Case Report

An unusual presentation of atrial myxoma in an elderly patient: a case report

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Abstract

Left atrial myxoma is the most common intracardiac tumour. It could be seen in patients between 3–83 years of age, with the majority presenting in fifth decade of life as sporadic cases (90%) and second decade as familial cases (10%) [1]. It is an important source of central nervous system embolism [2]. Elderly patients often present with non specific symptoms that are often overlooked in the absence of a supporting cardiac history which makes an early diagnosis challenging. This case report discusses an unusual presentation of left atrial myxoma in an elderly patient.

Case presentation

A 77 year old lady presented with a two week history of shortness of breath and acute onset palpitations and chest pain. She had also developed an acute confusional state for two days prior to her admission. She had been admitted five months ago with an episode of chest infection which was successfully treated with antibiotics. In the previous four years she had suffered from an episode of transient ischaemic attack (TIA).

Previous investigations had shown a normal computer tomography (CT) scan of the head, active rheumatoid arthritis and osteoarthritis. On this occasion she was afebrile, normotensive and had an irregularly irregular pulse rate of 150 bpm. Further cardiovascular, respiratory and abdominal examination was unremarkable and a detailed neurological examination did not reveal any significant abnormality.

An electrocardiogram showed fast atrial fibrillation (AF) for which she was treated with digoxin, warfarin & beta blocker for further optimisation. An in-patient trans-thoracic echocardiogram (TTE) demonstrated a mobile mass in the left atrium (figure 1). Subsequent trans-oesophageal echocardiography (TOE) revealed a mobile large left atrial mass (2.7 cm × 3.5 cm) attached to the fossa ovalis region in the inter-atrial septum, prolapsing into the left ventricle through the mitral valve in diastole (figure 2 &3).

The patient declined surgical treatment in view of the risks involved, & died a few weeks later whilst on conservative therapy.

Discussion

Atrial myxoma is the commonest (20–30% of all) primary intra-cardiac tumour in adults and two thirds of these arise in the left atrium [3]. Other locations are right atrium (next commonest), ventricles, superior vena cava or pulmonary veins. In 5 percent of cases myxomas can be multiple. Differential diagnosis [4] of atrial myxoma includes pedunculated thrombus, metastatic sarcoma and melanoma in left atrium. Other metastatic tumors could specifically metastasize via inferior vena cava to the right
side of the heart & include hypernephroma, hepatoma, melanoma and intravenous leiomyomatosis from uterus.

Left atrial myxoma is most commonly seen in women with 90% being solitary and pedunculated and 10% being familial, with an autosomal dominant pattern of inheritance [5]. The mean age of onset is between 30–60 years.

Most myxomas produce symptoms when they weigh greater than seventy grams. The presentation of atrial myxoma can in three different ways:

- **Obstructive symptoms** – dyspnea, cardiac failure, dizziness, collapse & syncope due to obstruction of the mitral valve.
- **Constitutional symptoms** – i.e. symptoms of autoimmune disease, vasculitis and various other non specific symptoms.
- **Embolic symptoms** – most frequently being cerebral emboli [6].

TOE has nearly 100% sensitivity for cardiac myxoma. Atrial myxoma is usually seen at the border of the fossa ovalis in the left atrium, attached to the inter-atrial septum as in this case. The tumor tissue manifests as spherical/pedunculated mass attached to the endocardial surface with hypoechoic areas [7]. TTE has less specificity than the TOE. Contrast CT demonstrates a well defined spherical or ovoid intracavitary mass. Magnetic resonance imaging (MRI) can visualise the point of attachment and helps differentiate a thrombus from a tumour. Differences in signal intensity between myocardium, tumor/thrombus is very helpful, especially with the use of contrast agent like Gadolinium-DTPA [4]. A cine MRI sequence is a very sensitive technique to distinguish between an thrombus and a tumor, intra-cardiac or intravascular. Surgical excision is the only definitive treatment for atrial myxoma. In relatively small tumors, TTE/TOE can be used to monitor the growth of the tumor, to decide the timing of the surgery [8]. Conservative management is of limited value in symptomatic patients with large myxomas. However, a conservative strategy with TTE/TOE monitoring, & anticoagulation is favoured in high operative risk patients,
asymptomatic patients, and slow growing atrial myxomas.

