A RARE CASE REPORT: PAPILLARY FIBROELASTOMA OF THE AORTIC VALVE IN THE EMERGENCY INSTITUTE FOR CARDIOVASCULAR DISEASES AND TRANSPLANTATION TG-MUREŞ

MATHE ZSOMBOR[1], RADULY GERGO[2], COSMIN BANCEU[3], VALENTIN IONUȚ STROE[4], ALEXANDRU INCZE[5]

[1,2,3,4]Phd Student, University of Medicine and Pharmacy Tg-Mureş, [5]University of Medicine and Pharmacy Tg-Mureş

Keywords: *aortic valve, papillary fibroelastoma, echocardiography*

Abstract: *Papillary fibroelastoma is the third most common primary tumor of the heart after cardiac myxomas and cardiac lipomas. Cardiac myxomas are six times more frequent than papillary fibroelastoma. We report a rare case of a young, 44 year-old patient with papillary fibroelastoma which was found incidentally on ecocardiography. The patient was without any other symptoms. Upon surgery, a tumor of 15 mm was revealed on the right coronary artery aortic cusp. The tumor was surgically resected. After surgery, in the second postoperative day the patient presented rhythm disturbances with atrial fibrillation. This was treated with specific drugs and after 24 hours the patient's rhythm was regular. Six days later, the patient was discharged. The main histological findings show a fibroelastoma. As a conclusion, early diagnosis of this rare tumor is very important because cerebrovascular and other embolic complications can be prevented.*

INTRODUCTION

Primary cardiac tumors are usually rare, but are a real challenge for a cardiac surgeon. Papillary fibroelastoma is the third most common primary tumor of the heart. The other two tumors are cardiac myxomas and cardiac lipomas. Cardiac myxomas are six times more frequent than papillary fibroelastoma. Until the second half of the century, cardiac tumors were diagnosed almost exclusively at autopsy. Nowadays, papillary fibroelastomas are recognised more frequently than in the past because of advances in imaging technology. Most of these tumors are discovered incidentally in many cases by transthoracical echocardiography, but using of transesophageal echocardiography is much better to evaluate the structural and functional anatomy of the aortic valve. Although patients with cardiac tumors may present with cardiovascular symptoms, the diagnosis is frequently incidental during an imaging examination performed for a different indication. Incidental masses in the heart are more likely to be thrombi or vegetations for which clinical correlation is paramount. Should the mass represent a primary cardiac tumor, it is likely to be benign, with myxoma being the most common primary cardiac tumor in adults and rhabdomyoma the most common in children. Primary malignant cardiac tumors are extremely rare, only 25% of the total cardiac tumors, and they are generally a variety of sarcoma.

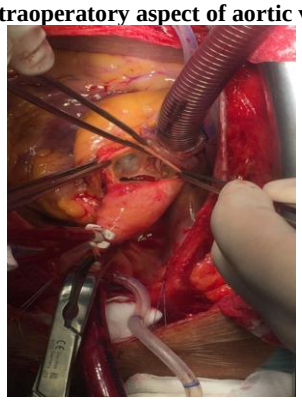
AIM

The aim of this article was to present a rare case of a young patient with cardiac benign tumor, discovered incidentally, during routine transthoracical ecocarddiography and to speak about the right strategy for managing these kind of tumors especially when they are incidentally discovered

CASE REPORT

In September 2018, a 44-year-young male was scheduled for surgical treatment in the Emergency Institute for Cardiovascular Diseases and Transplantation. The patient was without medical history and was of course totally asymptomatic. He was diagnosed incidentally, after a transthoracic echocardiography. Main echocardiography findings were: a tumor of aproximately 7 mm on the right coronary aortic cusp. At that moment we could not know exactly what kind of tumor it was. A coronary angiogram was also performed, which excluded the coronary stenosis. Preoperative ecocardiography revealed a left ventricular ejection fraction of 60% and no significant valvular abnormalities. The patient was scheduled for open heart surgery with sternotomy. After the induction in general anesthesia, we performed sternotomy, followed by pericardiotomy. After systemic heparinisation, the extracorporeal circuit was prepared. Cardioplegia called Calafiore was administrated with warm blood, of course after aortic crossclamping. At inspection, the aortic valve was without any insufficiency buton the ventricular part of the aortic valve adhering to the right coronary aortic cusp a 13 mm tumor was identified (figure no. 1). This 13 mm mass was mobile and pedunculated (figure no. 2).

