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Papillary fibroelastoma as a rare cause of recurrent strokes and myocardial infarction

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A 40-year-old male patient after two ischemic strokes was referred to our outpatient cardiac center for extended assessment. Previous evaluation revealed no significant causes of stroke. It excluded proximal atherosclerosis, arteritis, arterial dissection or atrial fibrillation (AF). Transthoracic echocardiogram demonstrated no intraventricular thrombus, valvular or regional wall motion dysfunction. Thrombophilia testing was negative: checked for lupus anticoagulant, anticardiolipin antibodies, anti-β₂ glycoprotein I antibodies, antithrombin deficiency, protein C or S deficiency, factor V Leiden, prothrombin 20210 mutation, hyperhomocysteinaemia. Screening for connective tissue disease revealed no inflammatory markers elevation, presence of ANA, cANCA or proteinogram abnormalities. Cancer of other organs and infection were excluded. Apart from mildly elevated troponin concentrations in the acute peri-stroke period, patient's blood tests were normal. ECG showed sinus bradycardia and non-specific ST-T changes in the inferior leads.

To search for a minor-risk cardioembolic sources we performed transesophageal echocardiography (TEE), which revealed neither cardiac thrombi, nor patent foramen ovale. It demonstrated a small, mobile, pedunculated mass arising from the anterior mitral leaflet (A1 segment), with maximum size of 4 mm (figure 1A), concerning for differential diagnosis of infective endocarditis, myxoma or papillary fibroelastoma (PFE). Patient was assessed as eligible for surgery. Coronary computed tomography angiography (CCTA) showed normal

coronary arteries with no lesions and confirmed suspicious formation attached to the mitral valve. We repeated prolonged ECG monitoring for 72 hours, which again did not show any AF episodes, but episodes of second- and third-degree atrioventricular block with a spectacular 17 second pause during blood collection, resulting in syncope. By heart team conclusion patient was planned for cardioneuroablation after surgery. To check for potential causes of conduction disorders we performed cardiac magnetic resonance imaging (MRI). Unexpectedly it revealed subendocardial areas of late gadolinium enhancement (LGE) in the inferior wall, indicating an old myocardial infarction [1]. Regarding normal coronary arteries, presumable cause of myocardial infarction must have been embolic.

Finally our patient was referred for surgical resection of the mitral lesion. He underwent surgery in another center according to his wish. The surgical procedure was performed using robotic da Vinci Surgical System. We present intraoperative images (figure 1B, figure 1C). Aspirin 75 mg was used in secondary thromboembolic prevention. Histopathological analysis of the tumour concluded it was a PFE. Since excision our patient has had no further thromboembolic complications so far. Further decisions regarding treatment of conduction disorders are at the discretion of center chosen by the patient.

Presented case illustrates how challenging the assessment of stroke causes may be. In approximately one-fourth of strokes their causes fail to be determined, thus many strokes are cryptogenic [2]. We present a case of a PFE causing two embolic strokes and a myocardial infarction. Although primary cardiac tumours are rare (0.02% prevalence) [3], with PFE as one of the most common, we should remember about the importance of wide diagnostic approach when determining the cause of thromboembolic events, especially in an otherwise negative workup [4]. Apparently secondary prevention with aspirin, P2Y12 inhibitors or VKA /NOAC anticoagulation might not always be enough.