The diagnosis of atrial myxoma can be elusive, especially when symptoms are suggestive of other diagnoses. In this case, the significance of this patient's past medical history of a transient ischemic attack only became apparent when the patient presented with new symptoms of AF, which led to various investigations looking for a source of cardio- genetic cerebral embolism, eventually revealing the left atrial myxoma. This seems to be an unusual case due to the age at presentation of the patient. Left atrial myxoma presenting in seventh decade is rare, with only few published case reports with this one of its first kind in the UK in the last ten years. Bire et al [9] studied the number of myxoma cases in patients over 75 years of age between 1962 and 1997 and found only 19 confirmed cases.

Consent
Written informed consent was obtained from the deceased patient's next of kin for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing Interests
The authors declare that they have no competing interests.

References
Cardiac myxomas: Clinical and echocardiographic profile

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Abstract

We reviewed our clinical and echocardiographic experience in 70 consecutive patients with 73 cardiac myxomas, diagnosed over an 11 year period. There were 21 males and 49 females, ages ranged from 18 to 80 years. Only in 5.7% cases was the diagnosis of myxomas made clinically. 88.6% cases were initially diagnosed as having: mitral valve disease (70%), tricuspid valve disease (10%), ischemic heart disease (5.7%), cardiomyopathy (2.9%), and the remaining 5.7% were detected during family screening and follow-up. The mean duration of symptoms was 10.6 months. The commonest symptom was dyspnoea (80%), followed by constitutional symptoms (45.7%), embolization (30%), palpitation (25.7%), syncope (15.7%), pedal oedema (15.7%) and pain chest (12.9%). The sites of myxomas were as follows: left atrium, 58; right atrium, 9; and, biatrium, 3. All myxomas except 3 were attached to the interatrial septum. The site, size, shape, attachment, mobility, prolapse into ventricle, and surface characteristic of myxomas were accurately assessed by 2D-echocardiography and confirmed in all (65 of 70) who underwent surgery. When the morphological characteristic of myxomas were studied and correlated with clinical features large left atrial myxoma size was closely related with constitutional symptoms, congestive heart failure, with syncope and auscultatory findings suggestive of mitral valve disease, whereas smaller myxoma size and irregular surface were associated with embolization. Constitutional symptoms were only present in left atrial myxoma. Post-operative mean echocardiographic follow-up of 60 months showed no recurrence except in 2 with familial myxoma. We conclude that the majority of myxomas mimic many cardiovascular diseases and were detected in symptomatic patients, so a high index of clinical suspicion is important for its early and correct diagnosis. The size and appearance of the myxomas correlated with the presenting symptoms. © 1998 Elsevier Science Ireland Ltd.

Keywords: Cardiac myxomas; Echocardiography

1. Introduction

Cardiac myxomas are uncommon [1], but surgical treatment offers a potential cure [2–4]. Early diagnosis is essential so that tumor can be promptly removed in order to reduce morbidity or mortality from atrio-ventricular valve dysfunction or embolic complications. Because of varied clinical manifestations, cardiac myxomas usually mimic many cardiovascular or systemic diseases [2,5–7]. Unless the physician has a high index of suspicion, the clinical diagnosis is usually missed, resulting in morbidity or mortality, in a otherwise curable disease. Once a diagnosis is suspected, it is easily confirmed by 2D-echocardiography. In this report we review the clinical and echocardiographic experience with cardiac myxomas at our centre and delineate the clinical features that allow the physician to suspect this rare fatal disease and to compare the echocardiographic findings with surgical findings and to correlate the
Case report

Atrial myxoma presenting with orthostatic hypotension in an 84-year-old Hispanic man: a case report

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Abstract

Introduction: Left atrial myxomas remain the most common benign primary cardiac tumors, and these cardiac growths can masquerade as mitral stenosis, infective endocarditis and collagen vascular disease. Atrial myxomas are found in approximately 14-20% of the population and can lead to embolization, intercardiac obstructions, conduction disturbances and lethal valve obstructions.

