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DIAGNOSIS OF CARDIAC PAPILLARY FIBROELASTOMA PRESENTING WITH ACUTE CORONARY SYNDROME AND CONCOMITANT SALMONELLA BACTERAEMIA, MIMICKING AORTIC ENDOCARDITIS

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ABSTRACT

Introduction: A case of fibroelastoma of the aortic valve in a patient with acute coronary syndrome has been reported. Cardiac papillary fibroelastomas (PFE) are rare cardiac tumours that are generally benign. In most cases they are incidental findings, their first clinical manifestation is often associated with embolic events.

Case presentation: We present the case of a 63-year-old man who experienced coronary embolization without any indications of underlying coronary artery disease. Further investigation led to the identification of the cause of embolization. The concomitant presence of Salmonella bacteraemia, associated with symptoms of gastrointestinal inflammation and elevated inflammation indices, led to the hypothesis of valvular endocarditis. Therefore, a transoesophageal echocardiogram was performed, which confirmed the presence of a mobile, well-demarcated, echo-dense mass identified on the left coronary cusp. Despite the diagnostic uncertainty between fibroelastoma and endocarditic formation, the occurrence of the embolic event and the marked hypermobility we decided that prompt intervention was required. Surgical cleavage of the valve formation was performed, revealing macroscopic morphological characteristics consistent with, PFE which was confirmed by the histological finding.

Conclusions: This case highlight the importance of a comprehensive diagnostic approach, including transoesophageal echocardiography, in patients with embolic events with no obvious evidence of an embolic causes.

KEYWORDS

Fibroelastoma, endocarditis, aortic valve, echocardiography, Salmonella bacteraemia

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LEARNING POINTS

- Cardiac papillary fibroelastomas (PFE) are rare cardiac tumours that are generally benign.
- Their first clinical manifestation is often associated with embolic events, also leading to acute coronary syndrome.
- In patients with embolic events with no obvious evidence of embolic causes, transoesophageal echocardiogram should be performed to look for PFE.

INTRODUCTION

Papillary fibroelastomas (PFEs) are rare tumours of the heart and accounting for approximately 10% of all benign primary cardiac tumours. In terms of incidence, they are the second most common cardiac tumours after myxomas[1]. They are usually located on the cardiac valves, but also on the chordae tendineae, papillary muscles, or atrial and ventricular septum[2]. Diagnosis is often incidental; embolic events are rarely the first clinical presentation.

We report the case of a 63-year-old man, who was referred for acute coronary syndrome who was found to have a PFE of the aortic valve.

CASE DESCRIPTION

A 63-year-old man was admitted to the emergency department of the "Card. Panico, Hospital" (Italy) by the emergency medical service. The patient following an out-of-hospital cardiac arrest (OHCA) was promptly resuscitated. In our emergency room the patient presented with chest pain and hypotension. The electrocardiogram (ECG) revealed a pattern indicative of an acute lateral myocardial infarction. As a result, the patient was promptly transferred to the cardiac catheterization laboratory for urgent coronary angiography. The patient has a history of arterial hypertension treated with angiotensin-converting enzyme (ACE) inhibitor. He was a non-smoker with no family history of cardiac disease. For 2 weeks prior to admission, the patient had experienced intermittent fever, abdominal pain, and diarrhoea. Upon arrival, the patient's vital signs were as follows: blood pressure 86/54 mmHg, heart rate 111 beats per minute, respiratory rate 18 breaths per minute, oxygen saturation 96%, and temperature 37.8°C. The clinical examination was unremarkable. An ECG revealed ST-segment elevation in leads I, aVL and V5-V6