Article information

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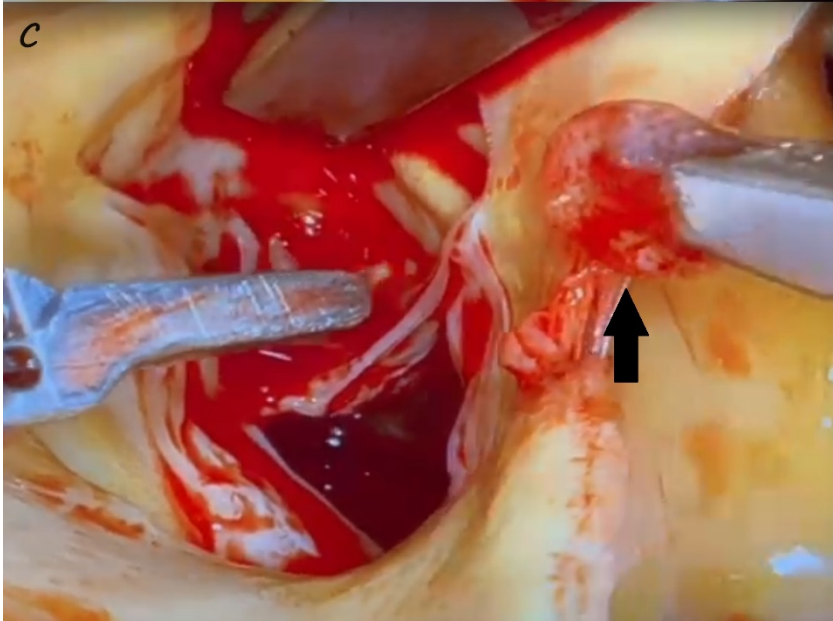
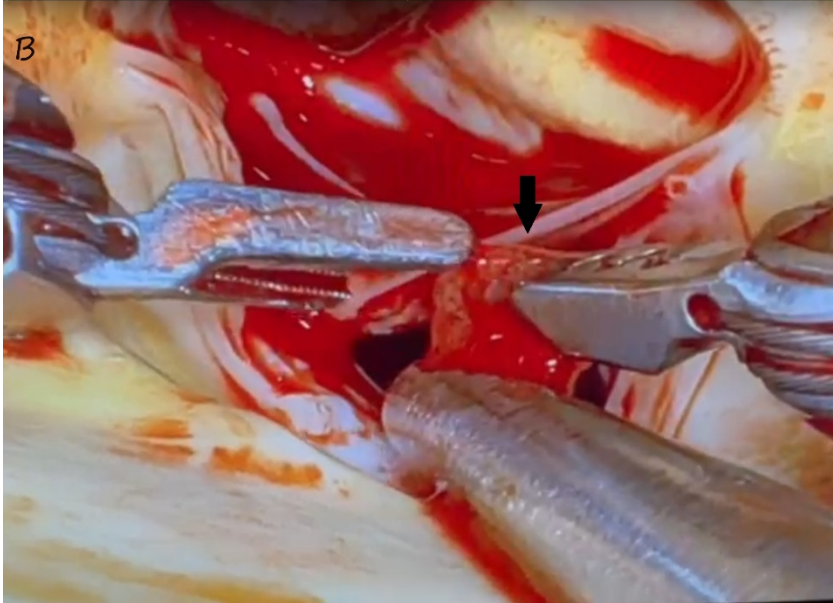
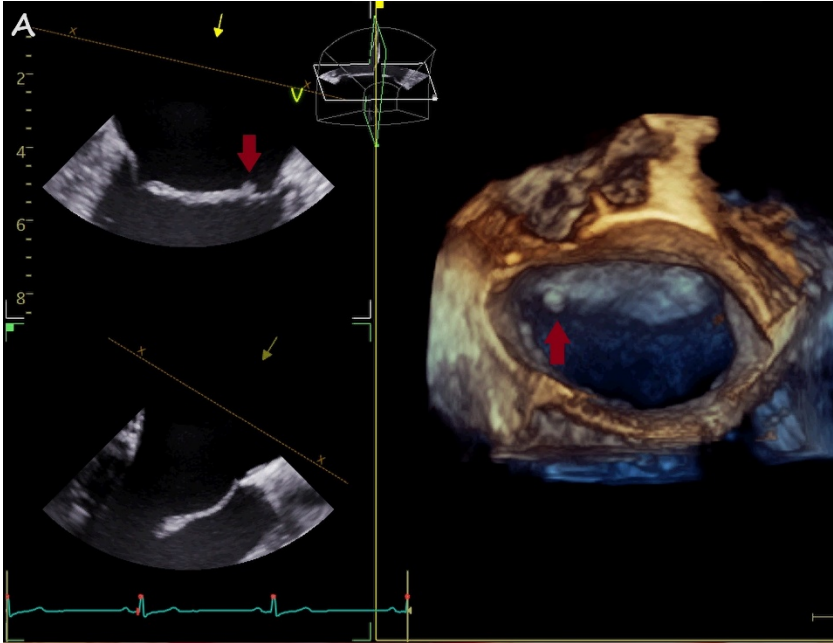


Figure 1. A. Transesophageal echocardiography (TEE) showing pedunculated mass arising from the anterior mitral leaflet (A1 segment). **B, C.** Intraoperative images.