CASE STUDY

Fibroelastoma: case report and review of the literature

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Abstract
A 64 year old woman presented with right hemianaesthesia and was found to have a pansystolic apical murmur with systolic and diastolic posture related plops. Echocardiography revealed a mobile mass on the mitral valve apparatus that was confirmed by magnetic resonance imaging. This was successfully excised and was proven on histopathological examination to be a fibroelastoma. Other cases of fibroelastoma from the literature are reviewed.

Keywords: fibroelastoma; mitral valve; transient ischaemic attack

A 64 year old woman presented with an episode of right hemianaesthesia that resolved completely within a few minutes. A diagnosis of transient ischaemic attack (TIA) was made. Her past medical history included: a murmur detected at the age of 14 years, herpes zoster infection, and right breast carcinoma treated with lumpectomy and radiotherapy with no evidence of disease at five years of follow up.

On examination she was in sinus rhythm with normal pulse and blood pressure. On auscultation, a pansystolic apical murmur with intermittent posture related systolic and diastolic plops were heard. There was no carotid bruit.

Routine blood tests were normal except for potassium of 3.2 mmol/l (normal range 3.5–5) and erythrocyte sedimentation rate (ESR) of 40 mm in the first hour. She had a normal electrocardiogram (ECG), 24 hour Holter ECG, chest radiograph, and duplex scan of the carotid and vertebral arteries. Computed tomography (CT) of the brain showed minimal cerebral atrophy. A mobile mass attached to the mitral valve and the chordae tendineae was detected on cross sectional transthoracic echocardiography (TTE) (fig 1), and confirmed by magnetic resonance imaging (MRI) of the heart (fig 2). Right heart catheterisation and coronary arteriography were normal. Left ventricular angiography was not attempted because of the risk of embolism.

Management
The patient was anticoagulated with intravenous heparin, and referred for surgery. At operation, a 1 cm papillary mass was found within a small left atrium. It was attached to both the middle of the free edge of the anterior leaflet of the mitral valve and to adjacent chordae tendineae (fig 3). The mass was excised, with a small piece of chorda. The patient made an uneventful recovery, her potassium was normal and she was discharged home on aspirin 75 mg daily.

After 44 months of follow up she remains asymptomatic with a pansystolic apical murmur. A recent cross sectional TTE proved the presence of mild mitral regurgitation. Her ESR is normal at 9 mm in the first hour.

Histopathology
Histopathology revealed papillary growth covered by a single layer of hypertrophied endothelium. The stroma was composed of a central dense amorphous fibrous core and an outer layer of loose connective tissue, consistent with a papillary fibroelastoma of the endocardium.

Discussion
DESCRIPTION
The majority of fibroelastomas are valvar. When they involve the semilunar valves, either side of the leaflets could be affected, whereas...
the atrial side is the most commonly affected when the atrioventricular valves are involved. Along the valve leaflet, the most commonly involved site is the midpoint, well away from the annulus. Papillary fibroelastoma can also involve the chordae tendineae, the endocardium of the right atrium, and both ventricles, including the papillary muscles and the ventricular septum. The size of the tumour varies, with 83% being less than 1 cm in diameter. The histological character of the lesion consists of multiple papillary villous fronds radiating from a central fibrocollagenous stalk with each frond showing three zones: a central core that contains collagen, reticulin, and elastin; a peripheral myxomatous zone; and an outer rim of hyperplastic endothelial cells. It is this architecture of the papillary fronds of fibroelastoma that distinguishes it from cardiac myxoma. In our case, it is intriguing that the macroscopic features were thought at the time of surgery to have been representative of a myxoma. The reason for that is unclear, we suspect that fusion of the fronds by thrombosis caused the smooth macroscopic appearance of the masses.

PRESENTATION
The majority of the cases described in the literature were incidental findings in the process of cardiac investigation for an unrelated problem, or they were found at necropsy. It is well recognised that cardiac tumours are great mimics of other forms of cardiac disease; fibroelastomas are no exception. In addition to the obvious risk of systemic emboli, patients can present in several ways.

Chest pain—This could be non-exertional pain. It may mimic the pain of aortic dissection with its radiation to the back, as in the case of Zull et al when the postmortem examination revealed a 1.5 cm papillary fibroelastoma on the aortic valve. The tumour was acting like a ball valve occluding the left coronary artery ostium, and had resulted in an embolus that affected the left anterior descending artery (LAD). Acute myocardial infarction (AMI) could result either from a fibroelastoma of the aortic valve dynamically occluding the ostium of one of the two main coronary arteries, or from coronary embolisation from a fibroelastoma of the aortic valve or the left ventricle.

