Anadolu Kardiyol Derg
2008; 8: 306-14

also a publication indicating that a good tomography interpretation is often adequate in detecting pulmonary artery agenesis (6).

In a literature publication from 1982; a series comprised of 19 cases was examined and ligation was performed on two patients because of frequent hemoptysis attacks (7). In a case report in 1993; pulmonary hypertension was detected in two cases from a series comprised of 4 cases and both of them died in two years. The most important complications affecting the survivals of these cases were; pulmonary hypertension and frequent infections.

In a literature publication from 2004, the average age of the eight cases detected was 11, and four of them were followed up until adult years and no complications were seen (8). One case of pulmonary artery agenesis with stenosis of the main bronchus that was diagnosed after being admitted to the out-patient clinic with asthma-like attacks was also reported (9). A patient with pulmonary artery agenesis who has shortness of breath and pleuritic chest pain was admitted with the classical symptomatology of the condition and diagnosed (10).

Our case had not shown any symptoms previously either and was diagnosed with this condition after the age of 20 upon shortness of breath with effort. Since the survival rates of patients diagnosed at younger ages are increased due to operations for complications, we may suggest that our case's not being symptomatic until his adult age will not affect his survival rate.

Conclusion

We wanted to present our case because it was an isolated case, the condition could be observed less frequently in the left pulmonary artery, it was detected at a rather late age, it did not have pulmonary hypertension, it did not follow a course with complications and it did not have any additional cardiac anomaly.

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Fibroelastoma of the posterior mitral leaflet

Posteriyor mitral yaprakçıkta fibroelastoma

Kaan İnan, Alper Uçak, Onur Selçuk Göksel, Veysel Temizkan, Murat Uğur, Eralp Ulusoy, Melih Hulusi Us, Ahmet Turan Yılmaz*

From Clinics of Cardiovascular Surgery and *Cardiology, GATA Haydarpaşa Education Hospital, İstanbul, Turkey

Introduction

A papillary fibroelastoma is a very uncommon primary tumor of the heart with benign pathology. The tumor is generally diagnosed accidentally during echocardiography or following the investigation of a thromboembolic patient (1, 2). Valvular involvement includes aortic and mitral valves. Most of the scarce data have been reported from patients older than 50 years old, it may be found at any age, from the neonate to the nonagenarian (2). We present a rare case of papillary fibroelastoma of the posterior mitral leaflet in a 52- year-old patient.

Case report

A 52-year-old lady was referred to our clinic with palpitation and dyspnea and a mobile mass was demonstrated with transthoracic echocardiography on the posterior leaflet of the mitral valve. She had no prior history of a thromboembolic event. The transesophageal echocardiography (TEE) demonstrated a sessile tumor 10 mm in diameter and mild mitral regurgitation (Fig 1A. and 1B). It is noteworthy that the mitral annular diameter was 26 mm on echocardiography. The surgical removal of the tumor was planned. Following standard

Address for Correspondence/Yazışma Adresi: : Dr. Murat Uğur, GATA Haydarpaşa Eğitim Hastanesi Kalp Damar Cerrahisi Kliniği, Üsküdar / İstanbul, Türkiye
Mobile: +90 532 570 89 18 Fax: +90 216 348 78 80 E-mail: drmugur@gmail.com

Note: This case report was presented at the 3rd News in Cardiology and Cardiovascular Surgery Congress, 28 November- 2 December, 2007, Antalya, Turkey

sternotomy and cardiopulmonary bypass with bicaval cannulation, 10x8x6 mm solitary verrucous tumor was seen on the posterior mitral leaflet through a transseptal approach (Fig 2A). Mitral annulus diameter was about 25 mm with similar finding of the echocardiography. The tumor invade into the leaflet tissue with involvement of some subvalvular apparatus. A quadrangular resection with plication of the posterior annulus was performed following complete excision of the tumor with the involved leaflet segment and chordae. A cystic area around the solid tumor was aspirated for microbiologic and pathologic investigation in addition to the primary tumor. Intraoperative TEE showed moderate mitral regurgitation with systolic anterior motion of the anterior leaflet causing a mean gradient of 30 mmHg in the left ventricular outflow tract (LVOT). Due to this LVOT obstruction we replaced the valve with posterior annular augmentation using Teflon felts (Fig 2B). The patient weaned off cardiopulmonary bypass easily following replacement with a mechanical prosthetic valve and TEE demonstrated functioning valve and the absence of gradient on left ventricular outflow tract. The result of pathologic examination was; papillary fibroelastoma and seroma around it (Fig 3A and Fig 3B). After an uneventful postoperative period, the patient was discharged from hospital at 7th postoperative day.

