Papillary Fibroelastoma of the Aortic Valve: Report of a Case

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Abstract This report describes a case of aortic papillary fibroelastoma causing transient ischemic attack (TIA), and is followed by a review of the relevant literature. A tumor measuring 1.5 × 1.5 cm was located in the right coronary cusp of the aortic valve, which was found to have a “sea anemone-like” configuration due to collagenous or elastic papillary projections. Aortic fibroelastoma can cause cerebrovascular symptoms, and this patient presented with TIA. Moreover, aortic fibroelastoma usually causes chest symptoms and occasionally results in death despite its small size of generally less than 1 cm. According to the literature, all of the patients who died of fibroelastoma had an aortic valve tumor in the right or left coronary cusp, which indicates that the location of the tumor rather than its size may be an important factor contributing to the case of death. Surgical treatment is usually indicated especially for aortic fibroelastoma because of the high associated risk or cerebrovascular and chest diseases. The recent evolution of echocardiography will promote the chance of establishing a preoperative diagnosis of this lesion. However, because it is not possible to differentiate fibroelastoma from other lesions, including malignancies, by echocardiography alone, this may be another reason for performing surgical removal.

Key words Papillary fibroelastoma · Aortic valve · Right coronary cusp · Transient ischemic attack · Echocardiography

Introduction

Papillary fibroelastoma is a relatively rare benign tumor of the heart, originating in the endocardium, including the heart valves.¹ It is usually found incidentally at autopsy or surgery and is rarely symptomatic,¹⁻⁷ but can cause myocardial infarction, cerebral infarction, and systemic embolism, even in young patients,¹⁻⁸⁻¹⁴ and sometimes results in sudden death.¹,⁴,¹⁵⁻¹⁸ For this reason, surgical removal is necessary, despite the benign nature of the tumor.¹,¹⁹ Recently, an increasing number of fibroelastomas have been found preoperatively by using echocardiography. We report herein a case of aortic valve papillary fibroelastoma causing a transient ischemic attack (TIA), which was detected by echocardiography. A review of the literature on this entity is also presented.

Case Report

A 64-year-old woman was admitted to our University Hospital 3 days after losing consciousness. She had no relevant medical history. On admission, her blood pressure was 132/70, her heart rate was 60/min, and her body temperature was not elevated. There was no cardiac murmur and blood examination data were within the normal range. No abnormality was found on chest radiography or electrocardiogram. The initial diagnosis was TIA as no abnormal findings were detected by a brain computed tomography scan. A mass measuring 1.5 × 1.5 cm was found in the right coronary cusp of the aortic valve by transthoracic and transesophageal echocardiography (Fig. 1). Vegetation was suspected and excision of the lesion with valvuloplasty was performed.

The chest was opened through a median sternotomy. The heart was arrested under cardiopulmonary bypass and the ascending aorta was opened. A soft, friable mass was found attached to the edge of the right coronary cusp of the aortic valve. This was carefully shaved off the valve leaflet and on gross examination appeared to be similar to an atrial myxoma. A valvuloplasty was performed with 7-0 silk interrupted sutures. The patient

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was rewarmed and weaned from cardiopulmonary bypass without difficulty. Postoperative transesophageal echocardiography demonstrated a normal aortic valve without aortic regurgitation. The patient was transferred to the intensive care unit in a fairly stable condition and her postoperative outcome was uneventful.

**Pathological Findings**

The tumor was 1.5 × 1.5 cm in size and resembled a sea anemone macroscopically (Fig. 2). Microscopically, it was avascular and composed of numerous papillary or villous projections with hyalinized or myxoid stroma. The surface of the tumor was covered by endocardial cells. In the proximal part, the stroma contained a core consisting of dense collagenous and elastic fibers. The distal part of the projection was mainly formed by a myxoid matrix. Fibrin was not observed on the surface of the lesion (Fig. 3).

**Fig. 1.** Transesophageal echocardiography. An arrow indicates the tumor

**Fig. 2.** Macroscopic appearance of the tumor, which resembled a sea anemone

**Fig. 3.**  
(a) The tumor was composed of numerous papillary projections. Elastic fibers were observed in the proximal part of the projection (Elastica von Gieson).  
(b) Myxoid matrix was obvious in the distal part of the papillary projection and the surface was covered by endocardial cells (H&E)