

**CASE REPORT**

Surgical Treatment of Aortic Valve Fibroelastoma: a Case Report

Stamenko Šušak^{1,2}, Bogdan Okiljević^{1,2}, Strahinja Mrvić¹, Jovan Rajić¹, Ranko Zdravković¹

ABSTRACT

A 45-year-old man was admitted to Clinic for the first time, with symptoms of chest pain and fatigue. Computerised tomography (CT) diagnostics of the chest showed a soft tissue vegetation of approximately 5x5 mm on the left aortic coronary cusp. A double-vessel coronary disease was also diagnosed. The patient underwent surgery, a complete resection of the tumour was achieved, which was confirmed by postoperative transoesophageal echocardiography (TEE). Because of the risk of valve damage, it was decided to replace the aortic valve. A bypass from left internal mammary artery (LIMA) to left anterior descending (LAD) coronary artery (LAD-LIMA) and right coronary artery (RCA) was also performed. The patient was discharged on the 14th postoperative day with satisfactory results.

Key words: fibroelastoma, aortic valve, tumour, cardiac surgery.

(1) Clinic of Cardiovascular Surgery, Institute of Cardiovascular Diseases Vojvodina, Sremska Kamenica, Serbia.

(2) Faculty of Medicine, University of Novi Sad, Novi Sad, Serbia.

Correspondence:

BOGDAN OKILJEVIĆ

E: bogdan.okiljevic@mf.uns.ac.rs

M: +381 64 102 96 53

ARTICLE INFO

Received: 12 July 2019

Revision received: 22 July 2019

Accepted: 26 July 2019

INTRODUCTION

Primary intracardiac tumours are not frequent findings. Their prevalence is below 0.3%. Intracardiac tumours develop mostly from the endocardium, rarely from the cardiac muscle or pericardium. The largest part of these tumours is benign (approximately 75%). However, their true incidence can be only assumed, because they are frequently asymptomatic.¹

Papillary fibroelastoma is a benign tumour which originates from endocardium. Share of papillary fibroelastomas in all cardiac tumours is less than 10%. Besides, papillary fibroelastoma is the second most common cardiac benign tumour and the most common tumour of the cardiac valves. This tumour can have a wide spectrum of clinical presentations. It can be asymptomatic or can cause severe embolic complications (eg stroke, myocardial infarction). Cardiac fibroelastomas are most often diagnosed in people between the 4th and 8th decade of life and it is more often seen in men. They are mostly slowly growing tumours with multiple vascular papillary fronds, which are made of proteoglycans, elastic fibres and a lot of collagen. Macroscopically, they are

small and friable masses. Their pathogenesis is unknown to this moment. Although benign, papillary fibroelastomas can cause life-threatening complications. The risk of complications can be assessed in dependence on their location, mobility and dimension.²⁻⁴

CASE HISTORY

A 43-year-old man started feeling chest pain and fatigue symptoms on exertion five years ago. Symptoms intensified in the last two months. Nine years ago, the patient was treated for cardiac arrhythmia caused by adenovirus. Chest X-ray and blood laboratory tests were normal. Coronary angiography showed 80% stenosis in the mid-left anterior descending coronary artery (LAD), 45% stenosis in the circumflex (Cx) artery and 50% stenosis in the right coronary artery (RCA). CT diagnostics of the chest showed a soft tissue vegetation of approximately 5x5 mm on the left aortic coronary cusp. Based on the above findings, a differential diagnosis was

made, and it included a thrombus, myxoma, fibroelastoma and inflammatory mass. Based on the potential embolic risk, either of the mass itself or of associated thrombus and the possibility of further enlargement, symptoms at the time of diagnosis and the findings of coronary angiography, the patient was referred for surgical excision of the mass and revascularisation.

The operation was conducted on extracorporeal circulation (ECC) with the ascending aorta and the right cavoatrial cannulation. The vent was placed in the upper-right pulmonary vein, to secure the suction of the left atrium cavity. After starting ECC, aortic clamping was performed on the ascending aorta. Cold blood cardioplegic solution was injected through coronary ostia. After achieving cardiac arrest, transversal aortotomy was performed. After the aortotomy, on the left aortic cusp, a pedunculated flower-like tumour was found (Figure 1). With simple shave, the excision of the tumour was undertaken with particular care to avoid embolisation and ensure that no remnants from fragmentation of this friable tumour were left behind both locally on the cusp and near the ascending aorta and left ventricle (Figure 2). A complete resection of the tumour was achieved. The aortic valve was three-leaflet, fibrously modified and of thin leaf-



Figure 1: Tumour on the left aortic cusp



Figure 2: Tumour after resection



Figure 3: Tumour and aortic cusps after resection

lets. Because of the risk of valve damage, it was decided to replace the aortic valve (Figure 3). Mechanical valve No. 23 was implanted. Also, a bypass to LAD branch with left internal mammary artery (LIMA) and saphenous vein graft on RCA was performed. Total cardiac arrest time was 87 minutes, and ECC time was 100 minutes. The lesion was histologically diagnosed as a papillary fibroelastoma. After 14 days of postoperative hospitalisation, the patient was discharged with optimal results of surgical treatment.

