Papillary fibroelastoma of the mitral valve in a patient with recurrent syncope

Papilárny fibroelastóm mitrálnej chlopne u pacientky s opakovanými synkopami

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Abstract. Cardiac papillary fibroelastomas (PF) are rare benign neoplasms. They can result in severe complications, mainly due to embolism, most frequently stroke and transient ischemic attack. Only a few case reports of PF as a culprit of syncope have been published. We herein describe, to the best of our knowledge, the first case report showing a PF of the mitral leaflet protruding into the left ventricular outflow tract in a patient with recurrent syncope. Syncopal workup should also include the evaluation of the infrequent causes involving the left heart masses. Fig. 3, Ref. 10, on-line full text (Free, PDF) www.cardiologyletters.sk

Key words: papillary fibroelastoma – mitral valve – syncope – echocardiography – cardiac surgery
Video at – Supplementary video Papillary https://youtu.be/FSu-M_q1zjw

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Abstrakt. Papilárne fibroelastómy (PF) srdca sú zriedkavé benígne nádory. Môžu spôsobovať závážne komplikácie, dané najmä embolizáciou, najčastejšie ischemickú mozgovú príhodu a tranzitórny ischemický atak. Publikovaných bolo len málo kazuistík, v ktorých bol PF príčinou synkopy. V tejto kazuistike prezentujeme podľa nášho najlepšieho vedomia prvý dokumentovaný prípad PF mitrálneho cípu, protrudujúceho do výtokového traktu ľavej komory, u pacienta s opakovanými synkopami. Diferenciálna diagnostika synkop by mala zahŕňať aj zhodnotenie zriedkavých príčin, vrátane más ľavého srdca. Obr. 3, Lit. 10, on-line full text (Free, PDF) www.cardiologyletters.sk

Kľúčové slová: papilárny fibroelastóm – mitrálna chlopňa – synkopa – echokardiografia – kardiochirurgia
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Introduction

Cardiac papillary fibroelastomas (PF) are rare benign neoplasms but they account for approximately 75% of all valvular tumors. PF are mostly incidentally diagnosed. The proportion of an asymptomatic course, in the case series with a relatively small number of patients with PF, is reported to be 54-60% (1-3). However, PF can result in severe complications, most frequently and primarily due to embolism, mostly stroke and transient ischemic attack. Syncope is a rare manifestation of PF. We report the case of a patient with recurrent syncope and an incidental finding of PF attached to the ventricular

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surface of the anterior mitral leaflet protruding into the left ventricular outflow tract (LVOT).

Patient description

A 72-year-old female was admitted to the Departement of surgery with ileus. Two years before admission she underwent endoscopic colon polypectomy with iatrogenic perforation. The lesion was subsequently treated with a suture. Her medical history also included hypothyreosis, asymptomatic myomatous uterus, hiatus hernia and 'minimally' four unexplained episodes of syncope. CT scan revealed an ileus of the small intestine. The patient was treated conservatively with the restoration of the intestinal passage. Repeated colonoscopy revealed colitis (suspicious for inflammatory bowel disease or ischemic colitis), numerous diverticula in the descending colon and adhesions after the previous surgery. Histology found only nonspecific chronic inflammatory changes without definitive signs of ischemic colitis or inflammatory bowel disease.

The patient was also examined on admission by the internist. On physical examination the cardiac finding was unremarkable. Echocardiography was indicated because of the P-pulmonale present in the ECG leads II, III and aVF. The ECG record was otherwise normal.

Transthoracic and transesophageal echocardiography revealed an incidental finding of an oval hypoechogenic mobile 19x11 mm mass attached by a short narrow pedicle to the ventricular surface of the anterior mitral leaflet. The mass presented a homogeneous appearance. It protruded into LVOT and floated in both directions with respect to the aortic valve **(Figures 1, 2**; **Supplementary material online**, *Video*). The mass was not causing LVOT obstruction by Doppler-flow interrogation, even after the Valsalva maneuver. The mitral valve appeared otherwise structurally normal. There was no valvular regurgitation. The remaining echocardiographic finding was free of significant abnormalities. The patient did not exhibit signs of endocarditis.

She underwent cardiologic workup of the mass and syncope. Cardiac magnetic resonance imaging did not contribute to an additional specification of the mass and was consistent with the echocardiographic finding. Ambulatory 24-hours ECG monitoring, carotid Doppler ultrasound and carotid sinus massage were noncontributory with the physiological findings. Therefore we considered an intermittent occlusion of the LVOT as the probable cause of the recurrent syncope. The patient was referred for cardiac surgery. Preoperative coronary angiography revealed normal findings. She underwent successful complete resection of the fragile, gelatinous valvular mass while completely preserving mitral valve integrity. The patient´s postoperative course was uneventful. She was discharged with no postsurgical complications.

