Case Report

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Cardiac papillary fibroelastoma: a rare cause of ST-segment elevation myocardial infarction: a case report

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Introduction and importance: Primary tumors of the heart are extremely rare occurrences. Among them, cardiac papillary fibroelastoma (CPF) is the second most common type. Although these tumors are usually benign, they can pose a risk of embolization, which may lead to severe complications like sudden death or embolization affecting the neurological, systemic, or coronary vasculature. Such complications can be life-threatening.

Case presentation: In this report, the authors present the case of a 68-year-old woman who experienced ST-segment elevation myocardial infarction due to embolization from a large papillary fibroelastoma. To address the issue, the authors performed a minimally invasive surgical removal and resection of the aortic valve, followed by a histological examination to confirm the diagnosis.

Clinical discussion: This case report discusses a rare occurrence of myocardial infarction caused by tumor embolization from a CPF. The patient presented with complete blockage of a coronary artery in the absence of atherosclerotic disease. Through a comprehensive workup, including transesophageal echocardiography, the CPF was identified as the source of embolization. Surgical resection of CPFs is curative, and recurrence has not been documented. Clinicians should consider CPFs in cases of coronary artery occlusion without atherosclerotic disease and employ transesophageal echocardiography for diagnosis. Prompt surgical intervention leads to an excellent prognosis and prevents recurrent embolization.

Conclusion: This report emphasizes the importance of recognizing the potential complications associated with papillary fibroelastoma-induced embolization to the coronary arteries and highlights the need to mitigate the risk of such complications occurring.

Keywords: aortic valve, cardiac papillary fibroelastoma, cardiac tumor, case report, STEMI

Introduction

Papillary fibroelastoma is an extremely rare primary cardiac tumor. Its prevalence is estimated to range from 0.0017 to 0.33% based on the autopsy series[1]. These tumors are typically incidentally discovered during autopsies and are classified as benign. While they are generally harmless, papillary fibroelastomas can have adverse effects on patients if complicated by the extremely rare occurrence of embolization, which may lead to myocardial infarction or stroke[1,2]. From the literature review, it is evident that tumor mobility is an independent predictor of adverse outcomes such as mortality or nonfatal embolization. Consequently, surgical intervention is recommended for symptomatic patients or when the tumor exhibits mobility[3].

In this report, we present the case of a 68-year-old woman who arrived at our emergency department with complaints of chest pain. The onset of pain followed a mechanical fall that occurred ten minutes prior to her arrival. Further diagnostic tests,

HIGHLIGHTS

- Myocardial infarction caused by tumor embolization from a cardiac papillary fibroelastoma (CPF) is a rare occurrence.
- A comprehensive workup, including transesophageal echocardiography, can identify CPF as the source of embolization.
- The curative treatment for CPF is surgical resection.
- CPFs should be considered in cases of coronary artery occlusion without atherosclerotic disease.
- Prompt surgical intervention offers an excellent prognosis and prevents recurrent embolization.

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including electrocardiography, percutaneous coronary intervention, and transesophageal echocardiogram, revealed ST-segment elevation myocardial infarction. The infarction was attributed to embolization from a papillary fibroelastoma located at the right coronary cusp.

This work has been reported in line with the CARE 2013 guidelines[4].

Presentation of case

A 68-year-old woman was admitted to the hospital due to chest pain. The patient presented at the emergency room with typical chest pain that was retrosternal in location and provoked by exertion. She described the pain as central, nonradiating, and characterized it as chest tightness, which she attributed to a fall that occurred ten minutes prior. She rated the pain as a seven out of ten on the pain scale and reported concomitant shortness of breath. The review of symptoms did not reveal any other significant findings.

The patient has a 15-year history of Parkinson's disease and had experienced multiple falls in the past. Two months prior, she was diagnosed with stage 3C ovarian cancer with peritoneal metastasis and had undergone one round of chemotherapy consisting of carboplatin and paclitaxel. Additionally, the patient underwent paracentesis to manage ascites. Two weeks ago, she was hospitalized for bilateral segmental acute pulmonary embolism with deep vein thrombosis. As a result, she was prescribed Apixaban oral anticoagulation. Other medical conditions that were present in the patient's past include hyperlipidemia, restless leg syndrome, gastroesophageal reflux disease, and insomnia.

