CASE STUDY

Fibroelastoma: case report and review of the literature

A Al-Mohammad, H Pambakian, C Young

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.

(Heart 1998;79:301-304)

Keywords: fibroelastoma; mitral valve; transient ischaemic attack

Cardiac Department,
Aberdeen Royal
Infirmary, Foresterhill,
Aberdeen AB25 2ZD,
UK

A Al-Mohammad

Department of
Histopathology, St
Thomas's Hospital,
London, UK
H Pambakian

Department of
Cardiothoracic
Surgery, St Thomas's
Hospital
C Young

Correspondence to:
Dr Al-Mohammad.

Accepted for publication
18 August 1997

Figure 1 Echocardiography showing a very mobile mass in the left ventricular aspect of the anterior leaflet of the mitral valve.

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

Figure 2 Magnetic resonance imaging of the heart showing thickened mitral valve leaflets and on the cine imaging a mass measuring 12 × 12 mm attached to the tip of the anterior leaflet of the mitral valve, extending into the outflow tract of the left ventricle during systole. Turbulence in the ascending aorta was noted and is compatible with disturbed flow by the presence of the mass in the left ventricular outflow tract.

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.¹ Papillary fibroelastoma can also involve the chordae tendinae, the endocardium of the right atrium,² and both ventricles, including the papillary muscles and the ventricular septum.³ The size of the tumour varies, with 83% being less than 1 cm in diameter.³

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.⁴ 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.⁵ It may mimic the pain of aortic dissection with its radiation to the back, as in the case of Zull *et al*,⁶ 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,^{7,8} or from coronary embolisation from a fibroelastoma of the aortic valve or the left ventricle.⁹

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.¹⁰

Cerebral embolisation—Caused by either a fibrin thrombus forming on the tumour that serves as a nidus,¹¹ or by a fragment of the tumour itself. This can lead to TIAs¹²⁻¹⁵ in several arterial systems,¹⁶⁻¹⁹ resulting in a variety of clinical presentations including partial loss of vision due to embolic occlusion of branch retinal artery²⁰ or complete loss of vision (amaurosis fugax).²¹ Unfortunately, the cerebral ischaemia could also result in completed strokes.¹¹

Asymptomatic or incidental finding—This was reported in variable circumstances, including asymptomatic patients,^{22,23} patients undergoing coronary artery bypass surgery (CABG),^{24,25} and in patients being investigated for a heart murmur,^{5,26,27} for heart failure or valvar

Figure 3 Macroscopic appearance of the tumour intraoperatively. (A) Relation to the mitral valve; (B) size of the tumour after excision.

regurgitation,²⁸ or for rheumatic valvar stenoses.^{4 29}

Pulmonary embolisation—This occurred from a right heart fibroelastoma that presented with repeated episodes of respiratory distress and fever.³⁰

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.³¹

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.³² 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.³³

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.⁶

The introduction of echocardiography³⁴ 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.³⁵ Transoesophageal echocardiography (TOE) is valuable, not only in confirming the presence of a cardiac mass^{8 35} but also in defining the mass²³ and in particular its attachment to the underlying tissue,³⁶ its exact location, and its relation with surrounding structures.³⁷ In addition to confirming the diagnosis and planning management and surgical approach, investigators have also used TOE to help the surgeon intraoperatively¹¹ by confirming complete excision²⁹ and testing the effectiveness of any accompanying valve repair or the presence of any remnant valvar regurgitation.³⁸

Thoracic CT scanning has not been used to diagnose fibroelastoma except in the case reported by Zull and colleagues,⁶ 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 Schwinger *et al*,³⁷ 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.¹⁶ At surgery, if a fibroelastoma is recognised simple excision of the tumour is usually all that is required,¹⁵ in contrast to the need for a wide base excision in cardiac myxomas.

When the valves are involved repair is often possible.^{5 10 11 13 16 18 39} If involvement of the valvar apparatus is more extensive, resection and valve replacement may be necessary.^{5 12 19}

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.

We thank Dr S Karwatowski and Professor R Underwood of the Royal Brompton Hospital, London, for their help in performing the MRI. We are particularly indebted to Dr William Travis of the US Armed Forces Institute of Pathology, Washington DC, USA, for reviewing the microscopic findings on the patient reported in this paper.