Macroscopically the excised yellowish lesion was made up of soft tissue with numerous papillary fronds. Histopathology revealed a villous tumor composed of loose avascular connective tissue and fronds covered by a single layer of endothelial cells without cellular atypia **(Figure 3)**. On histopathologic examination the mass was diagnosed as a PF.

Nineteen months after the surgery, the patient's condition is stabilized, with no syncope recurrence, and echocardiography showed no residual mass.

Discussion

Only a few case reports of PF as the culprit of syncope have been published. In four cases it was a tumor of the aortic valve, in one case a tumor of the tricuspid valve and

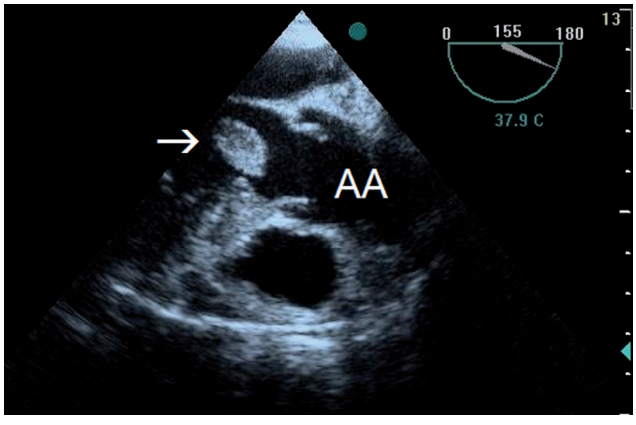

Figure 1 Transesophageal echocardiography. Papillary fibroelastoma (→) prolapsing during systole into the left ventricular outflow tract. AA – ascending aorta.

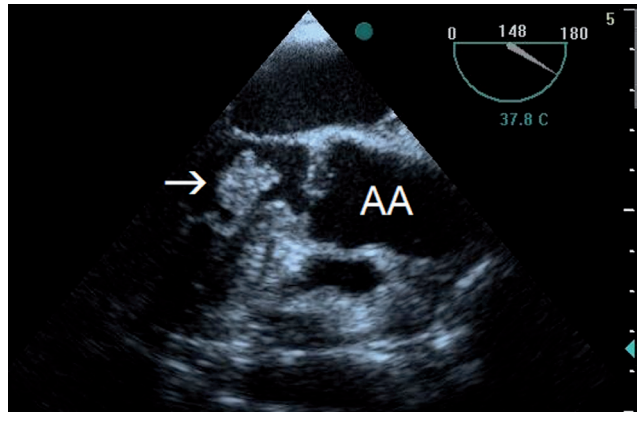

Figure 2 Transesophageal echocardiography. Position of the papillary fibroelastoma (→) in diastole. AA – ascending aorta.

in one case PF at the mitral valve annulus, obstructing the left ventricular inflow tract (4).

To the best of our knowledge, this is the first case report showing a PF of the mitral leaflet protruding into the LVOT in a patient with recurrent syncope. We believe that the intermittent LVOT obstruction, with decreased stroke volume, was the mechanism of syncope. This mechanism is most likely to occur in situations with reduced preload (relative hypovolemia, Valsalva maneuver) or increased left ventricular contractility (excercise induced or after premature ventricular contraction).

Khoueiry et al. described a case of a large PF at the mitral valve annulus, intermittently obstructing the left ventricular inflow tract. The patient was referred for syncope as in our patient (4). Butz et al. published a case report hemodynamically similar to our patient – PF attached to the endocardium of the head of the papillary muscle. The tumor prolapsed during systole into the LVOT. Unlike our patient, their patient did not present with syncope but with exertional dyspnea and atrial flutter (5). In both articles the tumors were said to mimic left atrial myxoma. Wickendon el at. presented a case of a PF attached to the basal septum, protruding into LVOT, with a 15 mmHg gradient which increased to 70 mmHg with a Valsalva maoeuver (6). This patient also did not present with syncope but with chest pain.

In the literature, a relatively small sets of cases of patients with PF have been published, covering 11-54 patients (1-3,7, 8). Usually these were solitary tumors. Multiple lesions are rare. Cianciulli et al. and Val-Bernal et al. found that they affected 13% of patients (1,3). The average number of tumors per patient is reported to be 1.1-1.4 (2, 3, 7). Exceptionally, the large "carpet-like" confluent appearance has been reported (9). The tumors are usually small, with the mean size 11-14 mm (1, 3). Mostly they have a maximal dimension of less than 15 mm (7). Although PF usually have a pedicle, fewer than half are mobile (1).

PF are friable, yellow to yellow-white masses typically having a pedunculated appearance with multiple fronds, causing them to look like a sea anemone. Microscopically, these tumors are composed of numerous avascular fronds with a hypocellular fibrohyalinous matrix. The fronds are lined with endothelial cells.

