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Tricuspid Valve Papillary Fibroelastoma: A Rare Tumor with a Diagnostic Dilemma

Hairulfaizi Haron, MS, Mohd Rizal Mohd Yusof, MD, Oteh Maskon, MRCP, Joanna Ooi, SuMin, MMed, Mohd Ramzisham Abdul Rahman, MS

Heart and Lung Centre, Universiti Kebangsaaan Malaysia Medical Centre, Kuala Lumpur, Malaysia

ABSTRACT

Papillary fibroelastoma is a rare primary tumor of the heart valves. This lesion can occur on any of the valves or endothelial surface of the heart and has been detected by echocardiography, by cardiac catheterization, during open heart operations for other conditions, and at autopsy. Because of the potential for comorbidities, this tumor should be removed. We present the case of an elderly man with a diagnosis of severe mitral valve regurgitation and moderate tricuspid valve regurgitation who was suspected to have a tricuspid valve vegetation. Mitral valve replacement, tricuspid valve repair, and excision of the lesion were performed successfully. A histologic examination of the vegetation confirmed it to be a papillary fibroelastoma. We present this case to emphasize the rarity of this tumor and the importance of a correct diagnosis to avoid delaying its prompt and definitive management.

INTRODUCTION

Papillary fibroelastoma (PF) is a rare benign cardiac tumor with an incidence of less than 10% of all primary cardiac tumors. It is most commonly found on the valves, predominantly the aortic valve (44%), followed by the mitral (35%), tricuspid (15%), and pulmonary (8%) valves [Shimode 2007]. The tumor infrequently occurs on the left atrium, atrial septum, right atrium, eustachian valve, right ventricle, and left ventricle [Shimode 2007]. PF is usually asymptomatic and is found incidentally during routine echocardiography or at autopsy. Surgical resection is the treatment of choice owing to the potential for complications. We share this case in view of its rarity and highlight the importance of an accurate preoperative diagnosis to avoid an unnecessary delay in its curative treatment.

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Correspondence: Professor Dr. Mohd Ramzisham Abdul Rahman, Division of Cardiothoracic Surgery, Department of Surgery, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob, Latif, Cheras, 56000 Kuala Lumpur, Malaysia; 60391456201/2; fax: +60391456684 (e-mail: ramzisham@hotmail.com).

CASE REPORT

A 68-year-old man with a background of hypertension and diabetes mellitus was referred for both severe mitral valve regurgitation and moderate tricuspid valve regurgitation secondary to chronic rheumatic heart disease. A detail

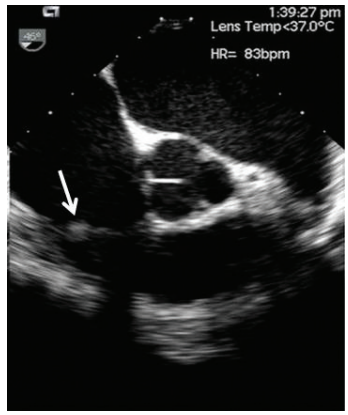

Figure 1. Transesophageal feature of the papillary fibroelastoma on the anterior leaflet of the tricuspid valve (arrow).

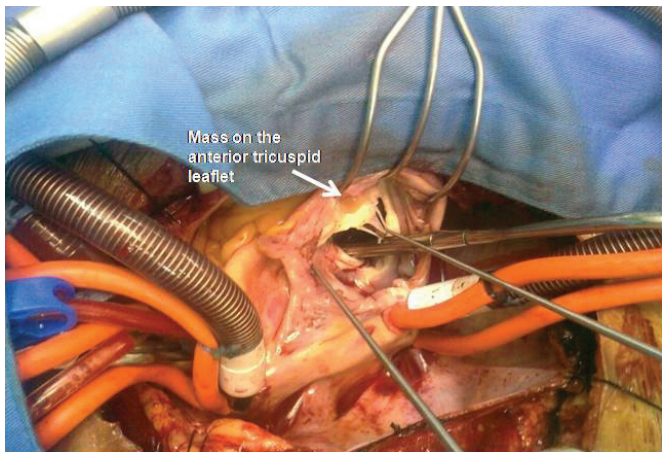

Figure 2. Intraoperative view through the right atrium of the papillary fibroelastoma on the anterior leaflet of the tricuspid valve.

preoperative transesophageal echocardiography (TEE) evaluation revealed a lesion of 1 cm × 1 cm adhering to the anterior leaflet of the tricuspid valve (Figure 1). Because the patient had a diagnosis of chronic rheumatic heart disease and presented with a history of nonspecific fever, we suspected a tricuspid vegetation. Despite negative results for a blood culture, our patient was treated with a broad-spectrum antibiotic prior to the valvular surgery. The patient's fever and general condition improved clinically; however, the lesion remained the same after approximately 6 weeks of antibiotic therapy. He eventually underwent successful mitral valve replacement and tricuspid annuloplasty, along with resection of the lesion. Intraoperatively, we found the suspected vegetation to be a firm yellowy gelatinous lesion adhering to the atrial aspect of the anterior tricuspid leaflet (Figure 2). A histopathologic examination of the lesion showed avascular papillae composed of collagen and covered by an endothelial lining, which confirmed the diagnosis of PF. The patient was discharged well 6 days following surgery with no significant postoperative complications; no recurrence of a similar lesion was found at the 1-year follow-up examination.

DISCUSSION

The diagnosis of PF is important because it represents a surgically correctable cause of such morbidities as stroke, myocardial infarction, and pulmonary embolism. Compared with lesions on the right side, which are usually asymptomatic, left-sided lesions are more common and more likely to cause complications [Wolfe 1991; Georghiou 2003]. Because of the potential for complications, early surgical resection is advocated for such lesions [McFadden 1987]. Because our patient was very ill with fever and heart failure when the diagnosis was made, the surgery was delayed. This delay was mainly because the surgery was deemed to be of high risk given the patient's medical condition at that time. Furthermore, the right-sided vegetation was not considered as serious as one on the left side. Optimizing the patient's overall condition while preparing him for surgery with antibiotics and anti–heart failure treatment for a few weeks seemed to be the best option at the time.

The diagnosis of PF can be obtained only if one has a high index of suspicion for such a rare lesion. The diagnosis can readily be achieved, however, by using echocardiography. A mobile, finger-like appearance and attachment to the valve by a small stalk has been reported to be a typical echocardiographic feature of PF [Yee 1997]. The use of TEE preoperatively can add more information by confirming the presence of the tumor and determining its location. TEE provides better resolution, which permits location of the central collagenous core of the tumor and distinguishing it from the rest of the mass by its bright echocardiographic appearance [Yee 1997]. If the mass is clearly identified, this central echodense contrast should allow this tumor to be differentiated from other intracardiac masses, especially myxomas, vegetations, mural thrombus, valve calcifications, and Lambl excrescences. Despite this ability, the typical appearance is rare, and there has been an atypical presentation of PF mimicking vegetations in suspected cases of subacute bacterial endocarditis [Lee 1993], such as in our case.

Because the PF was mistaken as an infective vegetation and because of the patient's sepsis and heart failure condition, we delayed prompt surgical treatment of the valves and the lesion. Fortunately, none of the complications described by previous authors arose. As in our case, we believe that diagnosis can be more accurate and that unnecessary worries can be avoided if there is a high index of suspicion for this relatively rare tumor, especially if the patient has a nonspecific history of pyrexia with negative blood cultures. We also support the use of preoperative TEE in every case because of its superiority to transthoracic echocardiography for the study of any suspected lesions in greater detail.

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<<AUTHORS: Please confirm that author names are presented correctly.>>