Upon arrival, the patient's vital signs were as follows: blood pressure of 129/74 mmHg, heart rate of 100 beats per minute, respiratory rate of 18 breaths per minute, oxygen saturation of 98% on 15 l/min, and a temperature of 98.4°F. Despite appearing uncomfortable, the physical examination revealed no abnormalities. Recurrent pulmonary embolism was considered as one of the differentials, and the pulmonology team was consulted. A negative D-dimer result contributed to the assessment and helped rule out pulmonary embolism.

Electrocardiography indicated sinus rhythm with ST elevations in leads II, III, and aVF, as well as T wave inversion in lead aVL (Fig. 1). Emergency coronary angiography revealed complete occlusion of the distal second posterolateral artery branching of the right coronary artery, which was treated with balloon angioplasty (Fig. 2). A 50% stenosis was observed in the mid to distal left anterior descending artery. Due to the distal location and small caliber of the culprit lesion in the second posterolateral artery, as well as the relatively smooth caliber of the rest of the right coronary artery, the cause of the infarction remained unclear, whether it resulted from ruptured atherosclerotic plaque or embolization.

To investigate the presence of a patent foramen ovale or an embolism in transit, an echocardiogram with definity contrast agent was performed after angiography, but no intracardiac shunt was detected. However, a transesophageal echocardiogram revealed the presence of an intrapulmonary shunt and an atrial septal aneurysm. Furthermore, a mobile, well-demarcated, echodense mass measuring 1.46 cm was identified on the right coronary cusp, indicative of papillary fibroelastoma (Fig. 3). Following consultation with cardiothoracic surgery, the mass was surgically removed, along with aortic valve resection. Pathology findings confirmed the diagnosis of papillary fibroelastoma for the aortic mass.

Clinical discussion

Primary cardiac tumors are extremely rare and often discovered incidentally. Among them, myxomas are the most common type[1,2]. Papillary fibroelastomas, also known as cardiac papillary fibroelastomas (CPF), are the second most prevalent primary cardiac tumors. CPF exhibit sporadic occurrence, and the exact etiology leading to their development remains unknown[3,5]. Factors that increase the risk of developing CPF comprise a history of endocardial surgery, a history of rheumatic heart disease, thoracic radiation treatment, and cardiac valve disease leading to trans-valvular pressure differences. Additionally, CPF has been linked to other concurrent conditions like hypertension, diabetes, hyperlipidemia (as seen in our case) and chronic obstructive lung disease[6]. These tumors predominantly manifest on the valves, with the aortic valve being the most commonly affected site as seen in our case[3]. Echocardiograms are useful in distinguishing CPFs from cardiac myxomas. Grossly, CPFs appear as papillary fronds attached to the endocardium by a central stalk. They are avascular papillomas composed of proteoglycans, elastic fibers, and collagen, lined by endocardial cells[1–3]. Histologically, CPFs exhibit a fibrous plaque-like appearance. This composition gives the tumor a firm consistency and reduces the risk of fragmentation or embolization[2]. However, CPFs have a slow growth rate and can potentially accumulate fibrin clots[1,3].

CPF are generally considered benign and often remain asymptomatic, even when located on the left side of the heart[2]. However, despite their relatively benign nature, CPFs have been associated with embolization events, as evidenced by documented cases. Embolization commonly manifests in the cerebral, systemic, or coronary circulation. Instances of heart failure and sudden deaths have also been reported[3]. A comprehensive study conducted over 41 years, analyzing 17 280 patients undergoing heart surgery at a single center, found that embolic phenomena were observed in 30–40% of patients with cardiac tumors. While embolization of the coronary arteries was relatively uncommon, embolization to the central nervous system, kidney, spleen, and extremities was more prevalent[5]. Furthermore, CPF embolization has been linked to complications such as angina pectoris and myocardial infarction[3], which have been well-documented. Another case–control study investigating embolization risks estimated an embolization rate of 25% for cardiac tumors, with structural and anatomical factors significantly impacting the occurrence. CPFs located on the aortic valve and in the left atrium were found to have the highest rates of embolization[7]. Additionally, the presence of coronary pulmonary fistulae at the coronary aortic cusp increases the risk of prolapse into the coronary ostia, potentially leading to cardiac arrest or unexpected death[1,2].

