Left atrial myxoma: new perspectives in the diagnosis of murmur free cases

PETROS NIHOYANNOPOULOS, PRADHIB VENKATESAN, JOEL DAVID, DAVID HACKETT, HANNAH VALANTINE, CELIA M OAKLEY

From the Department of Medicine (Clinical Cardiology and Rheumatology), Royal Postgraduate Medical School, Hammersmith Hospital, London

SUMMARY No cardiological signs or symptoms were seen in three patients with left atrial myxomas. The diagnosis was established by cross sectional echocardiography. In two patients this investigation was performed to exclude a cardiac source of systemic emboli. The third patient had constitutional signs only. All three had raised erythrocyte sedimentation rates and C reactive protein concentrations. In two patients the myxomas were successfully excised; the third patient who had presented with massive peripheral and central embolisation died during emergency operation.

Cross sectional echocardiography is the technique of choice for detecting atrial myxomas and the absence of the cardiological signs should not preclude referral for diagnostic echocardiography.

Left atrial myxoma is the most common primary cardiac tumour and is one of the few for which operation offers a complete cure. Clinical findings are variable and non-specific and may mimic almost any other cardiovascular disorder. Before echocardiography became available, diagnosis of left atrial myxoma, if suspected, was made by angiography or at necropsy or was an unexpected finding during cardiac operation. Cross sectional echocardiography has increased the number of myxomas that are diagnosed before operation.

We report the clinical presentation, diagnosis, and management of three patients with left atrial myxoma seen over the past 12 months. These patients are representative of a wide spectrum of manifestations, the true cause of which may be missed when the myxoma does not produce a murmur.

Patients and methods

CASE 1
A 48 year old man with chronic aortic regurgitation

was first seen at the Hammersmith Hospital at the age of 39 when he complained of lethargy and general malaise. M mode echocardiography and cardiac catheterisation suggested that the aortic regurgitation was only moderate and that aortic valve replacement could safely be deferred. An erythrocyte sedimentation rate of 50 mm in the first hour was also noted but a bacteriological search for endocarditis was negative.

Over the next six years the patient had continued constitutional symptoms and further enlargement of the heart was noted. At age 44 years his aortic valve was replaced with a Starr-Edwards prosthetic valve.

Surgical findings were consistent with the diagnosis. The aortic root was dilated and prevented competent apposition of the otherwise normal valve cusps. Although the postoperative period was uneventful, the patient continued to complain of lethargy and fatigue. There was no weight loss and numerous blood cultures and serological tests were all negative. Serum protein electrophoresis and quantitative immunoelectrophoresis showed a moderate increase in the \(\alpha_1\) and \(\gamma\) region which was polyclonal with no paraprotein. The concentration of C reactive protein was raised (48 mg/l; normal range for this laboratory <15 mg/l). The complement profile was normal and there were no heart muscle antibodies or rheumatoid factor. Six months later at
**Left atrial myxoma: new perspectives in the diagnosis of murmur free cases**

a routine visit for anticoagulation control he was found to have an erythrocyte sedimentation rate of 100 mm in the first hour. Cross sectional echocardiography showed an almost immobile mass the size of a tennis ball that was attached to the posterior left atrial wall (fig 1). A left atrial myxoma, 10 cm in diameter, was excised at operation. It had a broad (2 cm in diameter) very short stalk attached to the free wall of the left atrium adjacent to the superior pulmonary veins. This part of the atrial wall was also excised. Six months after operation the patient was fit and had returned to work.

**CASE 2**

In March 1979 a 41 year old woman, who had previously been well, had a fall associated with loss of consciousness. Over the next three years she was admitted to hospital on 25 occasions because of transient episodes of loss of consciousness. She suffered three episodes of hemiparesis involving the right side on one occasion and the left side twice. In April 1982 investigations including bilateral carotid arte-

riography, cerebrospinal fluid examination, computed tomography, and M mode echocardiography were all normal. Diagnosis included epilepsy, migraine, and hysteria. Several therapeutic treatments were tried without benefit. In August 1983 the patient attempted suicide.

