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Case Report

Papillary Fibroelastoma of the Aorta Presenting as a Non-ST Segment Elevation Myocardial Infarction

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ABSTRACT

A 42 yr female presented with atypical chest pain and was found to have a filling defect in the Sinus of Valsalva on CT angiography. Transesophageal echocardiography revealed that the defect was a mobile peculated mass with features suggestive of a fibroelastoma. She ruled in for a NSTEMI with abnormal cardiac enzymes and due to progressive chest pain coronary angiography was performed which revealed a thrombotic occlusion of the right coronary artery which was managed by percutaneous intervention. Post procedure she was managed non-operatively with aggressive systemic anticoagulation and a P2Y12 inhibitor. Interval imaging in six weeks documented a residual stump but there were no further complications.

Keywords: Primary aortic tumors; Papillary fibroelastomas; Angiography; Trans-esophageal echocardiogram

LEARNING OBJECTIVES

Utilizing a heart team approach to the diagnosis and treatment of a varied presentation of a primary aortic tumor.

INTRODUCTION

Primary tumors of the aorta are rare, usually malignant [1-4], and can mimic a wide range of conditions including aortic dissection and coarctation of the aorta. These tumors usually present incidentally or with embolic symptoms [5] and are usually treated by surgical excision with adjuvant radiation and chemotherapy in some cases [1].

Papillary Fibroelastomas [PF] are benign tumors and occur extremely rarely as a primary aortic tumor [3]. They are pedunculated, a vascular structures usually located on the endocardium or the cardiac valves. The tumor is covered with a single layer of endothelium with fibrils and whorls, which can be covered with thrombus or a gelatinous material [6]. PF are also found incidentally but can also present with embolic symptoms such as a cerebrovascular accident or a myocardial infarction. We aim to report a unique presentation of a papillary fibroelastoma arising in the Sinus of Valsalva, which presented with an acute myocardial infarction due to embolization as well as a ball and valve occlusion of the ostium of the right coronary artery.

CASE REPORT

A 42-year-old female with no significant risk factors for coronary artery disease presented to an outside medical facility with a 3-to-4-day history of recurrent and progressive chest discomfort radiating through to her back. Initial ECG, in the emergency room, documented no significant ECG changes. Due to her lack of significant risk factors, symptoms that were not specific for an acute coronary syndrome and the description of the pain, the initial assessment was to rule out a possible aortic dissection; therefore, a CT scan of the chest was obtained. The scan documented a filling defect in the ascending aorta at the Sinus of Valsalva, which protruded into the lumen (Figure 1A).The patient was emergently transferred to our institution with a presumed diagnosis of a type A aortic dissection. Upon careful review of her scan by our cardiac surgeons it was apparent that this was not an aortic dissection or a penetrating aortic ulcer. She did have abnormal cardiac enzymes and ruled in for a Non-ST Segment Elevation Myocardial Infarction [NSTEMI] with a troponin level of 5.7 ng/ml, which ultimately peaked at 37 ng/ml. We thought that further evaluation of the defect would be critical to determine further treatment especially in light of the elevated cardiac enzymes. We decided to perform a Trans-Esophageal Echocardiogram [TEE]to further characterize the mass. It revealed a pedunculated, mobile mass measuring 1.6cm x1.0 cm in the Sinus of Valsalva adjacent to the ostium of the

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right coronary artery but with no evidence of aortic disruption. (Figures 1B-1D).With progressive chest pain which was not relieved by optimal medical management she was triaged emergently to the cardiac catheterization laboratory where angiography documented a thrombotic occlusion of the right coronary artery in the mid segment with a proximal translucent filling defect (Figure 2A). We performed aspiration thrombectomy of the proximal, mid and distal segments with stent placement in the mid segment due to intimal dissection (Figure 2B). She was treated with systemic anticoagulation with Coumadin and a P2Y12 inhibitor.

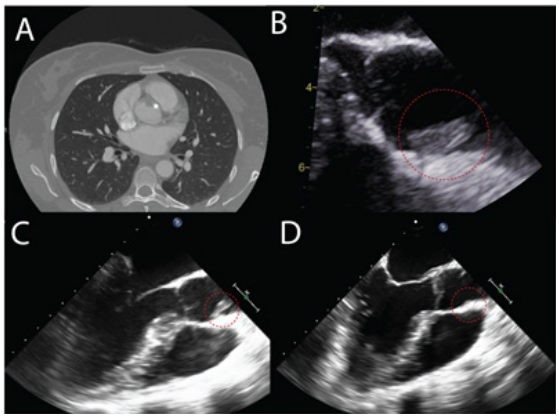

Figure 1(a-d): CT scan of the chest with pedunculated mass at the inferior sinotubular junction.

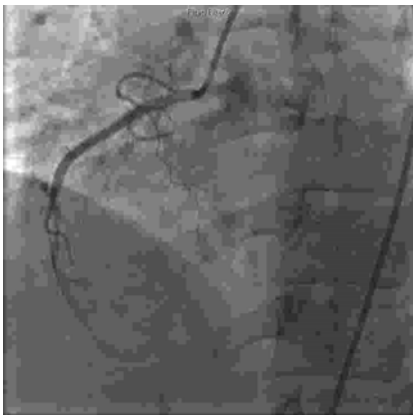

Figure 2A: Right coronary artery pre-procedure angiogram with total occlusion in the mid segment and haziness in the ostium.

A TEE was performed 6 weeks later to re-evaluate the mass and it documented a 2.0 mm stump at the site of the pedunculated mass (Figure 1D). A heart team discussion on the utility of surgical intervention concluded that non-operative management with continued systemic anticoagulation would be undertaken with accompanying interval imaging of the lesion. This was with the understanding that surgical excision of the mass would be performed if there were any recurrent symptoms or further complications resulting from the residual stump while being treated with continued systemic anticoagulation and P2Y12 inhibitors.

The patient was followed for 18 months without recurrence of symptoms or further sequelae. Systemic anticoagulation was discontinued after 3 months. Afterward she was maintained on dual anti-platelet therapy.

DISCUSSION AND CONCLUSION

Cardiac and aortic masses with concurrent complications require urgent imaging to assess their morphology and help determine further management. The first step in this case was to determine whether the presentation was due to a dissection, thrombus or tumor. With this pedunculated mobile mass, TEE is the diagnostic modality of choice due to its ability to visualize and characterize mobile structures as well as its ability to provide superior resolution of cardiac and proximal aortic structures. PF's remain largely asymptomatic and their diagnosis before complications can be difficult. The pedunculated mobile mass with flowery head noted on TEE suggested that this mass was a papillary fibroelastoma. The initial angiogram of the right coronary artery suggested thrombotic occlusion of the mid vessel but proximally at the ostium the haziness suggested occlusion proximally from the gelatinous material noted in pathologic samples. Depending on the length of the stalk, size, location and mobility of the tumor it has been known to obstruct cardiac valve resulting in syncope. The mobility and proximity of this pedunculated mass as well as the intermittent symptoms over 3 to 4 days before presentation suggest either a ball valve occlusion of the right coronary or intermittent embolization into the right coronary artery. While surgical management of these tumors are the treatment of choice the medical history, patient presentation in addition to the location and morphology of the tumor are typically the most important data to determine the optimal management plan for these patients. Furthermore, a multidisciplinary team of cardiac surgeons and cardiologists should decide the best treatment on a patient-by-patient basis weighing the risks and benefits of each treatment plan.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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