Surgical resection of CPF is considered curative, with no documented cases of recurrence. Resection is recommended when symptoms arise or when the tumor demonstrates mobility, as mobility is an independent predictor of death or nonfatal embolization[3–5,7]. Patients who are not suitable candidates for surgery should be offered long-term anticoagulation therapy;

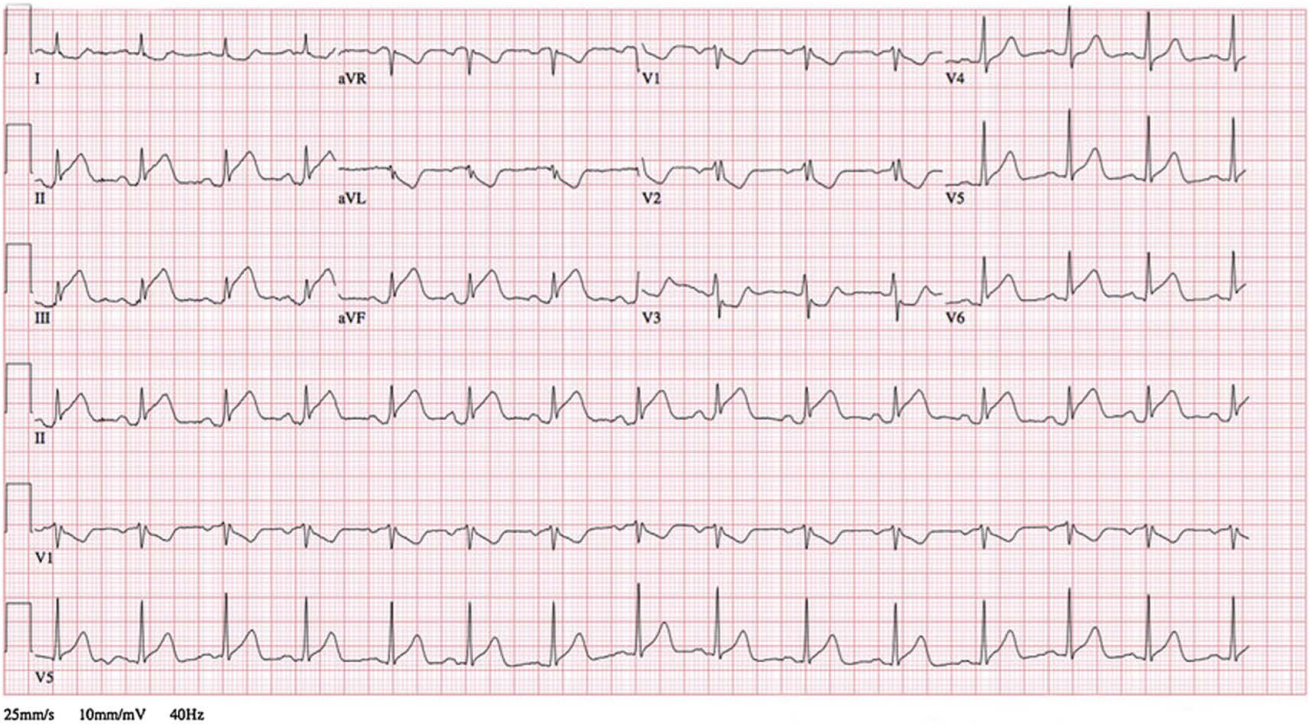

Figure 1. Electrocardiogram showing ST Elevation in leads II, III and aVF, reciprocal change with T wave inversion in aVL lead.

however, the effectiveness of this intervention has not been firmly established[2,3]. It is important to note that approximately one-quarter of CPFs may not be detected on a transthoracic echocardiogram but can be detected on transesophageal echocardiogram. Therefore, patients presenting with myocardial infarction but no evidence of coronary artery disease or other potential sources of embolization should undergo a transesophageal echocardiogram for CPF investigation[8].

