

A Rare Cause of Embolism: Cardiac Papillary Fibroelastoma

Nadir Görülen Bir Embolizm Nedeni: Kardiyak Papiller Fibroelastom

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Abstract

Cardiac tumors are rarely encountered pathologies, but they may have versatile and different clinical implications. Cardiac papillary fibroelastomas are the third most common primary heart tumors and these are often diagnosed incidentally. When these tumors cause symptoms, they often emerge with cerebral embolic events or myocardial ischemia. Surgical resection of the tumor is recommended to eliminate the risk of embolism and sudden death for symptomatic cases and when the tumor originates from the left side of the heart and mobile. In this case report, we present valve-sparing complete resection of a cardiac papillary fibroelastoma arising from aortic valve which causes neurological symptoms.

Keywords

Cardiac tumors, papillary fibroelastoma, embolism

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Kalp tümörleri, papiller fibroelastom, embolizm

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Öz

Kalp tümörleri nadir olarak karşılaşılan bununla birlikte çok yönlü değişik klinik yansımalara sahip olabilen patolojilerdir. Kardiyak papiller fibroelastomlar tüm kalp tümörleri arasında üçüncü en sık görülen primer tümörleridir ve bu tümörlere çoğunlukla rastlantısal olarak tanı konulmaktadır. Kardiyak papiller fibroelastomlar semptom verdiğinde, sıklıkla embolizme bağlı serebral olaylar veya miyokardiyal iskemi ile karşımıza çıkmaktadır. Semptomatik olgularda ve özellikle kitlenin kalbin sol tarafından köken aldığı ve mobil olduğu durumlarda emboli ve ani ölüm riskini ortadan kaldırmak için tümörün cerrahi rezeksiyonu önerilmektedir. Bu olgu sunumunda nörolojik semptomlar ile kliniğe yansıyan, aort kapağı üzerinde yerleşimli bir kardiyak papiller fibroelastomun kapak koruyucu tam rezeksiyon ile tedavi edilmesi sunulmaktadır.

Introduction

Cardiac tumors are rare pathologies with a frequency of 0.02% in the large autopsy series. Fibroelastomas are the third most common primary tumors after myxomas and lipomas and constitute 8-10% of benign heart tumors (1). The majority of the cases are asymptomatic and usually diagnosed by incidental echocardiographic examinations. However, cardiac papillary fibroelastomas may have variable clinical reflections. They may cause cerebral events due to systemic embolism and myocardial ischemia due to embolism in coronary arteries or tumor

prolapse (1-3). Surgical resection is recommended in symptomatic cases and for mobile tumors originating from the left side of the heart to eliminate the risk of embolism and sudden death (4).

In this paper, we present a case of cardiac papillary fibroelastoma located on the aortic valve causing neurologic symptoms. The tumor was resected surgically by preserving the aortic valve. The surgical techniques used in such cases are discussed and the literature is reviewed.

Case Report

A 37-year-old female patient with no previous history of neurological symptoms or transient ischemic attack admitted to the emergency service with left hemiparesis after syncope. Neurological status improved during the clinical follow-up. Routine blood tests, electrocardiograms, and direct chest X-ray were observed to be normal.

Cranial magnetic resonance imaging (MRI) and carotid doppler ultrasonography examinations did not reveal any pathology. Transesophageal echocardiography showed smooth, sessile, (1.4 x 1.2 cm) mobile mass on the aortic surface of the aortic valve, located between the left coronary and the non-coronary leaflets. The mass was moving in accordance with the heart cycle. There was no aortic valve dysfunction and intracardiac thrombus and other valvular structures were found to be normal. Cardiac MRI was performed to delineate the anatomy of the mass, detect the concomitant pathologies and to plan the surgical strategy. On imaging, the presence of supra-aortic left coronary fissure was confirmed (Figure 1). Considering the risk of recurrent embolism early surgical resection was planned because of the unknown pathology of the mass and the cerebrovascular event that the patient had experienced. Informed consent was obtained from the patient.