Case presentation: An 84-year-old Hispanic man presented with complaints of dizziness upon standing, and with no prior history of heart murmurs, syncope, shortness of breath, or chest pain. Physical examination revealed evidence of orthostatic hypotension and a soft grade 1/6 systolic murmur at the left sternal border. A transthoracic echocardiogram revealed a large atrial myxoma occupying the majority of the left atrium, with the posterior border of the large atrial mass defined by eccentric mitral regurgitation identified during cardiac catheterization. Left atrial myxoma excision was performed, revealing a 7 × 6.5 × 4.5 cm atrial tumor attached to a 4 × 3 × 2 cm stalk of atrial septal tissue.

Conclusion: This patient didn’t present with the common symptoms associated with an atrial myxoma, which may include chest pain, dyspnea, orthopnea, peripheral embolism or syncope. Two-dimensional echocardiography provides substantial advantages in detecting intracardiac tumors. We recommend a two-dimensional echocardiogram in the workup of orthostatic hypotension of unknown etiology after the common causes such as autonomic disorders, dehydration, and vasodilative dysfunctions have been ruled out. By illustrating this correlation between orthostasis and an atrial myxoma, we hope to facilitate earlier identification of these intracardiac growths.
Introduction

Although quite rare, left atrial myxomas account for 80% of all cardiac tumors. Diagnosis is often difficult due to the wide array of presenting symptoms. Atrial myxomas are associated with systemic embolization in 30 to 40% of cases [1]. These intracardiac growths may masquerade as mitral stenosis, infective endocarditis, and collagen vascular disease, which can further impede accurate diagnosis. The discriminatory marker for an atrial myxoma is often a tumor ‘plop’ heard upon auscultation at the apex of the heart.

We present the case of an 84-year-old man with a large atrial myxoma, who presented with complaints of positional dizziness and who was found to have a grade 1/6 systolic murmur, and significant orthostatic hypotension.

Case presentation

An 84-year-old Hispanic man presented with complaints of dizziness upon standing, which was relieved by lying down. Physical examination revealed a drop in the patient’s blood pressure from 124/80 mmHg supine to 99/70 mmHg one minute after standing. Pulse rate during the examination remained static. The patient had no prior history of heart murmurs, syncope, shortness of breath, or chest pain. Further physical examination revealed a soft grade 1/6 systolic murmur at the left sternal border, with no diastolic murmur present. There was no evidence of a tumor ‘plop’.

A transthoracic echocardiogram was performed that revealed a large atrial myxoma occupying the majority of the left atrium. Cardiac catheterization showed eccentric mitral regurgitation, defining the posterior border of the large atrial mass. Transesophageal echocardiography, carried out at the time of surgery, revealed a large myxoma prolapsing through the mitral valve leaflets into the left ventricle (Figure 1).

A left atrial myxoma excision was performed, resulting in successful removal of the tumor. Pathological analysis of the atrial mass revealed it to be 7 × 6.5 × 4.5 cm attached to a 4 × 3 × 2 cm stalk of atrial septal tissue (Figure 2). Four weeks postoperatively, the patient stated that the original complaint of ‘dizziness upon standing’ had disappeared, with no evidence of orthostatic hypotension during a follow-up physical examination. A follow-up echocardiogram showed no evidence of atrial myxoma recurrence, and the mitral valve leaflets separated normally without regurgitation.

Discussion

Our patient failed to present with the common symptoms associated with atrial myxoma including chest pain, dyspnea, orthopnea, peripheral embolism or syncope. Though cardiac myxomas are known to present with various non-specific clinical symptoms [2], orthostatic hypotension is not listed as a presenting symptom of atrial tumors in most textbooks of internal medicine or cardiology [3,4]. An extensive literature search revealed one case that reported orthostasis as a presenting symptom of a left atrial myxoma [5]. The patient in that case report had a principal complaint of dizziness upon standing, and orthostasis was observed with a blood pressure change from 90/50 mmHg supine to 64/40 mmHg standing. Upon echocardiographic investigation, a large atrial
Changing Management of Cardiac Myxoma Based on a Series of 40 Cases With Long-Term Follow-Up

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Background. Cardiac myxoma is generally considered to be a surgical emergency. However, as a result of progress in echocardiography and the increasing age of the patients presenting with this disease, the clinical presentation has changed and the management of cardiac myxoma now needs to be reviewed.

