

Five-year follow-up of a papillary fibroelastoma involving the mitral valve in a young patient

Genç bir hastada mitral kapak yerleşimli papiller fibroelastomanın beş yıllık takibi

Nezihi Küçükarslan,¹ Savaş Öz,¹ Eralp Ulusoy,² Erkan Kuralay,¹ Harun Tatar¹

¹Department of Cardiovascular Surgery, Gülhane Military Medical School, Ankara;

²Department of Cardiovascular Surgery, Gülhane Military Medical School Haydarpaşa Training Hospital, İstanbul

Papillary fibroelastomas are primary cardiac tumors of valvular tissue, located on valves, mostly the aortic valve. They are usually seen in elderly patients. In this report, we presented a 21-year-old male patient who underwent surgery for a papillary fibroelastoma involving the mitral valve and was followed-up for five years. No recurrence was encountered, but a minimal increase in mitral valve insufficiency was observed.

Key words: Cardiopulmonary bypass; echocardiography, transesophageal; fibroma/surgery; heart neoplasms/surgery; mitral valve; mitral valve insufficiency; papillary muscles/ultrasonography.

Primary cardiac tumors such as myxoma, lipoma, and papillary fibroelastoma (PFE) are benign tumors that are rarely seen,^[1] the latter accounting for less than 10% of all primary cardiac tumors.^[2] Papillary fibroelastomas usually originate from valvular tissues and they are localized on the surface of the valves. These tumors show a finger-like projection radiating from a central stalk.^[1] Usually a single valve is involved and mostly it happens to be the aortic valve. There are some cases involving the other valves.^[3] The lesions consist of a slender or broad fibrocollagenous stalk from which numerous papillary villous fronds radiate.^[4]

CASE REPORT

A 21-year-old male was hospitalized with a diagnosis of left hemiplegia that was partially recovered at the end of the first week. The history was not consistent with neither endocarditis nor prolonged fever. Physical examination revealed a regular pulse (95/min), normal body temperature, and blood pressure (115/65 mmHg) without any third or fourth heart sounds. There was a systolic murmur (1/6 grade) best heard at the apex of the heart. No signs or symptoms of peripheral endocarditis were found with electrocardiography, chest X-ray, complete blood count, and biochemistry evalua-

Papiller fibroelastomalar kalpte kapak dokudan köken alan ve sıklıkla aort kapağında yerleşen primer kardiyak tümörlerdir. Sıklıkla yaşlı hastalarda gözlenirler. Bu yazıda, mitral kapak yerleşimli papiller fibroelastoma nedeniyle ameliyat edilen ve beş yıl izlenen 21 yaşındaki erkek hasta sunuldu. İzlem döneminde nüks görülmedi, hastanın mitral yetersizliğinde hafif bir artış gözlemlendi.

Anahtar sözcükler: Kardiyopulmoner bypass; ekokardiyografi, transözofageal; fibroma/cerrahi; kalp neoplazisi/cerrahi; mitral kapak; mitral kapak yetersizliği; papiller kaslar/ultrasonografi.

tion. Computed tomography of the brain obtained to rule out cerebral ischemia or hemorrhage was consistent with a thrombotic ischemic area on the right cerebral hemisphere. Transthoracic and transesophageal echocardiography (TTE and TEE respectively) demonstrated a large mobile mass (1.5x1.5 cm) localized in the anterior leaflet of the mitral valve. Color Doppler examination suggested mild mitral regurgitation and hyperdynamic left ventricular wall motion. Based on these findings, surgical intervention was planned on cardiopulmonary bypass using a bicaval standard cannulation technique. After opening the left atrium, the tumor was observed in the anterior mitral leaflet and was excised with a 1-2 mm extensive peritumoral mitral tissue. The defect on the leaflet was primarily closed with a polypropylene suture and Kay annuloplasty was performed to overcome mitral regurgitation. The mitral valve was examined with TEE after cardiopulmonary bypass, which showed mild mitral regurgitation without any residual tumoral mass in the anterior mitral valve. Histopathologic diagnosis of the specimen was papillary fibroelastoma. The postoperative period was normal and the patient was discharged within normal time limit. He was called for periodic examinations once a year for five years, during which time he was evaluated

Received: July 13, 2004 Accepted: February 5, 2005

Correspondence: Dr. Nezihi Küçükarslan, Gülhane Askeri Tıp Akademisi Kalp ve Damar Cerrahisi Anabilim Dalı, 06018 Etilik, Ankara.
Tel: 0312 - 304 52 71 e-mail: nkucukarslan@gata.edu.tr

with physical examination, complete blood analysis, electrocardiography, and echocardiography. During these years, a minimal increase was noted in mild mitral insufficiency, which caused no complaints and did not disturb the life quality of the patient. At the end of the fifth year, blood analysis was completely normal. On physical examination, there was a mild diastolic murmur on the mitral point. Echocardiography showed no mass on the anterior mitral leaflet, but a minimal progress in the degree of mild mitral regurgitation (Fig. 1). Ventricular function, wall dimensions, and ejection fraction were found to be normal. Through the follow up period there was no recurrence.

