Case report

Tricuspid valve papillary fibroelastoma: an unusual cause of intermittent dyspnea

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Abstract

A 67-year-old woman was referred for investigation of intermittent dyspnea, which was known for years but had been worsening over the previous 3 months. Two-dimensional echocardiography revealed the presence of a mass attached to the anterior leaflet of the tricuspid valve. The patient was successfully operated on to excise the right atrial mass and preserve the tricuspid apparatus. Morphological examination of the excised tissue led to the diagnosis of papillary fibroelastoma. Surgical treatment should be considered when such a tumor is diagnosed, even in asymptomatic patients, because of the possible risk of embolization. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Papillary fibroelastomas are rare, accounting for fewer than 10% of all primary cardiac tumors [1]. Most often they are asymptomatic, and detection is usually incidental during echocardiography, cardiac catheterization, cardiac operations, or at autopsy. Symptoms, if reported, are usually due to involvement of the left-sided valves; fibroelastomas of the right side of the heart are largely asymptomatic. We report a case of intermittent dyspnea found to be due to papillary fibroelastoma of the tricuspid valve.

2. Case report

A 67-year-old woman was referred for investigation of intermittent dyspnea, which was known for years but had been worsening over the previous 3 months. Her medical history was noncontributory, and the results of a physical examination and routine investigations, including a chest X-ray film and an electrocardiogram, were unremarkable. Two-dimensional echocardiography and transesophageal echocardiography (TEE) revealed normal-sized heart chambers, with mild tricuspid regurgitation (TR). A mobile solitary round mass, 2.5 × 1.6 cm, was attached by a short stalk to the anterior leaflet of the tricuspid valve, protruding into the right atrium (Fig. 1). The pedunculate mass oscillated with the flow of blood between the right atrium and the right ventricle. Pulmonary ventilation/perfusion scan was normal and excluded recurrent pulmonary emboli originating from this mass as a cause of the intermittent dyspnea. Pulmonary function test revealed obstructive pulmonary disease with partial reversion after bronchodilators, consistent with chronic bronchitis. Preoperative angiography revealed no coronary or carotid artery disease.

The patient underwent elective surgery. Cardiopulmonary bypass was performed under moderate hypothermia, using aortic and bicaval cannulation. After aortic cross-clamp arrest of the heart by cold antegrade crystalloid cardioplegia, and additional topical cooling using ice slush, the right atrium was opened. A single 2 × 2-cm shaggy, gelatinous mass (Fig. 2) was noted, attached to the atrial aspect of the anterior leaflet of the tricuspid valve by a short pedicle. The mass was removed, leaving the valve intact. There was no need for any surgical maneuver to repair the tricuspid valve. On examination, the tumor had a furry grayish-white surface. There was no thrombus. Grossly, the neoplasm resembled a sea anemone and was composed of many multiple papillary fronds.

Post-bypass TEE revealed mild TR but no evidence of any residual tumor. The postoperative period was uneventful, and the patient was given warfarin for 3 months. There were no further episodes of intermittent dyspnea during the 4 months of postoperative follow-up.

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Pathologic study showed that the specimen consisted of two fragments of delicate, white, myxoid tissue. Microscopically, polypoid projections lined by one layer of endothelial cells were noted. The projections were composed of fibrous tissue, which was partially hyalinized and partially myxoid. The features were typical of a papillary fibroelastoma.

3. Discussion

Primary cardiac tumors have an estimated incidence of 0.0017–0.33% in autopsy series and an estimated echocardiographic incidence of 0.019% in one clinical series [2]. Papillary fibroelastomas are the third most common of the primary cardiac tumors after atrial myxomas and lipomas, accounting for approximately 7% of cases [3,4]. Although they may be found anywhere in the heart, approximately 80% are found on the valvular endocardium. They represent the most common of the tumors of the heart valves accounting for 70–80% of patients [3–5]. In autopsy series, they are found with almost equal frequency on both sides of the heart and on all heart valves [6]. Most papillary fibroelastomas are connected to the underlying endothelium by a pedicle allowing for tumor mobility. The pedicle most often originates from the leaflets or from the valve apparatus. Rarely is a papillary fibroelastoma attached to the free wall of a cardiac chamber.

The etiology of papillary fibroelastoma is unknown. One explanation is that they result from mechanical trauma as they are more common in elderly patients with long-standing heart disease, including rheumatic disease, and are rarely seen in children [3]. Other theories suggest that they represent neoplasm, hamartomas, and inflammatory nodules [3].

Most often these tumors do not cause symptoms or produce physical findings. Those symptoms that do occur are seen with fibroelastomas of the left side of the heart and include angina, myocardial infarction, transient ischemic attacks, varying degrees of stroke, and even sudden cardiac death. Embolization of tumor fragments or of platelet fibrin clots forming on the surface of the tumor may be responsible for these events [1]. In contrast, fibroelastomas of the right side of the heart, of which tricuspid tumors are the most common, are less clearly related to symptoms. There have been far fewer reports of echocardiographic diagnosis in right-sided heart valve lesions [7]. It may be that the usually asymptomatic right-sided lesions do not necessitate medical attention and are thereby underreported.

Only a few reports have been published in which clinical signs and symptoms could actually be attributed to tricuspid fibroelastomas. These have a tumor ‘plop’ on auscultation [8], intermittent right ventricular outflow tract obstruction with cyanotic episodes [9], pulmonary embolization [10], and congestive heart failure [4].

A survey of the literature did not disclose any reports of tricuspid valve fibroelastoma in an otherwise healthy adult coming to a clinician’s attention solely due to intermittent dyspnea. In our case, when evaluated during surgery, the tumor seemed capable of almost entirely obstructing the right ventricular outflow tract, which could have caused intermittent obstruction to the pulmonary blood flow and episodes of dyspnea.

At present, conventional transthoracic echocardiography (TTE) and TEE with multiplanar studies are the best means of establishing diagnosis. Prophylactic anticoagulation is advised once the diagnosis of papillary fibroelastoma is suggested by echocardiography, and surgical removal with preservation of the inflicted valve apparatus is recommended even in asymptomatic patients so as to avoid the possible risk of recurrent or future embolism. Clinicians should be aware of intermittent dyspnea as a possible symp-
tom in the relatively uncommon condition of papillary fibroelastoma of the tricuspid valve. Recurrence after surgical excision is at yet unknown; however, careful follow-up is warranted.

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References