Methods. Between 1978 and 2001, 40 patients (16 men and 24 women) between the ages of 6 months and 82 years (mean age, 55.6 years) were operated on for cardiac myxoma. Signs of heart failure with pulmonary congestion (22%) or pulmonary embolism (20%) indicated a high-risk emergency situation in some cases, whereas, in other cases (58%), the patient’s condition was stable and the clinical presentation was less worrying. However, the tumor was always removed within 24 hours of admission. Most cases of cardiac myxoma observed over the last decade correspond to stable forms, as echocardiography has revealed smaller tumors in generally elderly patients.

Results. The postoperative mortality was 7.5% (3 patients). No patients were lost to follow-up, and the mean follow-up was 13.6 years. One patient was reoperated for recurrence 3 years postoperatively. Five patients required further cardiac surgery: three mitral valve replacements, one coronary artery bypass graft, and one angioplasty. The 15-year survival rate was 69%.

Conclusions. Myxoma tends to be observed in a more elderly and higher risk population, often at an early stage. The classic approach of emergency surgery is not always appropriate in these stable forms, allowing more thorough preoperative assessment of these patients.


Cardiac myxoma is usually considered to be a definite indication for urgent surgery. This is obviously true in certain symptomatic forms, such as cases presenting with embolism or heart failure. However, the development of echocardiography now allows more precise diagnosis, so that these severe forms are in now less frequently observed. We therefore wanted to determine whether, on the basis of our own experience and a review of the literature, this aggressive attitude is still appropriate.

Material and Methods

Between September 1978 and September 2001, 40 patients (16 men and 24 women) between the ages of 6 months and 82 years (mean age, 55.6 years) were operated on for cardiac myxoma. Signs of heart failure with severe dyspnea due to pulmonary congestion (22%) or pulmonary embolism (20%) indicated a high-risk emergency situation in some cases, whereas, in other cases (58%), the patient’s condition was stable and the clinical presentation was less worrying. However, the tumor was always removed within 24 hours of admission. Most cases of cardiac myxoma observed over the last decade correspond to stable forms, as echocardiography has revealed smaller tumors in generally elderly patients.

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factors. Relatively noninvasive surgical techniques were used, with simple resection in 87% of patients (ie, avoiding large resection of the atrial septum below the tumor implantation). Extensive resection therefore appears to be mainly justified in high-risk cases [15]. Our one case of recurrence, not presenting any particular risk factors, was probably due to incomplete resection.

However, our series comprised a significant number (6/40) of concomitant mitral valve procedures, plus another three cases of secondary mitral valve replacement. Similar results were reported in another series of 22 other three cases of secondary mitral valve replacement. Kabbani and associates [17] reported four mitral valve replacements in a series of 24 tumor resections and Semb [18] reported four cases of mitral valve insufficiency in a series of 11 patients and one tricuspid valve replacement in two cases of right cardiac myxoma. The valves should therefore be very thoroughly examined during initial surgery to lower the reoperation rate. A conservative mitral procedure, such as annuloplasty, would no doubt be feasible as part of the first step, and transoesophageal echocardiography should be systematically performed. In this abundant literature [15, 19], the classic attitude of emergency management of cardiac myxoma is rarely questioned [20]. This approach is perfectly logical in the case of acute symptomatic forms, such as heart failure with pulmonary edema, or embolism [21]. It is also justified in high-risk forms in stable patients presenting threatening echocardiographic images: a clapper-shaped tumor intermittently prolapsing into the mitral orifice, or a large, multinodular tumor suggesting a risk of embolism. However, emergency management appears to be much less clearly indicated in stable patients, in whom the only real risk is that of embolism [22], as the risk of embolism is probably low for tumors less than 2 cm in diameter. All 8 patients in our series presenting with embolism had tumors larger than 3 cm. There is little mention in the literature about the correlation between size and risk of embolism, but, to the best of our knowledge, no case of embolism has ever been reported for small tumors. Such small tumors are no longer rare with the development of echocardiography (Table 1), which questions the dogma of emergency surgery, proposed at a time when acute symptomatic forms were the most frequent. Moreover, cardiac myxoma, initially described as a disease of early adulthood or middle age, is now increasingly diagnosed in sometimes very elderly patients [Fig 2]. This trend also applies to cardiac surgery in general, which tends to concern an aging population with specific problems. In these older patients with a higher operative risk, more delayed surgery allows time for a more complete preoperative assessment, including more systematic coronary angiography. With the exception of real emergency situations, there is no reason why surgery for cardiac myxoma should not comply with the usual recommendations for preoperative coronary angiography before any form of cardiac surgery. Coronary angiography was performed in only 4 patients with a history of angina in this old series. In the 14 patients over the age of 65 years, two coronary revascularisations were performed, 6 months and at 2.5 years after tumor resection, but these cases could have been detected by preoperative coronary angiography. For all of these reasons, we therefore consider that emergency surgery is only indicated in patients with severe dyspnoea and presenting a high risk of embolism (ie, with a history of embolism or with large, mobile tumors on echocardiography). In all other cases, preoperative assessment should be performed to allow better patient preparation and to detect any associated coronary or valvular heart disease. This assessment should only take 1 or 2 days, which does not constitute a high risk for stable patients, but allows surgery to be performed under better conditions, particularly in elderly patients. Finally, in some exceptional cases in very debilitated patients, surgery may even be unnecessary, as illustrated by the case of an 86-year-old patient with a nonoperated small myxoma, who was followed for 4 years without incident [20].