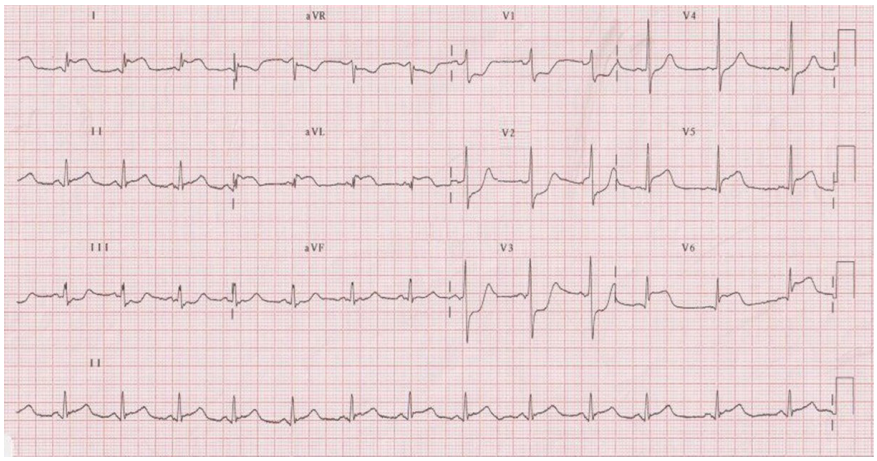


Figure 1. Electrocardiogram on admission. Typical signs of lateral STEMI are present. ST-segment elevation in leads I, aVL and V5-V6 with reciprocal ST depression in leads III, aVF and V1-V3.

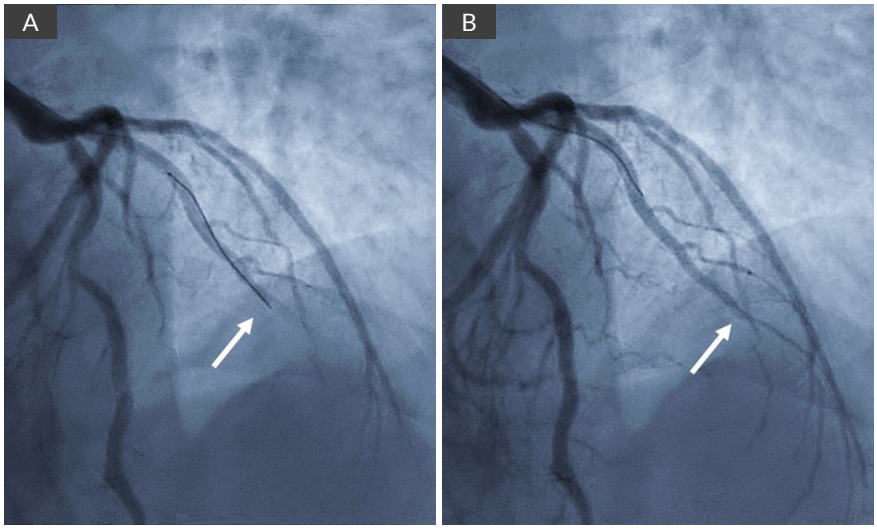


Figure 2. Coronary angiography. A) Occlusion of the diagonal branch (arrow) and B) subsequent restoration of coronary blood flow after thromboaspiration are highlighted.

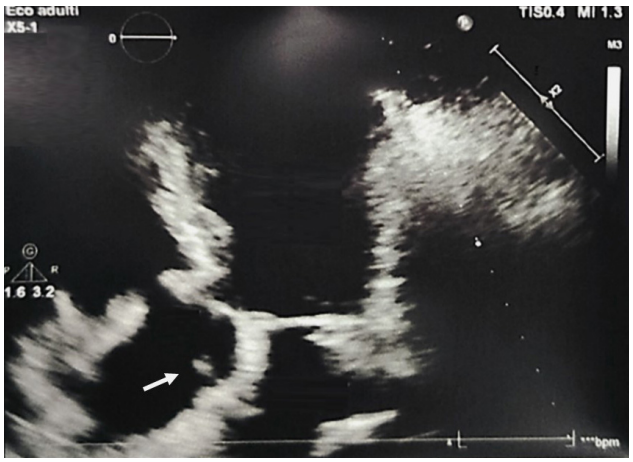
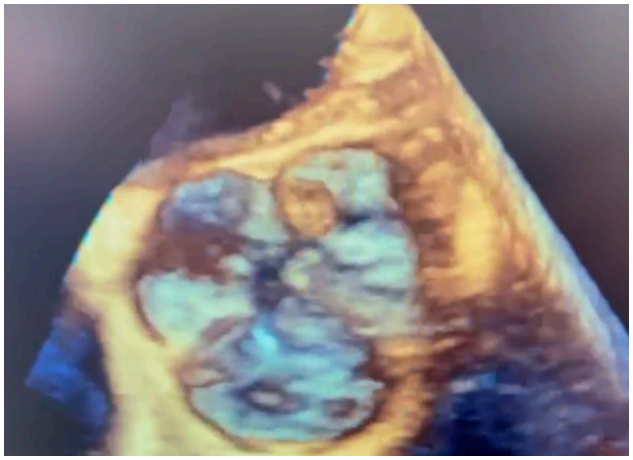


Figure 3. Two-dimensional transthoracic echocardiogram showing a small mobile mass on the left coronary cusp (5-chamber view).



