Infected Papillary Fibroelastoma Attached to the Atrial Septum

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A 61-year-old woman had intermittent fever of 2 months’ duration following a dental extraction. On admission, her body temperature was 39.2°C. A mid-systolic murmur was heard at the apex on auscultation. A 2-dimensional echocardiogram revealed a mobile, heavy stick-like mass with vegetation (5.0×1.5 cm) attached to the left atrial septum. Multiple blood cultures grew Streptococcus constellatus. On diagnosis of an infected left atrial myxoma, antibiotics were administered daily and 4 weeks later, the left atrial tumor was resected. The tumor was 5.3 cm long, 1.5 cm in diameter at the inter-atrial wall and had vegetation on the free edge. On microscopic examination, colonies of Gram-positive cocci were found in the thrombus, on the papillary fibroelastoma. After treatment with antibiotics for a further 4 weeks, the patient was discharged. This is the first report of infected papillary fibroelastoma. (Circ J 2002; 66: 305 – 307)

Key Words:  Echocardiography; Infection; Left atrial tumor; Myxoma; Papillary fibroelastoma

Primary tumors of the heart are rare and are often classified histologically as ‘benign’. One-sixth of these are papillary fibroelastomas, which usually originate in the heart valves. A few previous reports have shown papillary fibroelastomas arising from the mural endocardium of the heart chambers, papillary fibroblastos attached to the atrial septum are extremely rare.

Case Report

A 61-year-old woman was in good health until 2 months prior to admission to the Yamamoto General Hospital in June 1998. On admission she complained of progressive fatigue. The week before, she had had intermittent fever peaking at 39.5°C and 2 months prior she had had a tooth extraction.

On physical examination, the patient was anxious, but did not appear to be ill. Her body temperature was 39.2°C, pulse rate was 84 beats/min, respiratory rate was 16 breaths/min and her blood pressure was 108/80 mmHg. The first and second heart sounds were normal with no gallop. A grade 2/6 mid-systolic murmur was heard at the apex. The lungs were clear on auscultation. The abdominal examination was unremarkable and the extremities showed no clubbing, edema, arthritis, Osler’s nodes, or Janeway lesions. Neurological examination was unremarkable. Funduscopic examination demonstrated 3 spots of retinal hemorrhage in the left eye.

Laboratory studies revealed the following results: leukocyte count 10,500/μl, hemoglobin 9.0 g/dl, hematocrit 27.8%, mean corpuscular volume (MCV) 82 fl, mean corpuscular hemoglobin (MCH) 26.5 pg, Fe 7 μg/dl, unsaturated iron binding capacity (UIBC) 207μg/dl, ferritin 210 ng/ml, C-reactive protein (CPR) 7.0 mg/dl, erythrocyte sedimentation rate (ESR) 80 mm/h, albumin 2.7 g/dl, aspartate aminotransferase (AST) 54 IU/L, alanine aminotransferase (ALT) 31 IU/L, lactate dehydrogenase (LDH) 515 IU/L, alkaline phosphatase (ALP) 591 IU/L and, γ-glutamyl transpeptidase (γ-GTP) 75 IU/L. Chest roentgenography, 12-lead electrocardiography, and urinalysis were normal.

Two-dimensional echocardiography showed a mobile, heavy stick-like mass attached to the atrial septum, in the left atrium during ventricular systole and passing freely into the left ventricle in diastole. The head of the mass had a high-density echo that was thought to be vegetation (Fig 1). Doppler examination demonstrated moderate mitral regurgitation.

Fig 1. Two-dimensional echocardiogram showing a heavy stick-like mass (5.0×1.5 cm: arrow) arising from the atrial septum (long axis view). The free edge of the mass has a high-density echo. Ao, aorta; LA, left atrial; LV, left ventricle.
Four days following the patient's admission, 3 sets of cultures of blood obtained on admission grew Streptococcus constellatus, so a diagnosis of infected left atrial myxoma was made and imipenem (1 g every 12 h) and gentamicin (40 mg every 8 h) were administered. The patient's body temperature varied from 37.0°C to 38.5°C, but returned to normal over the course of 3 weeks. The patient was then transferred to the Department of Thoracic Surgery Mie University.