Conclusion

The circumstances of discovery of cardiac myxoma have changed over recent decades. They are more often detected at an early stage, but also in a more elderly and higher-risk population. Emergency surgery is still appropriate in patients with severe dyspnoea or a high risk of embolism, but in all other cases, a more complete preoperative assessment is recommended to allow better patient preparation and to detect any associated coronary or valvular heart disease. Surgery could therefore be more frequently performed with a minimally invasive approach with video-thoracoscopy.

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References

CARDIAC MYXOMAS

KLAUS REYREN, M.D.

Primary tumors of the heart are rare, with an incidence between 0.0017 and 0.19 percent in unselected patients at autopsy. Three quarters of the tumors are benign. Nearly half the benign heart tumors are myxomas, and the majority of the rest are lipomas, papillary fibroelastomas, and rhabdomyomas. Fibromas, hemangiomas, teratomas, and mesotheliomas of the atrioventricular node are found less frequently; granular-cell tumors, neurofibromas, and lymphangiomas are very rare. Whereas rhabdomyomas are the most common primary tumors of the heart in children, myxomas clearly predominate in adults.

A left atrial myxoma was first described in 1845. Before 1951, the diagnosis of intracardiac tumors was made only at postmortem examination; in that year the diagnosis of an intracavitary left atrial tumor was confirmed by angiocardiography. The first successful excision of a left atrial myxoma was reported in 1955. The introduction of echocardiography has greatly facilitated the antemortem diagnosis of cardiac tumors.

Supplemental diagnostic imaging methods include computed tomography (CT) and nuclear magnetic resonance imaging (MRI).

Epidemiology

Myxomas occur in all age groups but are particularly frequent between the third and sixth decades of life. The youngest known patient was a stillborn infant, and the oldest a 95-year-old woman. Women predominate in most series. Myxomas usually occur sporadically, but familial myxomas have been reported. “Complex” cardiac myxomas are those that occur in families, in combination with two or more of the following conditions: skin myxomas (single or multiple), cutaneous lentiginosis, myxoid fibroadenomas of the breast, pituitary adenomas, primary adrenocortical micronodular dysplasia with Cushing’s syndrome, and testicular tumors (characteristically, large-cell calcifying Sertoli-cell tumors).

At the time of the diagnosis, patients with familial myxomas are usually considerably younger than those with nonfamilial myxomas. Family studies suggest an autosomal dominant pattern of inheritance with a variable phenotype.

Location

Cardiac myxomas usually develop in the atri. About 75 percent originate in the left atrium, and 15 to 20 percent in the right atrium. Most myxomas arise from the interatrial septum at the border of the fossa ovalis, but they can also originate, in descending order of frequency, from the posterior atrial wall, the anterior atrial wall, and the atrial appendage. Only 3 to 4 percent of myxomas are detected in the left ventricle, and only 3 to 4 percent in the right. Myxomas are rarely multilocular, but cases of several myxomas within one atrium have been reported, as well as bialtral myxomas. In such cases, the tumor usually extends through the foramen ovale into the contralateral atrium. There have also been reports of combined atrial and ventricular tumors and biventricular tumors. Multiple tumors and atypical locations are more frequent in cases of familial myxoma.