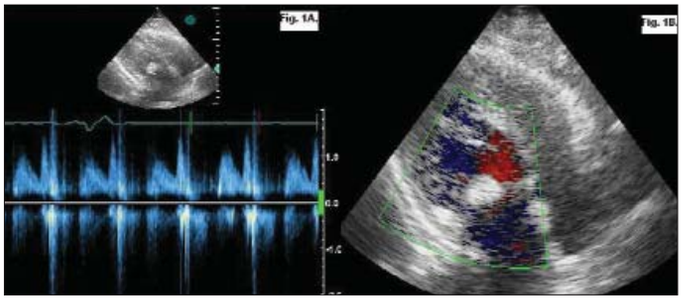

Figure 1. Echocardiographic images of the intracardiac tumor.

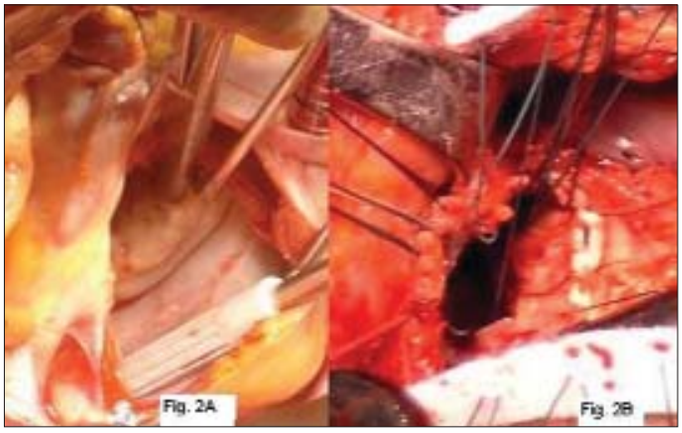

Figure 2. Tumor on the mid-posterior mitral leaflet (A). Augmentation of the posterior mitral annulus with Teflon felt strips from two sides for valve replacement (B).

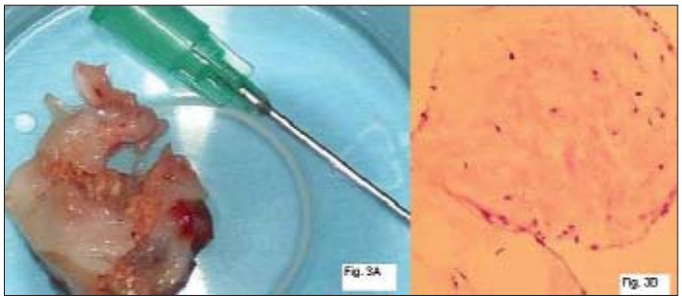

Figure 3. Gross appearance of the tumor excised with the leaflet tissue involved (A). Hematoxylin and Eosin (40X) staining of the excised mass demonstrating an elastic core surrounded by with a single layer of endothelium and papillary lesion (B).

Discussion

Cardiac papillary fibroelastoma is a rare benign tumor that involves most commonly heart valves and may cause thromboembolism or mechanical interference with the valvular function. Nearly 10% of the all cardiac tumors have been estimated to be papillary fibroelastoma (1, 3) Diagnosis is usually adequate even in the asymptomatic patient as an indication for surgery in spite of benign nature of the lesion.

It usually involves left-sided cardiac valves; commonly aortic and anterior mitral leaflet respectively. Involvement of the posterior leaflet is even more uncommon. However, a simple tumor shave excision off the leaflet was adequate more commonly in aortic valve lesions and valve replacement, mostly in mitral position with the subvalvular involvement, was required in 6% of the patients. In our patient, tumor involved valvular tissue in mid-posterior leaflet and some chordae. Primary goal was to repair the valve as no local recurrences have been reported after complete excision. Although unfavorable hemodynamics is very uncommon due to small anatomical annulus of the patient, consequently, replacement with a prosthetic mechanical valve was performed. In authors' current practice, valve replacements are performed exclusively with valve-preservation techniques; however, posterior leaflet preservation was not possible in this case due to small annulus as well as excision of some of the leaflet tissue in addition to tumor alone. Therefore, a posterior augmentation technique with two Teflon felt strips was adapted to reinforce posterior annulus (Fig 2B).

Papillary fibroelastoma can be radiopaque as in our previous experience depending on the histopathologic content of the tumor (4). Myxoid or calcific degeneration is consistent with the rare incidence of accidental visibility during coronary angiography. Tumor was not radiopaque in our patient and the degenerative cystic fluid aspirated around the verrucous mass (Fig 2B).

Conclusion

Irrespective of the presence of symptoms, prompt excision with preservation of the valvular anatomy and function whenever possible is the primary goal of the surgical treatment. Valve replacement is also safe and effective treatment option for selected cases.

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