

Case Report

Papillary Fibroelastoma of the Cardiac Valves: A Rare Cause of Embolic Stroke

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Papillary fibroelastomas are rare, primary, benign cardiac tumours most frequently located in the heart valves. They are a potential cause of systemic emboli, stroke, myocardial infarction and sudden death. We present two cases of papillary fibroelastomas located in the mitral and aortic valves of patients who had suffered multiple strokes. The fibroelastomas were diagnosed using transoesophageal echocardiography and the patients were treated surgically, with broad excision of the tumours and preservation of the valves. The echocardiographic and microscopic characteristics of the fibroelastomas are analysed and methods for their differential diagnosis from other cardiac tumours are discussed in the context of the available literature.

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Papillary fibroelastoma is a rare, primary, benign cardiac tumour that is most frequently located in the heart valves.¹ In order of frequency, it is the third primary cardiac tumour, after myxoma and lipoma. Fibroelastomas are usually found by chance in post mortem examinations. However, their prompt detection is of great importance because they are potential causes of systemic emboli, stroke, myocardial infarction and sudden death.²⁻⁴ Even rarer are multiple fibroelastomas, which show an echocardiographic picture of multiple pedunculated formations most commonly located in the atrioventricular valves.⁵

We present two cases of papillary fibroelastomas located in the mitral and aortic valves in patients who had suffered multiple strokes.

Case descriptions

First patient

A man aged 52 years, with a history of successive severe cerebrovascular strokes resulting neurologically in right hemiplegia, underwent laboratory and paraclinical test-

ing to determine the cause of the neurological events. The patient was free of symptoms before the episodes and had no relevant history. Regarding risk factors for coronary artery disease, the patient suffered from hypertension, dyslipidaemia, mild diabetes mellitus controlled by diet; he was a smoker and had elevated homocysteine levels. The 24-hour Holter ECG showed no pathological findings. Computerised tomography of the brain showed an extensive low-density region in the left hemisphere near the basal ganglia and the occipital area, and oedema of the left hemisphere with left-to-right displacement of the median line structures. Echocardiographic examination of the carotid arteries showed that the common carotids were patent bilaterally, showing non-calcified atheromatous plaque, especially in the right common carotid, without any notable stenosis or disturbance of blood flow. Transoesophageal echo found two pedunculated, mobile structures in the left atrium (the larger one 0.9 x 0.4 cm in size) at the entrance to the atrial appendage, protruding through the mitral valve annulus, with features suggestive of a fibroelastoma, although

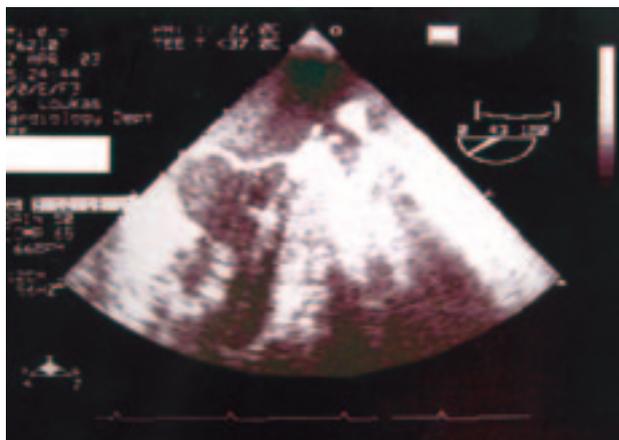


Figure 1. Transoesophageal echogram (long axis 40-60°) showing the two pedunculated formations (fibroelastomas) on the mitral annulus at the entrance to the left atrial appendage.

myxoma could not be ruled out (Figure 1). Coronary angiography revealed two-vessel coronary artery disease with severe stenoses (anterior descending and diagonal branches, right coronary artery). The patient was diagnosed as having “multiple ischaemic infarcts of cardioembolic aetiology due to processes in the left atrium” and was referred for triple coronary bypass surgery and removal of the structures in the left atrium (broad excision of the structures with their base and direct suturing). The surgical findings showed two pedunculated structures each about 1.5 cm in size, which were found on the mitral valve annulus between its P1 section and the left atrial appendage.

The patient’s postoperative course was smooth and uncomplicated. On re-examination twelve months after the surgery he was clinically improved following intensive physiotherapy during that period. Twenty-one months after the procedure the patient has had no new neurological episode.

Microscopic findings

A section of the mitral annulus and left atrial appendage were examined. In the wall two small, anomalous, cystic regions were seen, surrounded by a very thin band of myocardium and covered with broadened endothelial cells. The regions described macroscopically correspond to regions that mainly consisted of myxomatous connective tissue. A few free micropapillary formations were found ipsilaterally, made up centrally from homogenised eosinophilic material, peripherally from a thin band of myxomatous material, and coated

with atrophic endothelium. These findings were consistent with papillary fibroelastoma (Figure 2).

Second patient

A woman aged 62 years with a history of multiple transient strokes came to our clinic for a transoesophageal echo examination aimed at investigating emboli of probable cardiac origin. The patient had no cardiological history and physical examination showed no pathological findings. Computerised tomography showed a small low-density region in the left parietal lobe, without compression phenomena, and an absence of cerebral oedema. The echocardiographic examination of the carotid arteries was normal. The transoesophageal echo showed a pedunculated, mobile structure on the aortic surface of the non-coronary cusp of the aortic valve (0.7 cm long), probably a fibroelastoma or vegetation, although myxoma could not be ruled out (Figure 3). The patient underwent coronary angiography, which showed normal coronary vessels, and was operated on for excision of the base of the structure with preservation of the valve. The perioperative transoesophageal echogram confirmed the integrity of the valve.

The patient’s course was uncomplicated and twelve months later she remains free of symptoms without the appearance of a new neurological syndrome.

Microscopic findings

Vegetations from vitrified connective tissue with a structure similar to that seen in the chordae tendineae.

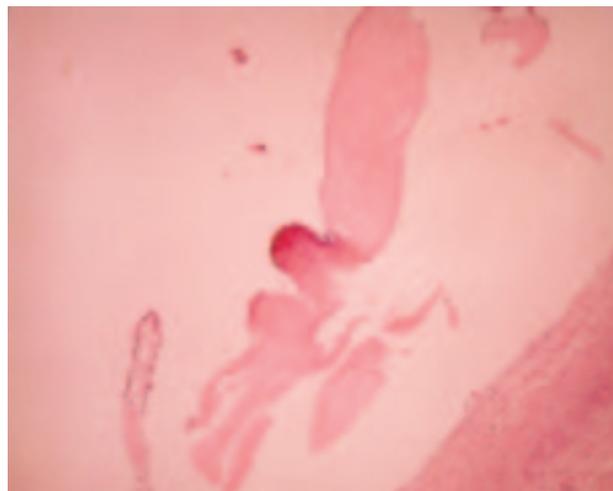


Figure 2. Papillary formations with homogenised stroma. The atrial wall is on the lower right (H+E, 10x).

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