- 1 Edwards FH, Hale D, Cohen A, *et al*. Primary cardiac valve tumours. *Ann Thorac Surg* 1991;52:1127–31.
- 2 Gallas MT, Reardon MJ, Reardon PR, *et al*. Papillary fibroelastoma. A right atrial presentation. *Texas Heart Inst J* 1993;20:293–5.
- 3 Abu Nassar SG, Parker JC Jr. Incidental papillary endocardial tumour. Its potential significance. *Arch Pathol* 1971;92:370–6.
- 4 Almagro UA, Perry LS, Choi H, *et al*. Papillary fibroelastoma of the heart. *Arch Pathol Lab Med* 1982;106:318–21.
- 5 Shub C, Tajik AJ, Seward JB, *et al*. Cardiac papillary fibroelastomas. Two-dimensional echocardiographic recognition. *Mayo Clin Proc* 1981;56:629–33.
- 6 Zull DN, Diamond M, Beringer D. Angina and sudden death resulting from primary papillary fibroelastoma of the aortic valve. *Ann Emerg Med* 1985;14:470–3.
- 7 Israel DH, Sherman W, Ambrose JA, *et al*. Dynamic coronary ostial obstruction due to papillary fibroelastoma leading to myocardial ischaemia and infarction. *Am J Cardiol* 1991;67:104–5.
- 8 Etienne Y, Jobic Y, Houel JF, *et al*. Papillary fibroelastoma of the aortic valve with myocardial infarction: echocardiographic diagnosis and surgical excision. *Am Heart J* 1994;127:443–5.
- 9 Mazzucco A, Bortolotti U, Thiene G, *et al*. Left ventricular papillary fibroelastoma with coronary embolisation. *Eur J Cardiothorac Surg* 1989;3:471–3.
- 10 Amr SS, Abu Al Ragheb SY. Sudden unexpected death due to papillary fibroma of the aortic valve. Report of a case and review of the literature. *Am J Forens Med Pathol* 1991;12:143–8.

