Aortic Valve Fibroelastoma: Case Report

Aort Kapağı Fibroelastomu

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ABSTRACT Fibroelastomas are the most common benign cardiac tumors involving the cardiac valves. Although this tumor can be found anywhere in the heart, mostly it is located at the valvular endocardium. Although papillary fibroelastomas are detected coincidentally as an autopsy finding, several advancements and increased availiability in echocardiography enable us to diagnose this entity earlier. Fibroelastomas may result in several clinical conditions especially such as embolization (ischeamic stroke, myocardial infarction). We report a 68-year-old male patient admitted with chest pain and diagnosed by echocardiographic evaluation to have a large mobile mass with a pedicle adhered to the aortic side of right coronary cusp. The tumor was surgically removed to avoid an embolic event and histopathological examination confirmed the diagnosis to be a cardiac papillary fibroelastoma, which is the most common benign valvular neoplasm of the heart.

Key Words: Heart neoplasms; aortic valve

ÖZET Fibroelastomlar kalp kapakçıklarını tutan en yaygın iyi huylu kardiyak tümörlerdir. Her ne kadar bu tümör kalbin her yerinde görülebilse de, genellikle en yaygın lokalizasyonu valvular endokardiyumdur. Bununla beraber, her ne kadar papiller fibroelastomlar otopsi sırasında tesadüfen bulunsa da, ekokardiyografinin yaygın olarak kullanılması bu olguları erken dönemde teşhis etmemize olanak sağlamaktadır. Fibroelastomlar embolizasyon başta olmak üzere bazı hastalıklara yol açarak karşımıza çıkabilir (iskemik inme, miyokard enfarktüsü). Burada, kliniğimize göğüs ağrısı şikayeti ile başvuran 68 yaşındaki bir erkek hastayı sunuyoruz. Hastanın ekokardiyografik incelenmesinde sağ koroner kapakçığın aortik tarafına yapışık büyük mobil pediküler kitle tespit edildi. Olası bir emboliyi önlemek amacıyla, kitle cerrahi operasyonla alındı. Kitlenin histopatolojik incelenmesi en yaygın kalp kapakçığı tümörü olan kardiyak papiller fibroelastom tanısını doğruladı.

Anahtar Kelimeler: Kalp tümörleri; aort kapağı

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Incidence of primary cardiac tumors in necropsy series ranges between 0.0017% and 0.33%, and the echocardiographic incidence is reported as 0.019%.[1] Papillary fibroelastoma is the third most common primary cardiac tumor (accounting for approximately 7% of all primary cardiac tumors) after myxoma and lipoma. It is the most frequent cardiac valvular tumor (70-80%).[1,2] Although this tumor can be found anywhere in the heart, it is mostly located at the valvular endocardium (80%). Before the echocardiography era, papillary fibroelastomas were detected coincidentally at autopsies. However, several advancements in transthoracic (TTE) and transesophageal (TEE) echocardiography enable us to diagnose this entity earlier. A number of cases have been identified as complications of throm-

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boembolism adhered to a tumor embolism, tumors, valvular disease or are found incidentally.[3,4] We report a case with fibroelastoma admitted with angina. A mass located at the aortic valve was detected on echocardiography, and it was successfully resected surgically.

CASE REPORT

A 68-year-old male was admitted to our clinics with chest pain. The medical history revealed that the patient underwent coronary angiography in January 2007, 90% stenosis was found in first small diagonal branch of left anterior descending artery and medical treatment was given for this lesion. His physical examination and routine laboratory tests were unremarkable. The electrocardiogram was also normal. TTE demonstrated normal left ventricular systolic functions, but there were left ventricular hypertrophy and left atrial dilatation. Moreover, a mobile mass (1.6 x 1.2 cm) with a pedicle adhered to the aortic side of right coronary cusp was found. There was no valvular dysfunction. TEE (Figure 1) confirmed the size and the location of the mass and normal valvular functions. Surgical resection was decided due to risk of thromboembolism. The patient underwent tumor excision with aortatomy (Figure 2). On the second day after the operation, atrial fibrillation developed and medical cardioversion with amiodarone restored the sinus rhythm. The patient was discharged four days after the operation. In the follow-up visit one week later, TTE showed that there was no residual mass over the aortic valve (Figure 3)

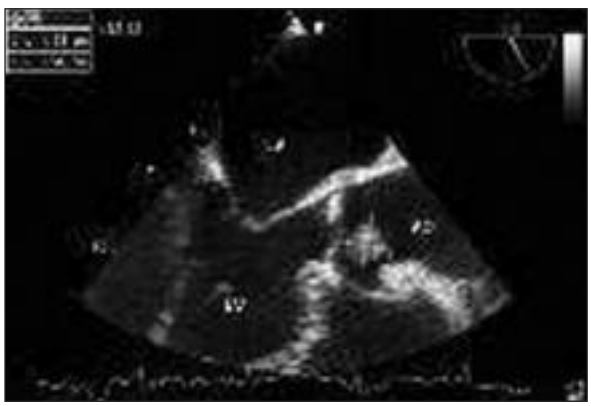

FIGURE 1: Transesophageal echocardiographic images before surgery.