Pathological Findings

Myxomas are neoplasms of endocardial origin. The tumor usually projects from the endocardium into the cardiac chamber. The cells giving rise to the tumor are considered to be multipotential mesenchymal cells that persist as embryonal residues during septation of the heart and differentiate into endothelial cells, smooth-muscle cells, angioblasts, fibroblasts, cartilage cells, and myoblasts. The prevalence of myxomas in the atrial septum is therefore understandable. The rate of growth of myxomas is unknown, but they generally appear to grow rather quickly. There is one report, however, of a left atrial myxoma that did not change in its appearance during a period of 28 months.

The malignant potential of cardiac myxomas remains doubtful, but there are a few reports of the remote growth of myxomatous material that has embolized.

Macroscopic Appearance

Myxomas are generally polypoid, often pedunculated, rarely sessile, and round or oval, with a smooth or gently lobulated surface (Fig. 1). The mobility of the tumor depends on its consistency, which varies with the amount of collagen, the extent of attachment, and the length of the stalk. Polypoid myxomas are usually compact and show little tendency toward spontaneous fragmentation. The less common villous or papillary myxomas have a surface that consists of multiple fine or very fine villous extensions (Fig. 2). These extensions are gelatinous and fragile and tend to break off or into pieces. Myxomas can appear white, gray–white, yellow-
require permanent cardiac pacing because of atrioventricular conduction disturbances.35,124

RECURRENT MYXOMA

Recurrences of myxomas, including second recurrences, have been observed.12,20,46,121,122,125-127 Since an initial report in 1967,121 more than 35 recurrent myxomas have been reported. Most were diagnosed during the first 4 years after surgical removal of the first tumor, but the tumor recurred in two cases after 10 and 14 years.125 Incomplete resection, intraoperative displacement of tumor material, embolization, transformation from a benign to a malignant lesion, and — above all — multifocal genesis have been proposed as possible explanations.27,46,122,125,126 Multifocality may explain the high rate of recurrence and the possibility of remote21,125,126 or multilocular47,125,128 recurrence in cases of familial myxoma. In most cases of sporadic myxoma, however, incomplete resection is thought to be the reason for a recurrence.27,125,126 The overall risk of recurrence is about 12 and 22 percent for familial and complex myxomas, respectively, whereas it is only 1 to 3 percent for sporadic tumors.27,126 Semiannual echocardiographic follow-up examinations are indicated in all cases.

SUMMARY

Although cardiac myxomas are histologically benign, they may be lethal because of their strategic position. They can mimic not only every cardiac disease but also infective, immunologic, and malignant processes. Myxomas must therefore be included in the differential diagnosis of valvular heart disease, cardiac insufficiency, cardiomegaly, bacterial endocarditis, disturbances of ventricular and supraventricular rhythm, syncope, and systemic or pulmonary embolism. The symptoms depend on the size, mobility, and location of the tumor. Echocardiography, including the transesophageal approach, is the most important means of diagnosis; CT and MRI may also be helpful. Coronary arteriography in patients over 40 years of age is generally required to rule out concomitant coronary artery disease. Surgical removal of the tumor should be performed as soon as possible; the long-term prognosis is excellent, and recurrences are rare. In follow-up examinations as well, echocardiography is essential.

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**Background**

The cardiac myxoma is a benign neoplasm arising within the heart. It is sometimes referred to as an atrial myxoma, since the vast majority arise within the atrial septum. For many years, there was a debate on the origin of this tumor. Some thought it represented an organizing thrombus. Today, it has clearly been shown that this is a true neoplasm.

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Only in 5.7% cases was the diagnosis of myxomas made clinically. 88.6% cases were initially diagnosed as having: mitral valve disease (70%), tricuspid valve disease (10%), ischemic heart disease (5.7%), cardiomyopathy (2.9%), and the remaining 5.7% were detected during family screening and follow-up. The mean duration of symptoms was 10.6 months. The commonest symptom was dyspnoea (80%), followed by constitutional symptoms (45.7%), embolization (30%), palpitation (25.7%), syncope (15.7%), pedal oedema (15.7%) and pain chest (12.9%). The sites of myxomas were as follows: left atrium, 58; right atrium, 9; and, biatrium, 3. All myxomas except 3 were attached to the interatrial septum. The site, size, shape, attachment, mobility, prolapse into ventricle, and surface characteristic of myxomas were accurately assessed by 2D-echocardiography and confirmed in all (65 of 70) who underwent surgery.

