CASE REPORT

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Papillary fibroelastoma of the aortic valve

Papilarni fibroelastom aortnog zaliska

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Abstract

Introduction. Primary tumors of the heart are rare, usually benign and occur mostly in adults, and usually originate from the endocardium, followed by the myocardium and rarest of the pericardium. Papillary fibroelastoma accounts for less than 10% of all cardiac tumors, but they are most common valvular tumors. The clinical presentation of papillary fibroelastoma varies from asymptomatic cases to cases which have severe clinical presentation that is most likely due to embolic complications. Tumor can usually be discovered by echocardiography or during autopsy. **Case report.** We reported a case of 53-year-old man submitted to routine echocardiographic examination. The patient had the history of hypertension for five years, without any other symptoms. Echocardiography found a round tumor attached to the noncoronary cuspis of the aortic valve. The tumor was surgically removed and pathohistological examination confirmed diagnosis of papillary fibroelastoma. After surgery the patient fully recovered without tumor recurrence or aortic regurgitation. **Conclusion.** Histologically, papilary fibroelastoma is benign tumor of the heart. As demonstrated in this case, a papillary fibroelastoma can be an incidental finding discovered during echocardiography in patients with the history of hypertension.

Key words:
heart neoplasms; fibroma; aortic valve; diagnosis; echocardiography; histological techniques.

Apstrakt

Uvod. Primarni tumori srca su rijetki, uglavnom benigni i javljaju se najšće kod odraslih. Najčešće ovi tumori potiču od endokarda, ređe od miokarda, a najređe od perikarda. Papilarni fibroelastom čini manje od 10% svih tumora srca, ali ovaj tumor predstavlja najčešći tumor koji se javlja na srčanim valvulama. Klinički, ovaj tumor može biti asimptomatski, ali može i dati teške kliničke simptome koji se uglavnom javljaju zbog embolijskih komplikacija. Ovaj tumor se najčešće otkriva ultrazvukom ili prilikom autopsije. **Prikaz bolesnika.** Prikazan je bolesnik, star 53 godine, koji se liječio pet godina od hipertenzije i kod koga je na rutinskom ultrazvuku otkriven okruglast tumor na nekoronarnom kuspisu aortnog zaliska. Tumor je hirurški odstranjen, a patohistološki je imao strukturu papilarnog fibroelastoma. Nakon hirurškog odstranjenja tumora bolesnik se u potpunosti oporavio i nije imao znakove aortne regurgitacije. **Zaključak.** Papilarni fibroelastom histološki je benigni tumor srca. Ovaj prikaz pokazuje da se papilarni fibroelastom može slučajno otkriti u toku ultrazvučnog pregleda srca kod bolesnika sa hipertenzijom.

Ključne reči:
srce, neoplazme; fibromi; zalistak, aortni; dijagnoza; ehokardiografija; histološke tehnike.

Introduction

Primary tumors of the heart are rare, usually benign, and occur mostly in adults. The prevalence of these tumors ranges from 0.002 to 0.3% in autopsy series. Usually, these tumors originate from the endocardium, followed by the myocardium and the rarest by the pericardium [1–4]. The majority of primary tumors of the heart are benign [4, 5]. Papillary fibroelastoma (PFE) accounts for less than 10% of all cardiac tumors, but it is the most common valvular tumor [6–9]. The clinical presentation of papillary fibroelastoma varies from asymptomatic cases to cases with severe clinical presentation most likely due to embolic complications. Most PFEs are not discovered during echocardiography or autopsy. We presented a case of PFE of aortic valve accidentally discovered during echocardiography.

Case report

A 53-year-old man underwent routine echocardiographic examination without any other symptoms. He had been treated for hypertension five years. Echocardiography found a round tumor with the diameter of 13 mm attached with 6 mm length pedicle to the noncoronary cuspis of the

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aortic valve. The tumor was mobile, round, with echo dense and well-demarcated borders (Figure 1). The tumor did not cause any aortic insufficiency.

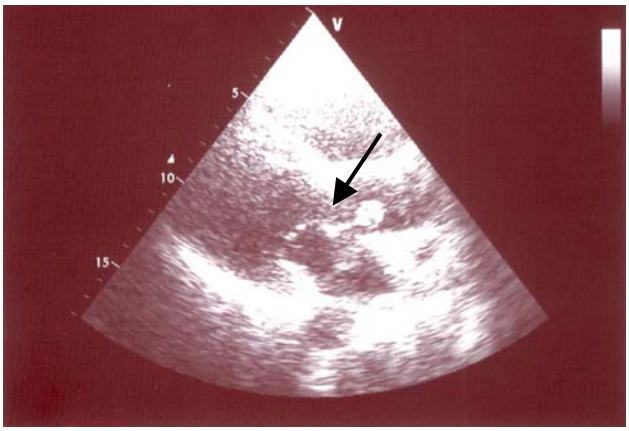

Fig. 1 – Echocardiography (parasternal long axis view) demonstrates a pedunculates mass of 13 mm in size attached to the aortic valve (black arrow).