After median sternotomy cardiopulmonary bypass was initiated with ascending aorta and right atrium cannulation and the left atrial vent cannula was placed. Following the aortic cross-clamping, diastolic cardiac arrest was maintained via antegrade cardioplegia administration from the aortic root. Transverse aortotomy was performed immediately proximal to the aortic root. On the left coronary leaflet, approximately 1x1 cm sized tumor mass

in close proximity to the non-coronary leaflet was observed (Figure 2). The tumor was macroscopically bright and had a soft, gelatinous structure. The tumor was completely resected by preserving the basal area which is held by the leaflet tissue. Post-resection aortic valve coaptation was normal. The postoperative period was uneventful. Pathologic examination of the

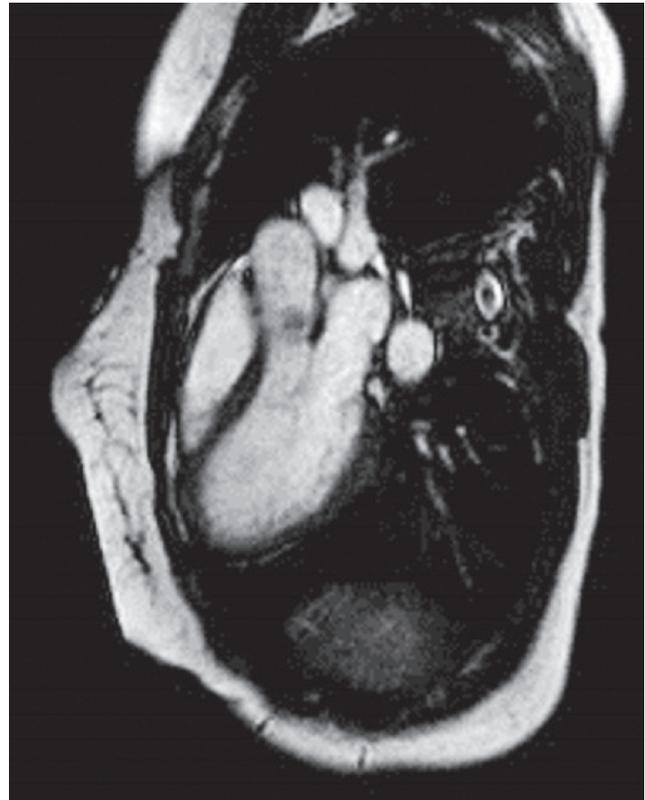


Figure 1. Appearance of the mass on the aortic valve on cardiac magnetic resonance examination

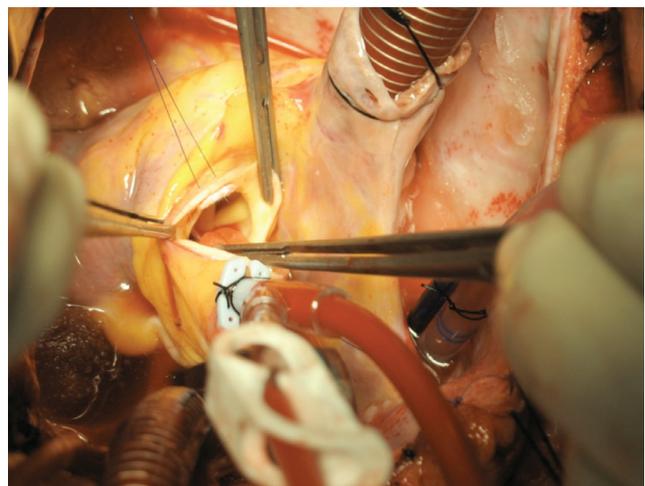


Figure 2. Surgical exposure of the tumor

mass revealed papillary proliferation and hyperplastic endothelial cells containing fibroblast, collagen and elastic fibers, confirming the diagnosis of cardiac papillary fibroelastoma (Figure 3).

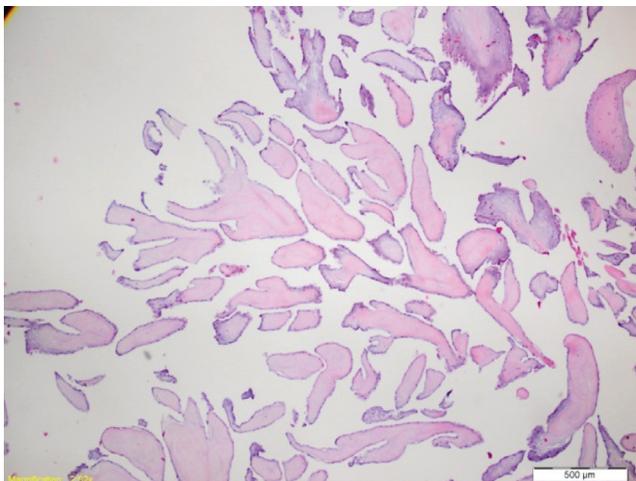


Figure 3. Papillary proliferation and hyperplastic endothelial cells containing fibroblast, collagen and elastic fibers detected on histopathologic examination of the resected mass (hematoxylin eosin staining, x40 magnification)