Six days later, a pedunculated tumor, attached to the atrial septum at the fossa ovalis, was excised from the interatrial septum and the defect at the fossa ovalis was closed directly. The tumor was 5.3 cm long and 1.5 cm in diameter at the interatrial wall. It was a partially pinched, heavy stick-like mass with vegetation on the free edge (Fig 2). The mitral valve had a small scar in the cuspis posterior, but no procedures were performed because the valve function appeared acceptable.

Microscopically, the heavy stick-like tumor from the atrial septum consisted of a fibrous base with papillary structures (Fig 3A). Each papilla consisted of a central collagen core with a small amount of elastic fibers, surrounded by loose connective tissue and covered by a single layer of hyperplastic endothelial cells (Fig 3B, C). The papillary structure was obscured by large thrombi that were adhered to the head of the tumor. Gram-positive cocci were found in the thrombus (Fig 3D). A diagnosis of infected papillary fibroelastoma was made.

Following surgery, the patient was treated with imipenem and gentamicin for 2 weeks and imipenem alone for another 2 weeks. There was no complication during the postoperative period and she was discharged on the 43rd postoperative day.

Discussion

The prevalence of primary heart tumors ranges from 0.001% to 0.28% in autopsy studies;1 75% of these tumors are benign and 25% are malignant. The most common benign tumors are myxomas (30%), followed by lipomas (10%) and papillary fibroelastomas (9%).1 Papillary fibroelastomas usually occur in elderly patients even though some have been described in neonates with cardiac congenital abnormalities1,3 More than 90% of papillary fibroelastomas are solitary;1,4 approximately 90% occur on a valve, most commonly the aortic or mitral valves.1 When present on the atrioventricular valve, these tumors are usually found on the atrial side, but when present on semilunar valves, they can be found on either side.1,4,6 To our knowledge, only one case of papillary fibroelastoma associated with the left atrial septum has previously been reported.1 Papillary fibroelastomas are usually small, less than 1 cm in diameter, so the present case is rare at 5.3 cm long, although there is a previous report of a 5.7 cm tumor.8

Papillary fibroelastomas resemble a sea anemone, consisting of multiple papillary-like fronds arranged on a stalk. The fronds have a collagen core surrounded by elastic fibers and loose connective tissue, and are covered by endocardial cells. There is controversy about their incidence and there are several theories: hamartomas,1 neoplasms, congenital abnormalities,3 organized thrombi and giant Lamble’s excrescence.2,9 Lamble’s excrescence is fibrosis hyperplasia and age related, but there are differences between Lamble’s excrescence and this tumor: (1) Lamble’s excrescences do not occur on the arterial side of semilunar valves or on the mural endocardium, whereas papillary fibroelastomas do;1 and (2) microscopically, Lamble’s excrescences contain fibrin, whereas papillary fibroelastomas do not. The fronds of these papillary fibroelastomas are similar in structure to normal chorda tendinea and these tumors replicate all the components of normal endocardium, which suggests that papillary fibroelastoma is a hamartoma1.

The present patient’s cardiac tumor with vegetation was found by echocardiography and was diagnosed as an infected myxoma and treatment begun. Infected myxoma has been reported in 21 cases10–15 Shimizu et al reported in 1992 that 33% of infected myxomas was related to dental treatment,16 suggesting that the process of infected myxomas was similar to infectious endocarditis. A similar case of left atrial hematoma infected with bacteria has been reported.17 In the present case, the patient had had dental treatment before she developed a fever, which suggests that the dental treatment may have caused the systemic bacterial infection. To the best of our knowledge, this is the first

Fig 2. Gross specimen of the tumor showing a partially pinched heavy stick-like mass (5.3 cm long, 1.5 cm in diameter). The free edge of the tumor has vegetation.

Fig 3. (A) Gross appearance of the tumor (H&E). B: Microscopic view of the papillary fronds (H&E, ×10). (C) Central collagen core with elastic fibers (arrows) (Victoria blue-H&E, ×20). (D) Gram-positive cocci found in the thrombi (Gram-stain, oil-immersion, ×100).
reported case of an infected papillary fibroelastoma.

The majority of patients with papillary fibroelastomas have no symptoms, but these tumors have been associated with sudden death, myocardial infarction, and stroke. There were no subjective symptoms in the present case, other than the left eye cerebral hemorrhage; an eye ground embolism was originally suspected.

Although treatment of infected papillary fibroelastoma has not been reported, resection of the tumor is most important, according to the standard treatment of infectious endocarditis with vegetation. The present patient was given antimicrobial drugs and had the tumor resected, and was discharged without any further complications.

References