Video 1. Transoesophageal echocardiogram. https://youtu.be/YZJCNDo5ZBE

with reciprocal ST depression in leads III, aVF and V1-V3 *(Fig. 1)*. The patient was promptly treated with intravenous bolus of heparin (5000 UI), 300 mg of acetylsalicylic acid, 180 mg of ticagrelor and transferred to the primary catheterization laboratory. Coronary angiography was performed, revealing occlusion of the diagonal branch of the left coronary artery *(Fig. 2)*. After direct aspiration thrombectomy, resolution of the thrombotic load was observed without the need for stenting. Later it was decided to complete tirofiban for 24 hours. Following coronary angiography, transthoracic echocardiogram showed normal left ventricular function, a normal pericardium, no valvular anomalies, and a suspicious mobile small mass on the left coronary cusp *(Fig. 3)*. Laboratory tests showed an increase in markers of myocardial cytonecrosis, and high inflammation indices (C-reactive protein and procalcitonin) *(Table 1)*. Considering the history of intermittent fever and gastrointestinal symptoms for 2 weeks and for the presence of high inflammatory indices and considering the embolic aetiology of acute coronary syndrome, blood cultures were obtained: Salmonella group bacteraemia was identified. Therefore, a transoesophageal echocardiogram was performed which confirmed the presence of a mobile, well-demarcated, echo-dense mass measuring 0.6 × 0.5 cm at the left coronary cusp *(Fig. 4, Video 1)*. Targeted antibiotic therapy was initiated with intravenous ceftriaxone (2 g/day) and ciprofloxacin (500 mg/bid). Following consultation with cardiothoracic surgery, the mass was surgically removed with optimal results *(Fig. 5)*. Surprisingly, a macroscopic morphological presentation of fibroelastoma was noted. Indeed, the pathology findings revealed a PFE

DISCUSSION

Cardiac PFEs are the second most prevalent primary cardiac tumours, exhibiting sporadic occurrence, with their exact aetiology still unknown[1]. These tumours predominantly manifest on the cardiac valves, with the aortic valve being the most commonly affected site[3]. Several factors are associated with an increased risk of developing PFE, including a history of endocardial surgery, rheumatic heart disease, endocarditis, and thoracic radiation treatment[4-5]. Despite their relatively benign nature, PFEs have been associated with embolic events, as evidenced by documented cases[5-7]. Embolization typically manifests in the cerebral, systemic, or coronary circulation[5]. PFEs located on the aortic valve and in the left atrium are associated with the highest rates of embolization[6-7].

Surgical resection of PFE is recommended for symptomatic patients and when the PFE demonstrates marked mobility for the potential embolic effect[5-6]. PFE may not be detected on a transthoracic echocardiogram. Therefore, inpatients with embolic events with no obvious evidence of embolic