DISCUSSION

Papillary fibroelastomas really are primary benign endocardial tumours. They originate from fibrous tissue, elastic fibres or smooth muscle cells - component that are otherwise normal constituents of endocardium. These tumours look like sea anemone - they are on short pedicle and they have multiple papillary fronds.¹ Usually papillary fibroelastoma originate from the valvular endocardium (85%). It is mostly seen on the aortic (29%) and mitral valves (25%), not so often on tricuspid (17%) and pulmonary valves (13%).⁵ There are several hypotheses about the development of cardiac fibroelastoma, but without unequivocally scientific evidence. The most accepted hypothesis is the microthrombus theory, which assumes that the lesions are acquired and that they arise as small thrombi on the coapting margins of the valves as a result of minor endothelial damage. Fibroelastomas are usually small tumours (with average diameter 9 to 12 mm). Although histologically benign, complications as acute valvular dysfunction, ventricular fibrillation, embolism, stroke and sudden death can be consequences of this type of tumours.⁴ The diagnosis can be made by trans-

thoracic or transoesophageal echocardiography (TEE). Additional diagnostic methods, as 3D echocardiography, cardiac magnetic resonance and multi-slice spiral computed tomography, can be used for more precise identification of the tumour.⁵

The question arises, when to treat such tumours. It depends on presence of symptoms and on risk of life-threatening complications. There is an agreement that all symptomatic patients should be surgically treated. Although asymptomatic, patients with large (>1 cm) left sided mobile masses, as in this case, should also be treated surgically, because of the danger of life-threatening complications. Patients who do not meet these criteria are being closely followed up echocardiographically.¹ Early anticoagulation is mandatory in the management of these tu-

mours. The surgical excision of tumour requires extracorporeal circulation, induced cardiac arrest and an access to the valve from which the tumour originates. The papillary fibroelastoma can be easily removed without a valve damage. Fragmentation of the tumour tissue can occur and therefore special care should be taken to avoid it. When there is a resultant valve defect after tumour excision, valve should be repaired or replaced, depending on institutional preference.⁵

The study of Gopaldas and colleagues described a case of a patient with fibroelastoma and elective coronary artery bypass grafting.⁴ In this case, the valve was not replaced, and only tumour removal was done. In the present case it was decided to perform valve replacement due to the changes described in the aortic cusps.

CONCLUSION

Fibroelastoma can cause a variety of symptoms or can be asymptomatic. Several authors stated that after the diagnosis, it is necessary to operate all symptomatic and asymptomatic tumours, except the asymptomatic ones with the tumour size less than 1 cm. Such tumours should be

monitored and checked regularly. The patient described in this paper had symptoms, which may have been the result of coronary artery disease, and even in that case the decision was made to remove the tumour of 5x5 mm in size.

ACKNOWLEDGEMENTS

None.

CONFLICT OF INTEREST

None.

REFERENCES

1. Parthenakis F, Nyktari E, Patrianakos A, Pitsis A, Asimaki A, Vardas P. Asymptomatic papillary fibroelastoma of the aortic valve in a young woman - a case report. *Cardiovasc Ultrasound* 2009 Sep 2;7:43. doi: 10.1186/1476-7120-7-43.
2. Baikoussis NG, Dedeilias P, Argiriou M, Argiriou O, Vourlakou C, Prapa E, et al. Cardiac papillary fibroelastoma; when, how, why? *Ann Card Anaesth* 2016;19(1):162-5.
3. Ikegami H, Andrei AC, Li Z, McCarthy PM, Malaisrie SC. Papillary fibroelastoma of the aortic valve: analysis of 21 cases, including a presentation with cardiac arrest. *Tex Heart Inst J* 2015;42(2):131-5.
4. Gopaldas RR, Atluri PV, Blaustein AS, Bakaeen FG, Huh J, Chu D. Papillary fibroelastoma of the aortic valve: operative approaches upon incidental discovery. *Tex Heart Inst J* 2009;36:160-3.
5. Jha NK, Khouri M, Murphy DM, Salustri A, Khan JA, Saleh MA, et al. Papillary fibroelastoma of the aortic valve - a case report and literature review. *J Cardiothorac Surg* 2010 Oct 17;5:84. doi: 10.1186/1749-8090-5-84.