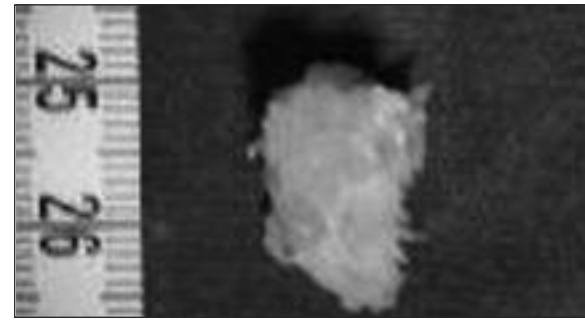

FIGURE 2: Surgical excision of the tumor.

FIGURE 3: Transthoracic echocardiographic image of the patient 2 weeks after surgery.

and that the valvular functions were normal. Histopathological evaluation revealed the mass to be a papillary fibroelastoma.

DISCUSSION

The differential diagnosis between fibroelastoma, vegetation and other tumors is of great importance. However, there is no definite echocardiographic criteria to differentiate fibroelastomas.

Echodensity of the tumor's central collagen core strongly supports the diagnosis and allows differentiation from other intracardiac tumors, vegetations, or mural thrombi.[4]

Etiology of papillary fibroelastoma is not known. Persistent turbulent flow in the heart or endothelial cell disorder due to a particular cause is generally thought to induce endothelial cell hyperplasia and lead to development of papillary fibroelastoma.[5,6] Similarly, Kurup et al.[7] have suggested that open heart surgery and thoracic radiation may cause fibroelastoma. Others claim that fibroelastoma may represent a neoplasm, hamartoma or a inflammatory nodule.[3,4]

The clinical presentation of papillary fibroelastoma (PFE) varies widely from asymptomatic to severe trombo-embolic complications. PFE is an incidental finding in most cases, although the clinical presentation is variable among symptomatic patients and dependent on the location, motility and size of the tumor.[3,6] Most cases originate from the left heart chambers. The most feared complication is systemic embolization, especially to the cerebral or coronary circulation.[1-6] The most common clinical presentation described is stroke or a transient ischemic attack. Other described manifestations are angina, myocardial infarction, sudden death, heart failure, syncope, pulmonary embolism, blindness and peripheral embolism with renal infarction.[1-6] Sudden death and myocardial infarction are the most common manifestations in patients with PFE on the aortic valve.

Approximately 90% of PFE affect cardiac valves usually as a single lesion on the atrial face of atrioventricular valves or on any of the sides of semilunar valves. They rarely occur as multiple lesions. Approximately 44% of PFE are found in aortic valves, followed by the involvement of mitral valve in 35% of cases, tricuspid valves in 15%, and pulmonary valves in 8%.[5] There are case reports of tumors involving all endocardial surfaces, including papillary muscles, tendinous chords, the septum and free walls of the cardiac chambers.[8]

Transthoracic, particularly transesophageal echocardiograms are the ideal methods for tumor diagnosis and characterization, as they usually show the mass with its variable proportions, its motility, delineation, whether it is pedunculated or sessile, and the configuration. They are mostly small (99% less than than 2.0 cm).[1-5]

For symptomatic patients, surgical excision is the choice of treatment in an attempt to preserve the valvular tissue and function.[9] In asymptomatic individuals, surgical procedures are controversial. Sun et al.[4] claimed that surgical resection was necessary in patients with highly mobile and large fibroelastomas and in patients who undergo cardiac surgery due to other cardiac problems. Scalia et al[10] recommended surgery for all left-sided tumors to prevent embolization, but stated that there was no consensus for right-sided tumors. Some investigators suggest surgical treatment to be performed as soon as possible after the diagnosis to prevent embolization.[11] Accordantly, in our case, the patient underwent surgery for a mobile, large mass on the aortic valve to avoid any embolization.

In conclusion, although cardiac papillary fibroelastoma is a benign tumor diagnosed by echocardiography, it can cause life-threatening complications such as stroke, systemic trombo-embolism and sudden death. Thus, prompt diagnosis and appropriate management are needed for this tumor. Surgery by simple excision can be accepted as safe, effective and a valve-sparing technique with good long-term results, especially in left-sided tumors.

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