- 11 Topol EJ, Biern RO, Reitz BA. Cardiac papillary fibroelastoma and stroke. Echocardiographic diagnosis and guide to excision. *Am J Med* 1986;**80**:129–32.
- 12 Fowles RE, Miller DC, Ebger BM, *et al.* Systematic embolisation from a mitral valve papillary endocardial fibroma detected by two-dimensional echocardiography. *Am Heart J* 1981;**102**:128–30.
- 13 Kroll W, Nellessen U, Hofig M, *et al.* Young patient with left brain infarct and transient right sided hemiparesis and cardiac papillary fibroelastoma. *Zeitschrift Fur Kardiologie* 1991;**80**:234–6.
- 14 Ragni T, Grande AM, Cappuccio G, *et al.* Embolizing fibroelastoma of the aortic valve. *Cardiovasc Surg* 1994;**2**:639–41.
- 15 Mann J, Parker DJ. Papillary fibroelastoma of the mitral valve: a rare cause of transient neurological deficits. *Br Heart J* 1994;**71**:6.
- 16 Ong LS, Nanda NC, Barold SS. Two-dimensional echocardiographic detection and diagnostic features of left ventricular papillary fibroelastoma. *Am Heart J* 1982;**103**:917–18.
- 17 Marvasti MA, Obeid AT, Cohen PS, *et al.* Successful removal of a papillary endocardial fibroma. *Thorac Cardiovasc Surg* 1983;**31**:254–5.
- 18 McFadden PM, Lacy JR. Intracardiac papillary fibroelastoma: an occult cause of embolic neurologic deficit. *Ann Thorac Surg* 1987;**43**:667–9.
- 19 Gorton ME, Soltanzadeh H. Mitral valve fibroelastoma. *Ann Thorac Surg* 1989;**47**:605–7.
- 20 Zamora RL, Adelberg DA, Berger AS, *et al.* Branch retinal artery occlusion caused by a mitral valve papillary fibroelastoma. *Am J Ophthalmol* 1995;**119**:325–9.
- 21 Schuetz WH, Welz A, Heymer B. A symptomatic papillary fibroelastoma of the left ventricle removed with the aid of transoesophageal echocardiography. *Thorac Cardiovasc Surg* 1993;**41**:258–60.
- 22 Ushida S, Obayashi N, Yamanari H, *et al.* Papillary fibroelastoma in the left ventricular outflow tract. *Heart Vessels* 1992;**7**:164–7.
- 23 LiMandri G, Homma S, Di Tullio MR, *et al.* Detection of multiple papillary fibroelastomas of the tricuspid valve by transoesophageal echocardiography. *J Am Soc Echocardiogr* 1994;**7**:315–7.
- 24 Autz L, Krieger KH, Yao FS. Transesophageal echocardiographic localization of an aortic valve papillary fibroelastoma during routine coronary artery bypass surgery. *J Heart Valve Dis* 1993;**2**:662–4.
- 25 Nakao T, Hollinger I, Attai L, *et al.* Incidental finding of papillary fibroelastoma on the atrial septum. *Cardiovasc Surg* 1994;**2**:423–4.
- 26 Flotte T, Pinar H, Feiner H. Papillary fibroelastoma of the left ventricular septum. *Am J Surg Pathol* 1980;**4**:585–8.
- 27 Shapira OM, Williamson WA, Dugan JM. Papillary fibroelastoma of the mitral valve. *Cardiovasc Surg* 1993;**1**:599–601.
- 28 Cha SD, Incarvito J, Fernandez J, *et al.* Giant Lambl's excrescences of the papillary muscles and the aortic valve: echocardiographic, angiocardiographic and pathologic findings. *Clin Cardiol* 1981;**4**:51–4.
- 29 Kalman JM, Lubicz S, Brennan JB, *et al.* Multiple cardiac papillary fibroelastomas and rheumatic heart disease. *Aust NZ J Med* 1991;**21**:744–6.
- 30 Waltenberger J, Thelin S. Images in cardiovascular medicine. Papillary fibroelastoma as an unusual source of repeated pulmonary embolism. *Circulation* 1994;**89**:2433.
- 31 Lee KS, Topol EJ, Stewart WJ. Atypical presentation of papillary fibroelastoma mimicking multiple vegetations in suspected subacute bacterial endocarditis. *Am Heart J* 1993;**125**:1443–5.
- 32 Lovett JL, Sandok BA, Giuliani ER, *et al.* Two-dimensional echocardiography in patients with focal cerebral ischaemia. *Ann Intern Med* 1981;**95**:1–4.
- 33 Greenland P, Knopman DS, Mickell FL, *et al.* Echocardiography in diagnostic assessment of stroke. *Ann Intern Med* 1981;**95**:51–3.
- 34 Gopal A, Li Mandri G, King DL, *et al.* Aortic valve papillary fibroelastoma. A diagnosis by transthoracic echocardiography. *Chest* 1994;**105**:1885–7.
- 35 Richard J, Castello R, Dressler FA, *et al.* Diagnosis of papillary fibroelastoma of the mitral valve complicated by non-Q-wave infarction with apical thrombus: transoesophageal and transthoracic echocardiographic study. *Am Heart J* 1993;**126**:710–12.
- 36 Thomas MR, Jayakrishnan AG, Desai J, *et al.* Transoesophageal echocardiography in the detection and surgical management of a papillary fibroelastoma of the mitral valve causing partial mitral valve obstruction. *J Am Soc Echocardiogr* 1993;**6**:83–6.
- 37 Schwinger ME, Katz E, Rotterdam H, *et al.* Right atrial papillary fibroelastoma: diagnosis by transthoracic and transoesophageal echocardiography and percutaneous transvenous biopsy. *Am Heart J* 1989;**118**:1047–50.
- 38 Gallo R, Kumar N, Prabhakar G, *et al.* Papillary fibroelastoma of mitral valve chorda. *Ann Thorac Surg* 1993;**55**:1576–7.
- 39 Lewis NP, Williams GT, Fraser AG. Unusual and intra-operative epicardial echocardiographic features of a papillary tumour of the aortic valve. *Br Heart J* 1989;**62**:470–4.