When the morphological characteristic of myxomas were studied and correlated with clinical features large left atrial myxoma size was closely related with constitutional symptoms, congestive heart failure, with syncope and auscultatory findings suggestive of mitral valve disease, whereas smaller myxoma size and irregular surface were associated with embolization. Constitutional symptoms were only present in left atrial myxoma. Post-operative mean echocardiographic follow-up of 60 months showed no recurrence except in 2 with familial myxoma.

We conclude that the majority of myxomas mimic many cardiovascular diseases and were detected in symptomatic patients, so a high index of clinical suspicion is important for its early and correct diagnosis. The size and appearance of the myxomas correlated with the presenting symptoms.
CASE REPORT

Management of a left atrial mass in an octogenarian

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Case presentation

An 83-year-old man was referred to outpatients with a 6-month history of some shortness of breath and intermittent palpitation. His past medical history was unremarkable with no history of angina or hypertension. On examination, he was in sinus rhythm with a systolic and diastolic heart murmur audible at the apex. No diagnostic abnormality was identified on either chest X-ray or electrocardiogram. Transthoracic echocardiography demonstrated no abnormality of the heart valves with normal left ventricular systolic function. The left atrium was dilated (5.8 cm) and it contained a smooth mobile mass of 8 x 3 cm thought to be either a left atrial myxoma or left atrial thrombus.

Transoesophageal echocardiography confirmed a pedunculated mass with mixed echo density but smooth contours arising from just above the fossa ovalis (Figure 1). There was no left atrial spontaneous echo contrast (this is a marker of slow blood flow and a tendency to intracardiac thrombus formation). Inflammatory markers were mildly elevated with an erythrocyte sedimentation rate of 45 mm/h and a C-reactive protein level of 25 mg/l.

We discussed management with the patient and explained that the probable diagnosis was left atrial myxoma; surgery was recommended because of the

Figure 1. Two-dimensional echocardiographic image of the mass with its echolucent core (MY). During atrial systole it prolapses through the mitral valve (MV). LA, left atrium. The excised tumour is shown in the adjacent panel.
embolic potential of such a mass. Full routine work up before cardiopulmonary bypass including coronary angiography was felt to be unnecessary—particularly in view of the low likelihood of clinically significant coronary artery disease.

We used a bi-atrial approach using cardiopulmonary bypass to resect a $10 \times 4 \times 4$ cm narrow smooth atrial myxoma. The pedicle, which was above the fossa ovalis in the left atrium, was removed and patched using pericardium. The patient required postoperative cardioversion for atrial fibrillation but made an otherwise uneventful recovery and remains well 9 months later.

**Discussion**

Left atrial myxoma is a rare condition with a variety of presentations. Myxomas represent 25% of all cardiac tumours, with 75% originating in the left atrium and 20% in the right atrium, although any cardiac chamber may be involved [1]. Most arise from the atrial septum, near the fossa ovalis. It affects women more than men with an average age at presentation of 50.

Major clinical symptoms in half to three-quarters of patients are attributable to heart failure, thought to represent obstruction of the mitral valve. Embolization of tumour material is the presenting complaint in one-third, with involvement of the systemic (including the cerebral) circulation [2]. Physical examination may reveal diastolic and or systolic murmurs, with an occasional extra early diastolic sound or ‘tumour plop’, sometimes misinterpreted as the opening snap of mitral stenosis.

The mainstay of diagnosis is two-dimensional transthoracic echocardiography allowing sizing and visualization of the tumour and providing other important information such as left ventricular and valve function. Transoesophageal echocardiography is useful in distinguishing thrombus from myxoma, when the stalk of the tumour may be visualised. Surgical removal using cardiopulmonary bypass and incisions in both atria has a high success rate [3] with an operative mortality in a young patient population of <2% [2]. Myxoma presenting in an octogenarian is extremely unusual. This makes it difficult to advise patients about appropriate management and risk, but the high likelihood of embolization/stroke in our view mandated a limited surgical approach.