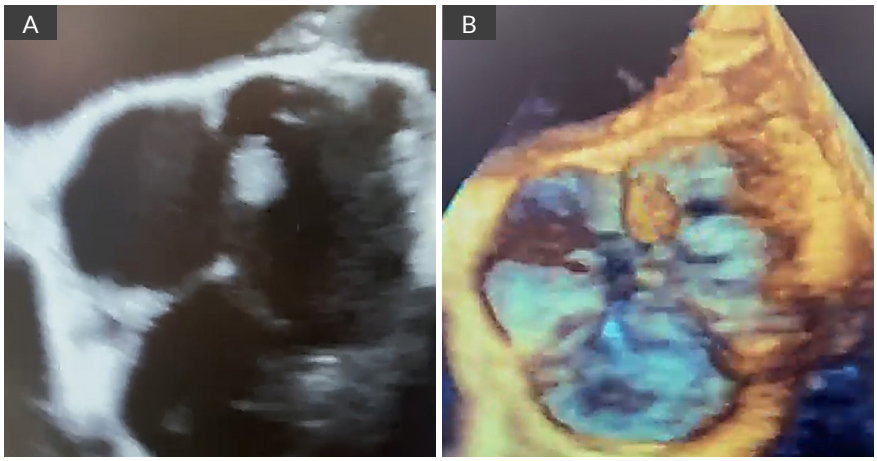


Figure 4. Transoesophageal echocardiogram: in short-axis view, a mobile, well-demarcated, echo-dense mass measuring 0.6 × 0.5 cm is evident on the left coronary cusp. A) Two-dimensional and B) three-dimensional echo imaging.

	Basal	2 Days	4 Days	6 Days	8 Days	10 Days
Troponin (mg/dl)	880	16000	6400	2300	990	344
CRP (mg/dl)	8.8	-	18	23	10	4
PCT (ng/dl)	0.54	-	2.2	-	-	0.2
Leucocytes (×103/µl)	8.20	-	17.10	-	12.40	10.20
Neutrophils (%)	65	-	80	-	69	-
Lymphocytes (%)	18	-	9	-	17	-

Abbreviations: CRP, C-reactive protein; PCT, procalcitonin.

Table 1. Laboratory test results: trend of inflammation, troponin, and blood count indices.

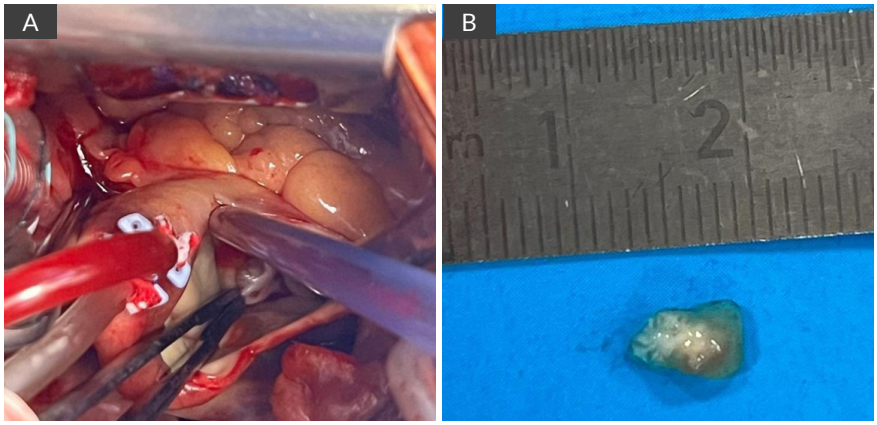


Figure 5. Intraoperative view. A) The moment of excision of the mass adhering to the aortic cusps and B) the specimen.

causes, transoesophageal echocardiogram should be performed to look for PFE[7].

We presented the case of a 63-year-old man who experienced coronary embolization. Notably, this patient exhibited complete blockage of the diagonal branch of the left coronary artery, without any indications of underlying coronary artery disease. Further investigation led to the identification of the cause of embolization. Therefore, transoesophageal echocardiogram was performed and confirmed the presence of a mobile, well-demarcated, echo-dense mass identified on the left coronary cusp. The concomitant presence of Salmonella bacteraemia, associated with symptoms of gastrointestinal inflammation and elevated inflammation indices, led to the hypothesis of valvular endocarditis. Salmonella endocarditis is a relatively rare condition, but several cases have been described, often presenting with embolic complications[8-9]. Despite the diagnostic uncertainty between fibroelastoma and endocarditic formation, the occurrence of the embolic event and the marked hypermobility we decided that prompt intervention was required. Surgical cleavage of the valve formation was then performed, revealing macroscopic morphological characteristics consistent with fibroelastoma, which was confirmed by the histological finding.

This case is especially interesting because PFE presented with acute coronary syndrome in a patient with concomitant Salmonella bacteraemia mimicking aortic endocarditis. A comprehensive diagnostic approach, including transoesophageal echocardiography and histopathological analysis of the mass is essential for a correct diagnosis.

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