The patient immediately underwent surgery because of the high risk of embolic complication. The removed tumor was round, soft, grayish, with the diametar of 13 mm. Histopathologic examination showed that the tumor was of papillary configuration. The papillary cord, being an elastic fiber, was surrounded by a mucous layer without blood vessels and covered by a layer of flattened cells. Based on these characteristics, the diagnosis of PFE was made (Figures 2 and 3).

autopsy [10]. The three quarters of hearth tumors are benign. The most common primary benign cardiac tumor is mixoma, while PFEs account for less than 10% of these tumors. The most frequent sites of their occurence are the aortic or mitral valves [11]. In some cases PFE was found in other parts of the heart such as the left ventricle or the atrial septum [12–14].

PFE occurs occasionally, usually in middle-aged and older patients, with a slightly higher incidence in males [2, 15]. Xu et al. [16] described a case of PFE of the tricuspid valve in a 1-mounth-old child. At the time of diagnosis PFE is usually a small tumor with papillary structures of collagen deposits, elastic material and proteoglycans.

To date it remains unclear whether this tumor is true neoplasm, hamartomatous proliferation or organized thromb. Kurup et al. [17] reported that PFE could be associated with irradiation of chest and cardiac surgery. Composition of PFE favors the hypothesis that this tumor represents organized thrombus. Based on the presence of dendritic cells and cytomegalovirus in some patients some authors [18] propose that PFE can be associated with viral endocarditis. De Feo et al. [19] reported a posibility of exposure to environmental pollution with the development of PFE.

Most patients with PFE have no symptoms and the tumors in this patients were incidentally diagnosed on echocardiography, catheterization, cardiac surgery or autopsy [2, 9, 20, 21]. PFE is benign tumor but in some patients torn apart parts of the tumor may lead to embolic complications. Stroke, transient ischemic attack, angina, myocardial infarction or sudden death were the most serious

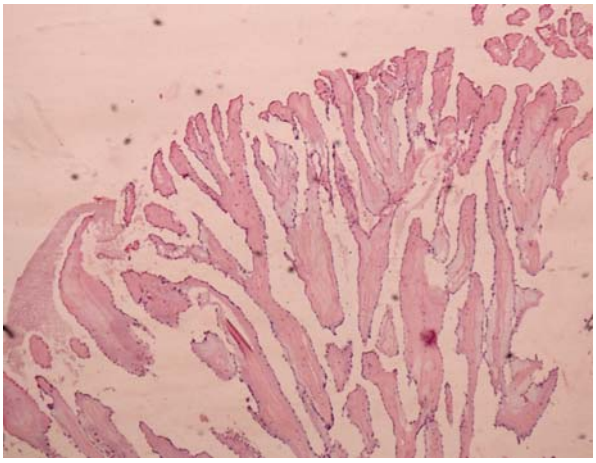

Fig. 2 – The tumor of long, thin papillary structures (HE, × 40).

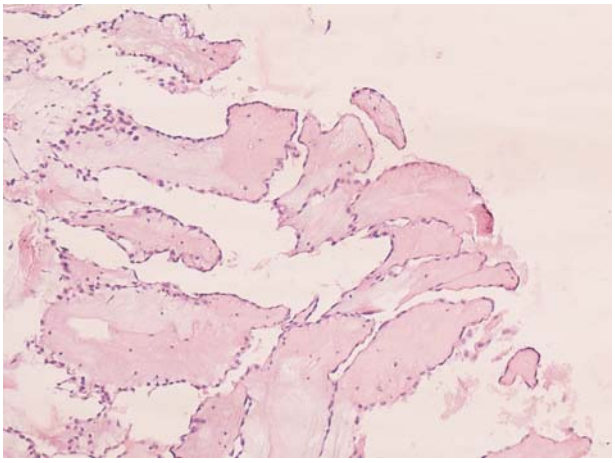

Fig. 3 – The papillary tumor with hyalinized collagenous matrix encountered by uniform single-layer cells without any blood vessel (HE, × 100).

The patient fully recovered after surgery, and echocardiogram 2 and 4 weeks after the surgery demonstrated no tumor recurrence and aortic regurgitation.

Discussion

Tumors of the heart are rare with the prevalence for primary tumors from 0.002% to 0.3% discovered on

complications described, embolism with parts of the tumor, being main reason for the early diagnosis of PFE to be of major interest [6, 15, 22, 23]. Interestingly, valve disfunction is rarely described, although the tumor is usually located on it.

Due to the risk of treatment complication, usually surgical excision of the tumor, it is done in either symptomatic and asymptomatic patients. Patients with tumor size more

than 1 cm, as in the presented case, should be surgically treated by tumor excision due to increased risk of embolization and sudden cardiac death [24, 25]. Asymptomatic patients with nonmobile tumor smaller than 1 cm can be closely followed-up with echocardiography until symptoms develop or tumors enlarges and becomes mobile, and after that tumor could be surgically removed [15, 22]. Patients who from other reasons are not candidates for surgical treatment should be treated with long-term anticoagulation therapy.

Conclusion

Papillary fibroelastomas are histologically rare benign cardiac tumors. Although benign due to their structure and localization these tumors can cause serious embolic complications and sudden cardiac death. Surgical removal of tumors is the best way to prevent possible complications. As demonstrated in this case, papillary fibroelastoma can be an incidental finding in echocardiography.

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