In June 1984 she was referred to Hammersmith Hospital. Cardiovascular examination was unremarkable. Her mental state was normal but there was mild dysarthria and a spastic quadriaparesis. The erythrocyte sedimentation rate was 45 mm in the first hour, haemoglobin 12.2 g/dl, and the biochemical profile was normal. The total protein concentration was 61 g/l, albumin was 41 g/l, and immunoglobulins were normal. C reactive protein was raised (55 mg/l); there was no complement consumption. Tests for rheumatoid factor and auto-antibodies were negative. Chest x rays and the electrocardiogram were normal. Cross sectional echocardiography showed a relatively large highly mobile mass attached to the posterior left atrial wall and protruding into the left ventricle in diastole (fig 2). A left atrial myxoma was diagnosed and in June 1984 the tumour was successfully excised. The left atrium contained a 4 cm diameter classic myxoma of the very friable frogs egg type (fig 3). It was attached by a thin friable stalk to the left atrial wall, just below the right inferior pulmonary vein. Both the atrioventricular valves were normal. The post-operative period was uneventful.

**CASE 3**

A 33 year old man was admitted from casualty in a comatose state after a sudden collapse. Fundoscopy showed attenuation of the blood vessels of the left eye but no papilloedema or haemorrhages. Cardiovascular examination was normal and peripheral pulses were all present. Laboratory data showed a white blood count of 19·4 x 10⁹/l and the erythrocyte sedimentation rate was 75 mm in the first hour. The total serum protein concentration was 76 g/l. Serum protein electrophoresis and quantitative immuno-electrophoresis showed a small increase in the γ region; IgA and IgM concentrations were normal. The IgG concentration was slightly raised with no paraprotein band. The concentration of C reactive protein was raised (38 mg/l). Cerebrospinal fluid was normal with a pressure of 14.5 cm. Chest and skull x rays, electrocardiogram, and a tomographic scan of the brain were all normal.

Six hours later the patient was noted to have cold and pale legs with absent pulses. Cross sectional echocardiography was then performed and a rather small but highly mobile mass attached to the left side of the atrial septum was identified (fig 4). Urgent embolectomy via the right femoral artery recovered

---

**Fig 1** Cross sectional echocardiographic still frame image from the apical four chamber view of patient 1, showing a firm left atrial mass, remaining impacted in the left atrium during diastole with no clear determination of point of attachment. LV, left ventricle; M, myxoma; RA, right atrium; RV, right ventricle.
Fig 2  Cross sectional echocardiographic image from the apical four chamber view of patient 2. Sequential views throughout the cardiac cycle demonstrate the myxoma (arrows) within the left atrium (a, b), engaging the mitral valve (c), and protruding into the left ventricle in diastole (d, e, f).

Fig 3  Photograph of the myxoma removed from patient 2. It measured 4.0 x 3.0 cm.
Left atrial myxoma: new perspectives in the diagnosis of murmur free cases

Fig 4  Echocardiogram taken from the apical four chamber view in diastole (a) mid-systole (b), and late systole (c) in patient 3. The small myxoma (M) moved along the left atrial side of the atrial septum without engaging the mitral valve. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Discussion

Atrial myxomas can mimic almost any cardiovascular disorder and they may produce symptoms and signs that direct attention away from the heart.9-16 The clinical features can best be considered as obstructive, resulting from occlusion of the mitral valve orifice, embolic, or constitutional. The diagnosis of left atrial myxoma should be strongly suspected when this triad is present11; however, when as in our patients there are no signs of left ventricular inflow tract obstruction the diagnostic search may be directed away from the heart.

There are two distinct forms of cardiac myxomas.17 The first form is semitransparent gelatinous with a frog spawn appearance (patients 2 and 3) and may give rise to multiple systemic emboli. As a consequence such a myxoma may never become large

a saddle embolus consisting of myxomatous tissue. The arteries to both legs were occluded. A 55 cm stratified embolus with alternating layers of myxoma tissue and thrombus was removed from the right femoral artery. Because of severe brain damage thoracotomy was not attempted and the patient subsequently died.

