Papillary fibroelastoma complicated by *Streptococcus sanguinis* bacteremia: a rare case of cardiac tumor with embolic events

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ABSTRACT

Fibroelastoma is a rare cardiac tumor that can cause embolization, stroke, myocardial infarction, heart failure, and cardiac arrest. Here, we report the case of a 45-year-old male who presented with right-sided weakness and fever. He was diagnosed with acute right frontal infarction and was found to have *Streptococcus sanguinis* bacteremia. Upon confirmation of a positive blood culture after 24 hours, treatment for endocarditis was initiated. Transesophageal echocardiography revealed findings highly suggestive of a papillary fibroelastoma (PFE). PFE ought to be regarded as a potential differential diagnosis in individuals who exhibit symptoms of fever, thromboembolism, and persistent bacteremia. Non-invasive imaging such as echocardiography is of great value in the diagnosis of PFE, while surgical resection remains the best treatment modality to overcome current and future associated complications.

KEYWORDS: papillary fibroelastoma; Streptococcus sanguinis; thromboembolic events

INTRODUCTION

Fibroelastomas are benign variants of cardiac tumors that are diagnosed incidentally in the vast majority of patients [1]. However, they can lead to serious medical conditions such as transient ischemic attack, stroke, myocardial infarction, syncope, and pulmonary embolism [2]. Pathological confirmation provides a conclusive diagnosis of fibroelastoma and superimposed infection [2]. However, transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are the most utilized techniques for diagnosis of PFE [3-6]. In this article, we present the case of an adult patient with papillary fibroelastoma (PFE) complicated by bacteremia and embolic events.

CASE PRESENTATION

We report the case of a 45-year-old male patient with a medical history of rheumatic fever 20 years ago, who presented with a new onset of slurred speech and right-sided weakness of 8-hour duration. He reported having a fever on and off for the last 2 months.

The patient had been admitted two months previously with fever and upper respiratory symptoms. At that time, the nasal swab for PCR SARS-CoV-2 was positive, and two sets of blood cultures grew *Streptococcus sanguinis* (sensitive

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to Ampicillin, Cephalosporins and Clindamycin), for which he received 2 weeks of intravenous ceftriaxone in addition to symptomatic treatment. Chest radiography revealed no consolidation, infiltration, or parenchyma-occupied lesions. Repeated blood culture 72 hours after the initial positive culture was negative. Our patient reported that, even after discharge, he experienced intermittent fever and did not recover completely.

Upon current presentation, he was febrile with a temperature of 38°C, hemodynamically stable with a BP of 140/80 mmHg, and a heart rate 105 beats/min. The patient was conscious, oriented, and cooperative. However, a neurological examination revealed weakness in the right upper and lower extremities and slurred speech. The left side was intact, gag reflex was present, and his face was not affected. Fundoscopic examination results were normal. A cardiac examination revealed a diastolic murmur.

CT tomography revealed faint right frontal suprainsular hypodensity, suggestive of an acute infarct. Laboratory tests showed elevated white blood cell count $18.0 \times 10^9/L$ (4-11x10⁹/L) with a significantly high C-reactive protein level of 136mg/L (<5mg/L) and normal liver and renal function tests. Chest X-ray and ECG were unremarkable. A septic workup, including blood and urine cultures, was performed. Ceftriaxone was administrated to the patient.

The patient underwent a comprehensive neurological evaluation by a neurologist who recommended the addition of aspirin. On the second day, MRI showed an acute infarction in the right frontal suprainsular region (cortical



and subcortical) and another small acute right frontal lacunar infarction (Fig. 1).

Twenty-four hours after presentation, two sets of blood cultures showed Gram-positive cocci. The identification of *Streptococcus sanguinis* was confirmed 24h later. The antibiogram demonstrated intermediate sensitivity to penicillin and adequate sensitivity to ceftriaxone.

Considering the previous history of rheumatic heart disease, diastolic murmur, fever, and bacteremia, infective endocarditis was on the top list of differential diagnoses. Therefore, transthoracic echocardiography (TTE) was performed. TTE showed a moderately dilated left ventricle, with an estimated LVEF of 45% and identified a small mobile mass attached to the anterior wall.

To further evaluate the cardiac findings, TEE was performed, which showed mild global LV dysfunction with a small mobile mass attached to the head of the papillary muscle (7×5 mm), consistent with papillary fibroelastoma,

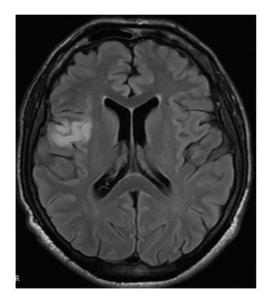


Fig. 1. MRI brain/T2 Flair showing acute infarction in the right frontal supra-insular region.

a mildly thickened aortic valve with severe aortic regurgitation, no vegetation on any cardiac valve, and no LAA thrombi (Fig. 2).

