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Case Report

Complete occlusion of the left main trunk coronary artery by a cardiac papillary fibroelastoma in a hemodynamically unstable patient



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ABSTRACT

Papillary fibroelastomas are benign cardiac tumors with high embolic potential. The majority of cases of complete obstruction of the left main trunk (LMT) of the coronary artery are diagnosed via autopsy following sudden death; survival is rare in this setting. We present the case of a 60-year-old woman who underwent stent placement in the LMT three years prior to developing chest pain and cold sweats. On coronary arteriography, the catheter could not be advanced into the LMT due to resistance in the ostium. Insertion of the catheter was achieved after the resolution of resistance via catheterization of the LMT by means of an intra-aortic balloon pump drive system. The LMT was normal, and the patient's circulatory failure improved. The cause of the LMT embolism was a cardiac papillary fibroelastoma. Primary surgical excision is the recommended therapy for symptomatic cardiac papillary fibroelastoma. If the patient is hemodynamically stable, it may be possible to delay surgery. However, the patient in question developed cardiogenic shock secondary to two-vessel obstruction by the tumor. Therefore, even if the tumor had been removed using an intra-aortic balloon pump, the patient may not have survived until surgery.

<Learning objective: Primary surgical excision is the recommended therapy for symptomatic cardiac papillary fibroelastoma. If the patient is hemodynamically stable, it may be possible to delay surgery. However, hemodynamically unstable patients may not survive until surgery. Therefore, emergent therapy is a useful stop-gap measure until surgery is feasible.>

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Introduction

Although the majority of patients with cardiac papillary fibroelastomas (CPFs) are asymptomatic, the clinical presentation of this entity may include severe thromboembolic complications, myocardial ischemia, infarction, stroke, and sudden cardiac death [1]. Acute myocardial infarction (AMI) secondary to CPF has historically been attributed to either adherent thrombi on the surfaces of or tumor fragments within the coronary arteries, as well as occlusion of the left coronary artery (LCA) ostium. The majority

of cases of LCA occlusion are diagnosed via autopsy following sudden death [2]; survival is rare in this setting [3]. Previous reports [4,5] have attributed MI to either intermittent obstruction of the LCA ostium or to the obstruction of a single vessel by the tumor; the patients described in these instances were hemodynamically stable. We describe a patient who presented with cardiogenic shock secondary to AMI. The patient's diagnosis was two-vessel obstruction secondary to a left main trunk (LMT) embolus from a CPF.

Case report

A 60-year-old female with a history of hyperlipidemia and prior angina pectoris underwent stenting of the LMT three years prior to hospitalization for chest pain and cold sweats. On examination, the patient's blood pressure was 94/68 mmHg, her pulse was

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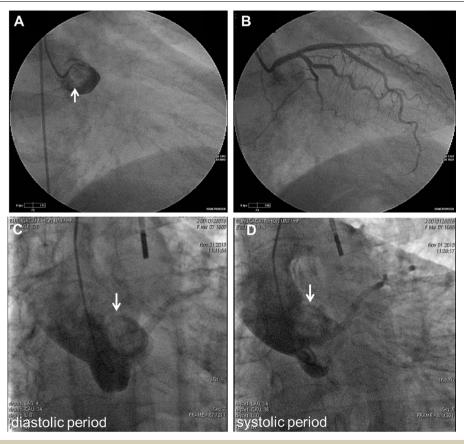
100 beats per minute, and her body temperature was 36.4 °C. Electrocardiography (ECG) revealed sinus rhythm, ST-segment elevation in leads I, aVL, and V1-V4, and ST-segment depression in leads II, III, aVF, and V5-V6. Chest radiography demonstrated an area of hemostasis at the pulmonary hilum. Transthoracic echocardiography (TTE) demonstrated severe anterior and lateral septal akinesis. Evidence of either aortic stenosis or regurgitation was not observed. At the time of the coronary arteriographic (CAG) examination, the right coronary artery was normal: a 5-Fr catheter could not be advanced into the LCA due to resistance in the ostium. Aortography demonstrated that the LMT was completely obstructed (Fig. 1A); the obstruction was attributed to a thrombus. The patient's blood pressure decreased to 56/34 mmHg, and circulatory failure subsequently developed. Both oxygen and catecholamines were subsequently administered for hypoxemia and hypotension, respectively, and an intra-aortic balloon pump (IABP) was inserted. The catheterization of the LCA under the IABP drive system was initially characterized by resistance in the catheter tip, but the successful insertion of the catheter was ultimately achieved following the resolution of the resistance. On CAG, the LCA was normal (Fig. 1B), and the patient's chest pain resolved. Her peak cardiac enzyme values were as follows: creatine kinase (CK), 7140 U/I; CK-MB, 635 U/I; and troponin I, 406.8 ng/ml. The patient underwent a repeat CAG on the 18th day of her admission due to improvement in her cardiac failure. Slight resistance was encountered at the same location as noted previously. The patient subsequently developed chest pain and dyspnea. Her ECG demonstrated ST-segment elevation, and her blood pressure decreased to 68/42 mmHg. Therefore, an IABP was

promptly utilized, and the patient's symptoms eventually resolved. A filling defect in the proximal portion of the LMT inlet was visualized via aortography with the assistance of the IABP (Fig. 1C and D). Therefore, the patient underwent emergency surgery. Preoperative transesophageal echocardiography demonstrated a pendulous mass filling the left coronary cusp (Fig. 2A). During the operation, a tumor attached to the rim of the left coronary cusp of the aortic valve was identified (Fig. 2B). Surgical removal of the tumor without valve replacement was performed without complications. The tumor measured 19 mm \times 13 mm \times 5 mm (Fig. 2C), and the results of the histopathological examination were consistent with a benign papillary fibroelastoma (Fig. 2D). The patient remained free of symptoms following surgery.

Discussion

Although most cases of MI are caused by atherothrombosis, MI occasionally occurs in patients with normal appearing coronary arteries on angiography. MI with normal coronary arteries can result from multiple conditions, including coronary embolism, coronary spasm, coronary artery anomalies, coronary dissection, hyper-coagulable states, and imbalances between blood flow and blood supply. A rare cause of coronary embolism is CPF, the second most common benign primary cardiac tumor after myxomas which represents 10% of all primary cardiac tumors and exhibits a frequency of approximately 0.02% [2,6].

CPFs are benign cardiac neoplasms. CPFs are always found incidentally during autopsy, during cardiac surgery, or via echocardiography. Sun et al. reported that the sensitivity and



(A) A coronary angiogram of the left main coronary artery demonstrating a filling defect (arrow). (B) Under intra-aortic balloon pump drive (IABP), a catheter was inserted into the left coronary artery after encountering resistance. The left coronary artery was normal. (C and D) The tumor (arrow) migrated toward the cardiac chamber in a diastolic period and toward the aortic arch in a systolic period after IABP insertion.

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