Necropsy confirmed the presence of embolised myxoma tissue and thrombus both in the legs and carotid arteries. There was a sessile left atrial myxoma, which measured 2.5 cm in its largest diameter, attached to the fossa ovalis (fig 5). The mitral valve showed roughening and thickening consistent with erosion by a much larger tumour. Microscopy of the tumour showed that it was avascular and relatively hypocellular with a very loose matrix and without any definite capsule; this meant that the tumour was extremely friable (fig 6).
Fig 5  Postmortem photograph of the myxoma in patient 3 attached to the fossa ovalis. The myxoma measured 2.5 x 1.0 cm.

Fig 6  Photomicrograph of a section of the myxoma in patient 3 showing the lack of vascularisation and the generally hypocellular consistency of the tumour.
Left atrial myxoma: new perspectives in the diagnosis of murmur free cases

enough to produce haemodynamic effects or clinical murmurs. The second type of myxoma is much firmer and usually produces signs of left ventricular inflow tract obstruction that mimic mitral stenosis. The myxoma in patient 1 belonged to the second category but did not produce a murmur because the pedicle was too short and the tumour too large to engage the mitral valve orifice. The myxoma could have been present for a long time and may have been missed by both M mode echocardiography and cardiac catheterisation nine years before the correct diagnosis was made. In both types of myxoma constitutional disturbances may occur in association with anaemia, an increased erythrocyte sedimentation rate, and a variable increase in the concentration of plasma immunoglobulins. One explanation is that the microemboli from the myxomatous tissue, which contain mucopolysaccharides, may function as polyclonal activators of B lymphocytes irrespective of the immunoglobulin receptor on the B cell surface. The presumed mitogenic signal for T cell dependent antigens would be generated through the interaction of T cells with the antigen held on the surface of the B cell. The autoantibodies produced reflect the characteristics of the population of myxoma cells and their fragments which react with the polyclonal activator. Other explanations such as impaired immune self-tolerance might also be feasible. All our patients had constitutional disturbance in association with high erythrocyte sedimentation rate, anaemia, and raised concentrations of C reactive protein.

Cross sectional echocardiography has made a great impact on the diagnosis of intracardiac masses and is more sensitive than M mode echocardiography. Its diagnostic accuracy arises from the ability to examine all four intracardiac chambers in several planes. This has made cardiac catheterisation, which is potentially hazardous and delays surgery, both unnecessary, and unjustifiable. Moreover, unless a myxoma is specifically sought, “routine” right and left ventriculography may not reveal the tumour. This happened in the case of patient one. In patients 1 and 2 the diagnosis would have been further delayed or would not have been made before necropsy if echocardiography had not been performed.

Although cross sectional echocardiography permits the accurate determination of tumour size, the point of attachment, mobility, and consistency of the intracardiac masses, these features are not always enough to characterise the great variety of cardiac tumours or distinguish them from thrombi. In patient 1 the left atrial mass was not typical of a myxoma in that it was firm and sessile, not deformable, and only moved slightly during the cardiac cycle. For these reasons it had been clinically silent. Fyke et al recognised similar echocardiographic appearances in five of their 20 patients with left atrial myxomas; others have reported cases of malignant atrial tumours simulating myxoma echocardiographically.

Surgical excision of cardiac myxomas should be performed without delay. In the Hammersmith experience there have been no recurrences. Recurrences have been reported after incomplete excision or growth from a second pretumorous focus. Excision should therefore include a full thickness of atrial septum or wall surrounding the pedicle attachment and the defect created should be patched.

In conclusion, cross sectional echocardiography remains the technique of choice for demonstrating the presence of left atrial myxoma. A high degree of clinical suspicion, extending beyond patients who present with specific cardiological signs, is needed to ensure that these patients are referred for echocardiography. The occurrence of systemic emboli or the presence of constitutional signs with raised erythrocyte sedimentation rate and C reactive protein of unknown aetiology should be regarded as an indication for echocardiography.

References


Left atrial myxoma: new perspectives in the diagnosis of murmure free cases.
P Nihoyannopoulos, P Venkatesan, J David, D Hackett, H Valantine and C M Oakley

Br Heart J 1986 56: 554-560
doi: 10.1136/hrt.56.6.554

Updated information and services can be found at:
http://heart.bmj.com/content/56/6/554

These include:
Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/