Along with antibiotics and anticoagulation, a multidisciplinary team, including a cardiothoracic surgical team, cardiologist, and infectious disease physician, recommended surgical intervention to remove the fibroelastoma and repair the aortic valve.

An additional workup was performed, including colonoscopy screening for bowel malignancy, in addition to cardiac catheterization. Ceftriaxone was continued for 4 weeks after documenting negative cultures.

Aortic valve repair and surgical resection of the tumor were planned; however, the patient declined surgical excision.

DISCUSSION

Papillary fibroelastomas (PFEs) are uncommon tumors that usually develop on the aortic valve [4,7]. However, PFEs were identified on other valves, papillary muscles, and ventricular walls as well. Most published cases have reported solitary tumors. A study that included 162 patients with PFEs reported that 45% (49/162) of the tumors were located on the aortic valve, most often on the right coronary cusp, followed by the noncoronary cusp, and least frequently on the left coronary cusp. Of the 40 tumors identified on the mitral valve, 23 were located on the anterior cusp and 17 on the posterior cusp. Furthermore, 32 were attached to the atrial surface, and some to the supporting apparatus [3]. Although rare, PFEs represent 10% of all primary cardiac tumors, and ranks second after cardiac myxomas [4].

Although most patients with PEFs are asymptomatic, thromboembolism is a serious complication related to PEFs. Other complications include myocardial infarction, stroke, and sudden death [7,8]. Cerebral arteries are the most common site of embolization, which subsequently manifests clinically as stroke [8]. Emboli can form as a result of tumor rupture, from a thrombus attached to it, or septic emboli due to an infected tumor.

To date, there is no clear explanation of the origin of fibroelastomas. However, endothelial damage with thrombus formation is considered a possible cause [5].

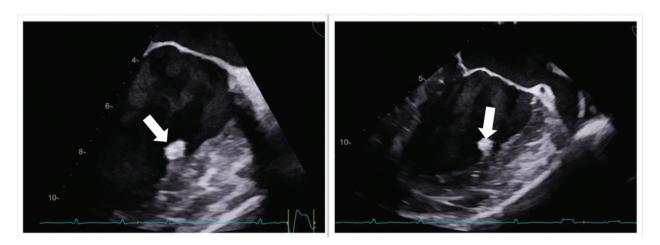


Fig. 2. Zoomed four chambers transesophageal echocardiology showing echogenic mobile mass attached to papillary muscle (fibroelastoma).

Our patient had a history of rheumatic fever, which could be linked to the development of PFE, as previously reported [5,9].

TTE and TEE are considered the main modalities for the detection and diagnosis of PFEs with sensitivities of 61.9% and 76.6% for PEFs <0,2 cm [3]. According to Klarish et al., PFEs are small in diameter (less than 1.5 cm), with a homogeneous speckled texture, mobile, and attached to the endocardium [5].

To avoid the formation of thrombi, the initiation of prophylactic intravenous anticoagulation is encouraged upon the diagnosis of PEFs [8]. Due to the high risk of embolic events, surgical removal is highly recommended for symptomatic patients [1,4].

Of particular interest, in this case, is the occurrence of *Streptococcus sanguinis* bacteremia concurrent to fibroelastoma, which could be misdiagnosed as infective endocarditis. *Streptococcus sanguinis* is a gram-positive, non-sporeforming, facultative anaerobe known as a colonizer of the oral cavity. It has been identified as main cause of infective endocarditis.

However, published data on bacteremia associated with fibroelastoma are limited. In 2002, Koji et al. reported the first case of an infected papillary fibroelastoma attached to the atrium. The case involved a 61-year-old female who presented with a fever for 2 months after dental extraction and was found to have *Streptococcus constellates* bacteremia. A left atrial tumor was seen on a 2-dimensional echocardiography. Surgical resection was done, and grampositive cocci were seen on microscopic examination of a thrombus on atrial fibroelastoma [10]. Another case reported by Faisal et al. described a young woman with a history of *Staphylococcus aureus* endocarditis, who was found to have a papillary fibroelastoma on the Chiari network [11].

CONCLUSION

The occurrence of infected fibroelastoma has been infrequently documented in the literature. Nevertheless, PFE ought to be regarded as a potential differential diagnosis in individuals who exhibit symptoms of fever, thromboembolism, and persistent bacteremia. Non-invasive imaging such as echocardiography is of great value in the diagnosis of PFE. Surgical resection remains the best treatment modality to overcome current and future associated complications.

Conflicts of Interest

The author(s) declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Informed Consent

Written informed consent was obtained from the patient. Available upon request.

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