Sudden death—The case of sudden unexpected death of a 21 year old male was ascribed to the embolisation of the left main stem and LAD by segments of a fibroelastoma of the aortic valve.

Cerebral embolisation—Caused by either a fibrin thrombus forming on the tumour that serves as a nidus, or by a fragment of the tumour itself. This can lead to TIAs in several arterial systems, resulting in a variety of clinical presentations including partial loss of vision due to embolic occlusion of branch retinal artery or complete loss of vision (amaurosis fugax). Unfortunately, the cerebral ischaemia could also result in completed strokes.

Asymptomatic or incidental finding—This was reported in variable circumstances, including asymptomatic patients, patients undergoing coronary artery bypass surgery (CABG), and in patients being investigated for a heart murmur, for heart failure or valvar...
regurgitation, or for rheumatic valvar stenoses. This occurred from a right heart fibroelastoma that presented with repeated episodes of respiratory distress and fever.

Atypical presentation—This is exemplified by the case of a woman with recurrent urinary tract infection, masquerading as culture negative infective endocarditis with multiple fibroelastoma masses in the left ventricle, aortic, and mitral valves. Interestingly that patient had a septal myectomy for hypertrophic obstructive cardiomyopathy 15 years earlier. Surgery was carried out when the patient developed cardiac failure secondary to worsening mitral regurgitation.

INVESTIGATIONS

Cardiac investigations in patients with TIAs

Echocardiography has been shown to be of limited value in defining a source of embolisation in patients without clinical evidence of cardiac disease, hypertension or atrial fibrillation. There are non-pathognomonic pointers to the heart as a possible source. These include aortic valve thickening, mitral valve annular calcification, mitral valve prolapse, or left ventricular wall motion abnormalities.

For fibroelastoma

There are no findings diagnostic of intracardiac masses on physical examination, blood tests, chest radiography or ECG. The rise in ESR is non-specific. The hypokalaemia noted in our case has been reported in another. The introduction of echocardiography has made the non-invasive diagnosis of this tumour possible; however, it is almost impossible to differentiate fibroelastoma from myxomas or thrombi using that investigation alone. Transthoracic echocardiography (TOE) is valuable, not only in confirming the presence of a cardiac mass but also in defining the mass and in particular its attachment to the underlying tissue, its exact location, and its relation with surrounding structures. In addition to confirming the diagnosis and planning management and surgical approach, investigators have also used TOE to help the surgeon intraoperatively by confirming complete excision and testing the effectiveness of any accompanying valve repair or the presence of any remnant valvar regurgitation.

Thoracic CT scanning has not been used to diagnose fibroelastoma except in the case reported by Zull and colleagues, where it was used to investigate the possibility of aortic dissection as a cause of the patient's chest pain. MRI was used in our patient. It confirmed the diagnosis but added little to the information already gained from echocardiography. This is the first report on the use of the MRI in the preoperative imaging of the lesion.

Transvenous biopsy was used to obtain histopathological diagnosis by Schwingen et al., where the cross sectional TTE and TOE confirmed that the fibroelastoma of the right atrium was unresectable.

TREATMENT

The patient should receive anticoagulation to reduce the risk of embolisation before surgical exploration. At surgery, if a fibroelastoma is recognised simple excision of the tumour is usually all that is required, in contrast to the need for a wide base excision in cardiac myxomas.

When the valves are involved repair is often possible. If involvement of the valvar apparatus is more extensive, resection and valve replacement may be necessary.

COMMENTS

We present another case of papillary fibroelastoma presenting with a TIA, highlighting the importance of including this condition in the differential diagnosis of TIA. In this case hypokalaemia was found preoperatively but disappeared spontaneously postoperatively. We can offer no explanation for this finding. The presence of hypokalaemia in two cases of papillary fibroelastoma raises the question of whether these tumours, or a subset of them, has any endocrine activity. This question can only be answered by reviewing all of the suitable tissue specimens still available, and by alerting clinicians and histopathologists to this question, which can then be investigated.

Although we demonstrated the use of MRI in the imaging of this tumour, we did not find that it yielded any diagnostic information not available from cross sectional TTE.

Finally, the curiously smooth macroscopic appearance of the tumour led us initially to suspect a myxoma, although the microscopy proved the tumour to be a fibroelastoma. We postulate that this may have been secondary to thrombosis leading to fusion of the fronds, which normally give the papillary fibroelastoma its distinctive macroscopic appearance.

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