Discussion

Cardiac papillary fibroelastomas are avascular tumors originating from fibrous tissue, elastic fibers and smooth muscle cells (5). These tumors, usually less than 1 cm in diameter, are held with a short stem in the endocardium. Although the pathogenesis of fibroelastomas is not known precisely, it is suggested that these tumors may be due to mechanical damage to the endothelium or to hamartomatous disease associated with organized thrombus (6). Macroscopically, the appearance of papillary fibroelastomas resembles a marine anemone, typically with extensions in the water (2).

Cardiac papillary fibroelastomas may be diagnosed at any age but are frequently detected in the seventh and eighth decades. Although the incidence of these tumors is not known precisely, it does not differ in terms of gender (1,4). The most commonly affected valve is the aortic valve (44%) however, the papillary fibroelastomas can also be located in the mitral (35%), tricuspid (15%) or pulmonary valves (8%) (7). In symptomatic patients, the clinical picture shows a broad spectrum depending on the location, size, and growth rate and embolization tendency of the tumor. Papillary fibroelastomas which are located in left

heart generally manifest with embolic neurological complications. In our case, papillary fibroelastoma on the aortic valve was also accompanied by history of syncope and hemiparesis.

The mechanisms of thromboembolism in cardiac papillary fibroelastomas involve tumor embolization and thrombus formation on the tumor. For this reason, there may be significant changes in tumor size at short time intervals (8). Since these tumors are usually located on the outflow tract of the left ventricle and exposed to high-velocity blood flow, the risk of thromboembolism is higher than the benign atrial myxomas which are more common heart tumors. Thus, tumor size may not be an indicator for the risk of thromboembolism in cardiac papillary fibroelastomas (9).

There is a consensus on surgical resection for symptomatic cardiac papillary fibroelastomas (10). The management in asymptomatic cases is controversial. Large, mobile tumors originating from the left side of the heart are suggested to be surgically removed to prevent sudden death and embolism. Serial echocardiography examinations and anticoagulation therapy can be adequate in the follow up of small and non-mobile tumors but surgical resection becomes mandatory if there is an increase in tumor size, tumor gets mobile or symptoms occur (4). However, there are no long-term and comprehensive studies evaluating the outcome of non-surgical treatments for cardiac papillary fibroelastomas. For this reason, some authors recommend surgical resection of all cardiac papillary fibroelastomas taking into account the potential risks (9).

In a majority of cases, surgical treatment of cardiac papillary fibroelastomas can be performed with low risk allowing complete removal of tumor tissue. As these tumors are usually pedicellate, simple excision of the tumor with preservation of the valve texture is often sufficient. The stalk and the endocardium to which the tumor is attached must be completely removed. If the defect formed in the tissue is too large, the repair can be performed by direct suture or it may be closed by means of a pericardial or Dacron patch. Replacement of the valve may be necessary in cases of severe mechanical damage to the valve structures or in cases where the tumor is widespread on the leaflet. Care should be taken to avoid fragmentation of the tumor during operation to prevent intraoperative

embolization. All heart cavities should be examined to notice additional tumor masses that may not have been detected before surgery (8).

In our case, the stalked tumor located on the left coronary leaflet of the aortic valve was excised by preserving the valve structure. There was no complication in the early postoperative period and no tumor recurrence was detected in one year follow up.

In conclusion, although cardiac papillary fibroelastomas are rare tumors, they are being more commonly detected with the development of diagnostic tools. Even though these tumors are benign and usually asymptomatic, their natural course is still unknown. Cardiac papillary fibroelastomas can cause embolization and mortality and therefore should be kept in mind in the differential diagnosis of cardiac masses. Early surgical resection is curative in symptomatic cases.

Ethics

Informed Consent: Patient informed consent form was received.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.B., S.D., U.G., Concept: S.D., M.B., Design: S.D., T.K., M.Y., Data Collection or Processing: S.D., M.Y., Analysis or Interpretation: N.Ç., Literature Search: S.D., M.Y., Writing: S.D., T.K.

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