**Key points**

- Exertional dyspnoea is common in older patients.
- This case illustrates the value of two-dimensional echocardiography and the importance of establishing a reason for dyspnoea.
- Cardiac surgery in octogenarians is performed increasingly, but the risks and potential benefits must continue to be calculated for each individual.

**References**


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Atrial Myxoma

Introduction

Atrial myxomas are the most common primary heart tumors. Because of nonspecific symptoms, early diagnosis may be a challenge. Left atrial myxoma may or may not produce characteristic findings on auscultation. Two-dimensional echocardiography is the diagnostic procedure of choice. Most atrial myxomas are benign and can be removed by surgical resection.

Pathophysiology

Myxomas account for 40-50% of primary cardiac tumors. Approximately 90% are solitary and pedunculated, and 75-85% occur in the left atrial cavity. Up to 25% of cases are found in the right atrium. Most cases are sporadic. Approximately 10% are familial and are transmitted in an autosomal dominant mode. Multiple tumors occur in approximately 50% of familial cases and are more frequently located in the ventricle (13% vs 2% in sporadic cases).

Myxomas are polypoid, round, or oval. They are gelatinous with a smooth or lobulated surface.
Left Sided Atrial Myxoma with Blood Supply from the Left Circumflex Artery in a 72 Year Old Women

History
This is the case of a 77 year old female patient, who suffered from atypical angina both during exercise and at rest, partially relieved by oral nitrates. Here medical record was unremarkable except for well controlled hypertension treated with an ACE inhibitor for several years. Because of here anginal pain, she was prescribed aspirin 100 mg daily.

Findings
Clinical examination was normal except for a 2/6 mitral regurgitant murmur. Laboratory values were within the normal range except for a moderately elevated ESR (18 mm /h). On echocardiographic examination a 5x3 cm large, smooth and poorly mobile mass in the left atrium was detected (Fig 1), presumably fixed to the atrial septal wall and without signs of obstruction to the pulmonary veins or left atrial outflow. Furthermore, mild eccentric left ventricular hypertrophy was noted (132 g/m2, relative wall thickness 42%) and signs of mild diastolic dysfunction were observed (E/A ratio < 0.8, velocity propagation 42 cm/s). CineMR of the tumor showed a quite homogenous but low signal intensity pattern, compatible with atrial myxoma with calcifications or hemosiderin deposits (Fig 2). Subsequent coronary angiography revealed normal coronary arteries and an abnormal vessel originating from the mid portion of the left circumflex artery supplying the tumor (Fig 3). On operation, the tumor was found to measure 4.5x3x3.5 cm, the consistency was jelly-like and the macroscopic diagnosis was left atrial myxoma. On histology, the tumor could be classified as myxoma not containing calcifications but substantial amounts of hemosiderin.

Discussion
Intracardiac primary tumors are infrequent (0.00017-0.28 per cent incidence on autopsy series) and myxomas are the most common type of primary cardiac tumor, comprising 30-50 per cent in most pathological series. The mean age of sporadic myxomas is 56 years, and 70 per cent are females. Approximately 86 per cent of myxomas occur in the left atrium, and over 90 per cent are solitary with a usual attachment to the fossa ovalis. The clinical signs and symptoms produced by cardiac myxomas include nonspecific manifestations, embolizations, and mechanical interference with cardiac function, but atypical or typical angina are rarely noted.

The vascularity of left atrial myxoma is usually poor, but some myxomas show high vascularity, usually with blood supply from coronary arteries, as noted by Van Cleemput to occur in 7 cases (37 per cent) out of 19 atrial myxomas, of which 3 myxomas (16 per cent) were supplied from the left circumflex artery. Left atrial myxoma blood supply from left circumflex arteries is a rare finding and has, to our knowledge, only been reported in 5 patients since 1988, of which only one patient suffered from atypical angina.

The exact reason for atypical chest pain in our patient could not further be elucidated because of a contraindication for ischemia testing. However, in view of the normal coronary angiogram and only mild left ventricular hypertrophy, a hemodynamic relevant steal phenomenon may have occurred.