristic (Fig. 2). Surgical treatment is indicated for symptomatic patients to prevent further ischemic or thromboembolic events. The surgery can be performed using conventional median sternotomy, upper mini-sternotomy or right mini-thoracotomy depends on the surgeon's experience and preference. Different surgical techniques for CPF excision have been reported; shaving excision, excision with valve repair and excision with valve replacement. In our patient shaving of the two left and non-coronary cups masses were done without interference with the integrity of both aortic leaflets. The surgical decision for asymptomatic patients with an incidental cardiac valve tumor is controversial. Gowda et al., [6] reported 12 tumor-related deaths from CVE and coronary obstruction in the cases of 25 medically-treated CPF patients, suggesting that the likelihood of developing symptom overtime should not be ignored and they recommend surgery if the tumor is mobile as the mobility of the tumor was a predictor of mortality and risk of embolism. The decision on

management of CPF is difficult as there have been no randomized controlled trials.

4. Conclusion

Cardiac papillary fibroelastoma is a rare benign tumor with the potential for devastating systemic embolization and should be considered in the differential diagnosis of a patient with CVE. Non-invasive diagnostic investigations like TTE, TEE, and dynamic cardiac CT are very beneficial in the detection of the systemic embolic source without increasing the risk of mobile mass detachment during coronary angiography. Fibroelastoma can be in one or multiple locations attached to the left side valves or cardiac cavities. Surgical shaving excision using the right mini-thoracotomy is safe and an accepted approach in symptomatic patients to prevent further embolic events.

Conflicts of Interest

The authors have no conflicts of interest relevant to this article.

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