PF are most often found on cardiac valvular surfaces (4). The aortic valve is affected more commonly than the mitral valve. The pulmonic and tricuspid valves are affected more rarely. PF arise on semilunar valves about as often as from the arterial and ventricular surfaces. In contrast, PF arising from the atrioventricular valves have a predilection for the atrial surface. In our patient, however, the mitral valve tumor was located on the ventricular surface. PF may also be located in both ventricles (including their outflow tracts), both atria and rarely they can be attached to the interatrial septum, Eustachian valve or Chiari network. Theoretically, PF may occur in any location

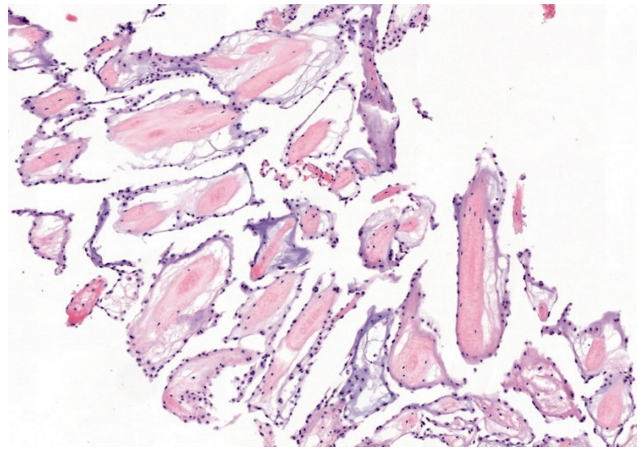

Figure 3 Histology revealing a papillary fibroelastoma with numerous papillary fronds composed of a loose avascular connective tissue and lined with endothelial cells without atypia (hematoxylin and eosin, original magnification x 100).

where the endocardium is located. Very exceptionally, also an extracardiac location in the aortic root has been described.

The unequivocal etiology of PF is unknown. Three etiologic theories can be found in the literature: 1. theory of the hamartoma (true neoplastic origin of PF), 2. reactive theory of traumatic endocardial injury, 3. theory of the organized thrombus.

Most PF are asymptomatic and therefore they are prevailingly incidentally diagnosed. This was also the case of our patient. The most common clinical presentation is embolism, mostly stroke or transient ischemic attack. The emboli can form either from fragments of the frond tissue or from the surface thrombi. Less frequently, PF have also been known to cause aortic or mitral regurgitation or obstruction, syncope, mesenteric, renal, and limb ischemia. Right-sided PF are usually asymptomatic and rarely present with pulmonary embolism. Other unusual complications of PF have been reported very rarely – embolic acute myocardial infarction, sudden death, retinal artery occlusion. In practice, however, it is sometimes difficult to decide definitively whether the symptoms are directly related to PF. An interesting finding is the fact that no significant relation between size, mobility, location and number of tumors, and the presence of embolism or other symptoms has been found (1, 2).

PF can mimic other tumors including myxoma, thrombi, vegetations or Lambl´s excrescences (valvular strands). The vegetations are attached to the "upstream" side of a valve leaflet (i.e. the ventricular surface of the aortic and pulmonary valves and the atrial surface of the mitral and tricuspid valves). PF may arise from the "upstream" or "downstream" side of the valves.

Some authors report that a controversy remains as to whether PF and Lambl´s excrescences represent two dis-

tinct entities, or a spectrum of the same pathologic process (10). Most authors, however, state that PF are, as opposed to Lambl´s excrescences, true neoplasms. Differentially, Lambl´s excrescences are located along the leaflets closure line and PF are typically located on the "midportion" of the leaflet.

There is general consensus that the patients with symptomatic PF should undergo surgical excision with valvular preservation when possible. The surgical procedures include simple tumor excision, excision with valvular reconstruction and valvular replacement. Controversy exists over the management of asymptomatic lesions. Various approaches are recommended – surgical resection for any PF, regardless of the presentation, versus resection for all left-sided PF or for those PF ≥ 1cm in size, or for all mobile tumors irrespective of their size. The operation may be justified by the fact that the surgical removal of PF is simple, safe, efficacious, and definitive (3). When considering the surgical therapy, symptomatology, tumor localization, size and mobility must always be taken into account. There is no clear consensus on whether anticoagulation therapy is indicated in unoperated patients.

Prognosis after the tumor excision is excellent. No death has been reported in the literature during hospitalization or within 30 days after the surgery (2,7). Tumor recurrence after its excision has not been observed.

Conclusion

Syncopal workup should include also the evaluation of the infrequent causes involving left heart masses. Heart tumors should not be overlooked. We have reported the very rare case of recurrent syncope in a patient with an incidental finding of PF attached to the ventricular surface of the anterior mitral leaflet with presumed intermittent LVOT obstruction.

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Supplementary material (video) is available at – Supplementary video Papillary https://youtu.be/FSu-M_q1zjw