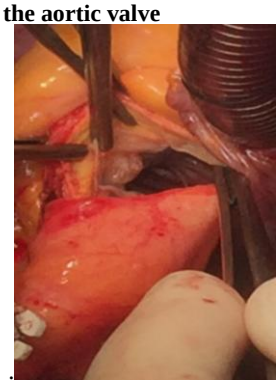
Figure no. 1. Intraoperatory aspect of aortic valve



[2]Corresponding author: Raduly Gergo, Str. Gheorghe Marinescu, Nr. 38, Târgu-Mureş, România, E-mail raduly_gergo@yahoo.com, Phone: +40747 714387

Article received on 20.07.2018 and accepted for publication on 22.11.2018
ACTA MEDICA TRANSILVANICA December 2018;23(4):44-45

Figure no. 2. Intraoperatory aspect of papillary fibroelastoma of the aortic valve



It was inserted in the aortic annulus and in the right coronary aortic cusp. We reduced the patient's temperature at 33 C degrees and excised the tumor with the aortic valve. The aortic valve was replaced with an aortic bileaflet St-Jude Medical prosthetic valve. Upon visual inspection, the resected tumor had negative margins, which was also confirmed by histopathologic analysis. The aortotomy was closed in two layers. After de-airing maneuvers the aortic-clamp was released and the patient's body was rewarmed. The total time of aortic-clamping was 42 minutes. Finally we have successfully weaned the patient from cardiopulmonary bypass. The patient did not require inotropic support. After 24 hours of stay in the ICU he presented atrial fibrillation in the second postoperative day, mainly due to low levels of K. Treatment with amiodarone i.v. was started. After the third postoperative day the patient was stable, in sinus rhytm. The patient was discharged from the hospital after six postoperative days.

DISCUSSIONS

The papillary fibroelastoma is the third most common benign heart tumor. It is mainly localized on the valvular apparatus. The places where we can usually find this kind of tumors are the aortic valve, the left atrium, interatrial septum, left atrium and right ventricle.(1,2) This benign tumor is diagnosed incidentally by echocardiography, during surgery or necropsy. Its size varies from 9 to 12 mm, but it may have the tendency to grow. On echocardiography it appears pedunculated and mobile, with speckled patterns and stippling along the edges.(3) It has papillar aspect, formed from collagen, elastic fibres and proteoglicans and has two layers: an outer, hyperplastic endotelial layer and a dense central core, its surface being covered with filiform projections.(4) The patient may be asymptomatic, but cardioembolic events may happen: stroke, acute valvular dysfunction ,embolism, ventricular fibrillation and sudden death (5) as most of the fibroelastomas are located in the left heart, on the valvular or mural side of the endocardium. It can occlude the coronary opening causing myocardial infarction, angina or ventricular tachycardia.

The management of the papilary fibroelastoma is surgical resection, but it depends on the tumor's clinical presentation. As discussed above, it may cause cardioembolic events or occlude the coronary arteries. Cerebrovascular embolism has been associated with the smallest tumors (diameters as small as 3 mm).(6) Smaller diameters may present asymptomatic patients who require periodic imagistic follow-up.(7) Prophylactic anticoagulation therapy should be initiated until surgical resection is accomplished. The optimal surgical procedure is valve-sparing resection for pedunculated tumors and valve resection in case of underlying degeneration or extensive destruction of the native valvular apparatus.(8)

The particularity of the case is that it is difficult to make the difference between a tumor and infectious endocarditis, the patient being firstly discovered and suspected with vegetant endocarditis. Although many authors claim that the papillar fibroelastoma is an organized thrombus rather than a tumor, it has a friable structure, and may be the source of cardioembolic events.(9)

CONCLUSIONS

Early diagnosis of cardiac tumors is very important. The management of papillary fibroelastoma depends upon its clinical presentation. Patients who are experiencing some embolic events undergo surgical therapy. In some cases, papillary fibroelastomas of the aortic valve have been associated with syncope or acute myocardial infarction. This is mainly due to total obstruction of the coronary ostia. This is the reason why in some patients with papillary fibroelastoma after sudden death of them we found exactly only after autopsy the cause of death. Aortic valve replacement is generally not necessary for papillary fibroelastomas unless there is underlying degeneration or extensive destruction of the native aortic valve. In some older patients with lots of comorbidities partial valve sparing procedure is an option for cardiac surgeon. Regrowth of the tumor after partial tumor resection is also an important fact especially in younger patients. This is the reason why in our case we replace the aortic valve with an prosthetic valve after resection of the papillary fibroelastoma. Although papillary fibroelastoma is benign, it is increasingly considered a matter for surgery because of such potential complications as stroke, acute myocardial infarction, ventricular fibrillation and sudden death. Surgical resection is strongly advised if any of the following situations is present: pedunculated lesions or tumor mobility. We recommend prompt transesophageal echocardiography, anticoagulation therapy to guard against surface thrombi and of course, which is the most important thing in a case with papillary fibroelastoma of aortic valve, early surgical referral.

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