In our case report, we presented the case of a 68-year-old woman who experienced coronary embolization resulting from a CPF located at the right coronary cusp. Notably, this patient exhibited complete blockage of the distal second posterolateral artery branching from the right coronary artery, without any indications of underlying coronary artery disease. Further investigation led to the identification of CPF embolization as the source of the problem. Although myocardial infarction is a rare complication of CPF embolization, it emphasizes the importance of prompt intervention. Surgical removal of CPFs is crucial due to the high risk of recurrent embolization, and the prognosis following tumor removal is excellent.

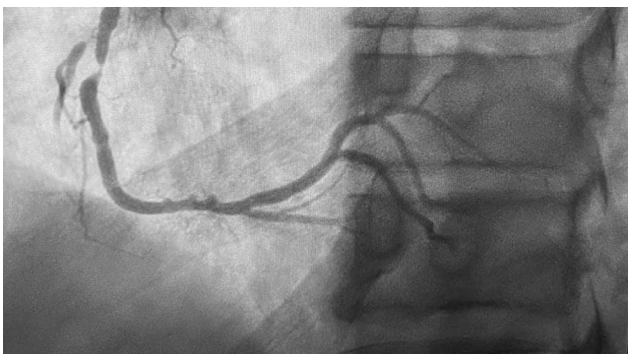

Figure 2. Coronary angiogram showing complete occlusion of the distal second posterolateral artery branching off the right coronary artery.

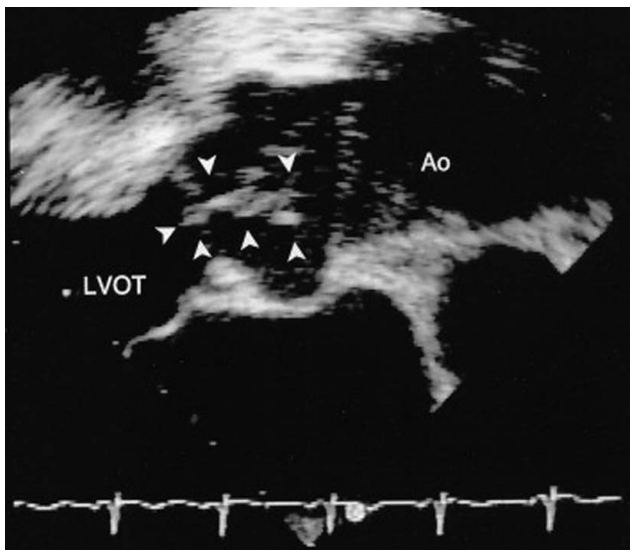

Figure 3. Transesophageal echocardiogram showing a cardiac on the aortic valve. Zoom view of an aortic valve papillary fibroelastoma (arrowheads), showing the linear morphology, with strands visible. Ao, aorta; LVOT, left ventricular outflow tract.

Conclusion

In conclusion, our case report highlights the rarity of papillary fibroelastomas as benign primary cardiac tumors. Despite their infrequency, tumor embolization can lead to severe complications such as myocardial infarction, as evidenced in our patient. When confronted with coronary artery occlusion in the absence of atherosclerotic disease, a comprehensive diagnostic approach, including transesophageal echocardiography, should be undertaken, as demonstrated in this case. By recognizing the potential for tumor embolization, appropriate diagnostic measures can be implemented to ensure timely management and improve patient outcomes.

Ethical approval

Ethical approval was not required for this case report by University of Toledo as it did not involve any interventions or experimentation on human subject and did not contain any identifiable information about the patient.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

F.R., MD: conceptualization, data collection, project administration, and supervision; A.S.T., MD: conceptualization and data collection; M.R., MD: manuscript drafting; M.J.F., MBBS: manuscript drafting; S.K., MBBS: manuscript drafting, editing, and reviewing; S.B., MBBS: manuscript drafting; S.M.G., MBBS: editing and reviewing; S.S., MBBS: editing and reviewing.

Conflicts of interest disclosures

There are no conflicts of interest to declare.

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