A Rare Location and Presentation of Papillary Fibroelastoma

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Papillary fibroelastomas are the second most common primary cardiac tumors. They can cause symptoms due to embolic phenomena or obstruction. We present a 60-year-old woman with new-onset atrial fibrillation along with complaints of shortness of breath and chest pressure. Her transesophageal echocardiogram and computed tomography angiogram revealed a mass (10 x 8 mm) involving the left coronary sinus and causing intermittent obstruction of the left main coronary artery. We proceeded with surgical removal of the mass due to its peculiar location, size, and hypermobility. Histopathology diagnosed the mass as papillary fibroelastoma. This case is unique in its location, invaginating into the left main coronary artery and causing obstruction. We also discuss the current treatment options and need of randomized studies for further recommendations.

Case Report
A 60-year-old Caucasian woman was referred for evaluation of new-onset atrial fibrillation. Her complaints included shortness of breath on exertion and chest pressure. On examination, her heart rate was 90 beats per minute, irregular and without any murmurs. She was started on warfarin and underwent nuclear perfusion scan, which revealed reversible ischemia in the anterolateral myocardial walls. She was admitted to the hospital for transesophageal echocardiogram-guided cardioversion followed by elective cardiac catheterization. Her transesophageal echocardiogram showed an echogenic, pedunculated mobile mass located at the junction of the left coronary sinus and the root of the aorta. This mass appeared to be obstructing the left main coronary artery intermittently (see Figure 1). Cardiac computed tomography confirmed the findings of the transesophageal echocardiogram (see Figure 2). The cardiothoracic team was consulted for excision of this mass. Intra-operatively, a pedunculated mass was noted, attached to the free surface of the aorta in close proximity to the commissure between the left and non-coronary cusps. Gross examination revealed a gray-tan, polypoid mass that measured 14 x 10 x 8 mm with numerous fine papillations covering the tissue. Histology showed papillary fronds with a myxoid stroma, lined by benign endothelial cells (see Figure 3). These features led to a diagnosis of papillary fibroelastoma. Her post-operative recovery was uneventful and she was discharged home on the seventh day of hospitalization.

Discussion
Papillary fibroelastomas are the second most common primary cardiac tumors. They account for 7.9% of all cardiac tumors.1 In the past they were thought to be benign in nature, but more recent studies suggested a high risk of embolic complications to coronary, cerebral, and systemic arteries.2,3 They are most commonly found on the cardiac valves and adjacent endocardium. Non-valvular fibroelastomas are rare, with isolated case reports involving the right atrium, left ventricle free wall, apex, and interventricular septum. The uniqueness of this case is its location in the aorta and that it intermittently obstructs the left main coronary artery.

Diagnosis is usually made by an echocardiogram. Transesophageal echocardiogram is superior to transthoracic echocardiogram. Cardiac computed tomography and magnetic resonance imaging studies can aid in the diagnosis, along with confirmation of the location and defining the

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type of cardiac tumor. The natural history of fibroelastoma is poorly understood. These tumors are friable and may lead to tumor embolization. Some experts believe that the true source of embolization is the fibrin strands alone and some others believe platelet and fibrin aggregation on the surface leads to the arterial embolism.4,5 There is no clear consensus about the exact treatment options for fibroelastomas. Around 30% of fibroelastomas were diagnosed incidentally and should be followed up by periodic clinical and cardiac imaging evaluation. Most experts advocate surgical removal of the large (>10 mm), highly mobile fibroelastomas causing recurrent peripheral embolism or symptoms such as an obstruction. Therapeutic long-term anticoagulation should be considered in all symptomatic non-surgical candidates.6,7 Post-surgical recurrence of the tumor has not been reported in the literature. In this case, we proceeded with surgical removal of the tumor due to its peculiar location, size, and hypermobility and in that it intermittently obstructed the left main coronary artery. In conclusion, this is a rare case report of fibroelastoma involving the left coronary sinus and causing intermittent obstruction of the left main coronary artery. Randomized studies are required to assess the treatment options and for further recommendations.