DISCUSSION

Papillary fibroelastomas are usually asymptomatic and are diagnosed at autopsy. However, they may be associated with strokes or transient ischemic attacks due to cerebral embolism.^[2] Angina, sudden death, and myocardial infarction are other clinical manifestations.^[2,5] The tumors range from 5x3 mm to 10x40 mm in size. In determining the localization, TEE is better than TTE and may be used perioperatively.^[6] On echocardiography, a mobile and pedunculated lesion with a homogeneous echo density is characteristic. Some authors suggest that a small, slowly growing PFE serves as a nidus for an extensive thrombus developing rapidly, so that the lesion can be seen echocardiographically. Based on the reports of many cases, if there is an embolic event suggesting the presence of a PFE, surgical intervention (a simple excision with or without valve repair) will be necessary.^[2] Recurrence of a PFE has not yet been reported.^[2,7]

The treatment of asymptomatic lesions differ from case to case. For small asymptomatic lesions diagnosed on echocardiography, warfarin or antiplatelet treatment may be useful to prevent thromboembolic events.^[8] Many reports suggest that patients with mitral valve tumors larger than 10 mm in diameter have a higher risk for systemic emboli. But a case of embolization due to a PFE of 3 mm in diameter has also been reported.^[2] Thus, the decision for surgical intervention must be taken on careful evaluation. Because of the probable complications such as emboli, myocardial ischemia, and sudden death surgical excision is suggested for larger lesions or for lesions close to the coronary ostia.

We performed both tumor excision and mitral valve reconstruction in our case. At the end of five years, there was no recurrence. Surgical intervention may be associated with valvular insufficiency. But the risk for

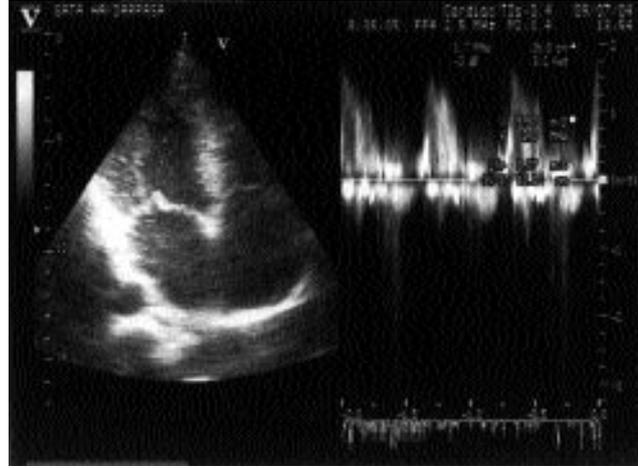


Fig. 1. Echocardiographic image five years after surgery.

minimal insufficiency can be taken to save the patient's life. Meanwhile, careful periodic examinations should be undertaken.

In conclusion, surgical approach is necessary for the treatment of symptomatic PFE cases. Tumor excision and valvular reconstruction were performed in our case. During five years of follow-up, we did not observe any recurrence in terms of ischemic embolism or events.

REFERENCES

1. McAllister HA Jr, Fenoglio JJ Jr. Tumors of the cardiovascular system. In: Atlas of tumor pathology. 2nd series, Fascicle 15. Washington DC: Armed Forces Institute of Pathology; 1978. p. 20-5.
2. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. *Ann Thorac Surg* 1995;59:538-41.
3. Colucci WS, Schoen FJ, Braunwald E. Primary tumors of the heart. In: Braunwald E, editor. *Heart diseases: a textbook of cardiovascular medicine*. 5th ed. Philadelphia: W. B. Saunders; 1992. p. 1451-64.
4. Almagro UA, Perry LS, Choi H, Pintar K. Papillary fibroelastoma of the heart. Report of six cases. *Arch Pathol Lab Med* 1982;106:318-21.
5. Zull DN, Diamond M, Beringer D. Angina and sudden death resulting from papillary fibroelastoma of the aortic valve. *Ann Emerg Med* 1985;14:470-3.
6. Topol EJ, Biern RO, Reitz BA. Cardiac papillary fibroelastoma and stroke. Echocardiographic diagnosis and guide to excision. *Am J Med* 1986;80:129-32.
7. Di Mattia DG, Assaghi A, Mangini A, Ravagnan S, Bonetto S, Fundaro P. Mitral valve repair for anterior leaflet papillary fibroelastoma: two case descriptions and a literature review. *Eur J Cardiothorac Surg* 1999;15:103-7.
8. Brown RD Jr, Khandheria BK, Edwards WD. Cardiac papillary fibroelastoma: a treatable cause of transient ischemic attack and ischemic stroke detected by transesophageal echocardiography. *Mayo Clin Proc* 1995;70:863-8.