

LETTER

Atrial myxoma: histological confirmation

Atrial myxoma is uncommon, with an overall prevalence in the developed world of 0.02%.¹ It is often undiagnosed until complications such as cardiac death or stroke occur. The incidence of primitive tumours of the heart at autopsy is between 0.0017% and 0.03%, and of these 50% are myxomas.^{1,2} We describe a case of left atrial myxoma which was only diagnosed after histopathological examination.

A 69-year-old woman with chronic hypertension and diabetes presented with cardiorespiratory arrest. She was well prior to the acute presentation. She was not a smoker and had no symptoms of malignancy. The family history was unremarkable. She appeared neither cachexic nor cushingoid. She did not have any features suggestive of Carney's complex. She was electively ventilated and successfully resuscitated. The praecordium examination was normal with absence of cardiac murmur or 'plop.' There was generalised crepitation in the lungs. The neurological examination was normal without any localising signs. Her complete blood count showed mild lymphocytosis. Other blood tests were normal except mild liver impairment. Cardiac biomarker was not elevated. Her chest x-ray was consistent with the diagnosis of acute pulmonary oedema. The ECG did not reveal any ischaemia. Her glycaemia control was good, with HbA1c below 6.1%. Fasting serum lipid showed hypercholesterolaemia. An echocardiography revealed a left atrial mass measuring 4.0 cm×3.2 cm. This homogeneously and well-defined mass was seen occupying the left atrium. The chamber's dimensions were within the normal range. We did not detect any pericardial effusion that can cause cardiac tamponade. There was no regional wall motion abnormality to suggest coronary artery disease. The finding led to the provisional diagnosis of atrial myxoma with differential diagnoses of thrombus or metastases. Further investigations to find the association with Carney's complex were normal. The ESR and CRP were elevated. There was mild hypergammaglobulinaemia. Blood cultures did not grow any organisms. The patient was stable and extubated a week later. A cranial computed tomography excluded stroke. The coronary angiogram showed mild coronary artery disease. She was referred for curative surgery. The excised gross specimen showed a 50 mm×40 mm×20 mm lobulated mass weighing about 38 g (figure 1). The histological examination revealed a lobulated polypoidal mass covered by endothelium and composed of abundant loose myxoid stroma with scattered round, polygonal and stellate cells with dense irregular nuclei (figure 2). This confirmed the diagnosis of atrial myxoma. She remained

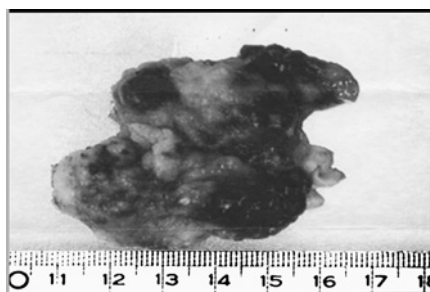


Figure 1 Gross specimen showing the lobulated mass excised from the left atrium measuring 50 mm×40 mm×20 mm. This cut section shows a soft myxoid surface with areas of haemorrhage.

in good health after 2 years of curative myectomy.

Atrial myxomas are endocardial tumours, and about three-quarters of these originate from the left atrium.² They can present variably from non-specific constitutional symptoms to sudden cardiac death. Common cardiac presentations include cardiac failure, syncope, palpitation, thoracic pain, dyspnoea and cough. The clinical signs and symptoms of cardiac myxoma are determined by their location, size, mobility and surface. Occasionally, the clinical signs may not be evident at the time of presentation, as illustrated here. The tumour can mimic mitral stenosis, and occasionally an early diastolic rumbling sound called 'tumour plop' may be heard during auscultation. Diastolic murmurs are the most common murmur heard in up to 89% of patients with cardiac myxoma.³ Non-specific systemic symptoms such as fever, fatigue, arthralgia, myalgia and weight loss are seen in 20–35% of cases.² This is associated with an elevated erythrocyte sedimentation rate and anaemia. These symptoms are mediated by the production and release of interleukin-6 by the myxoma itself.⁴ Following excision, the interleukin-6 levels return to normal, and the systemic symptoms eventually disappear with normalisation of interleukin-6.⁵

Multiple imaging modalities are recommended in patients with a suspected cardiac myxoma.⁶ Two-dimensional echocardiography has adequate sensitivity and specificity to detect the mass, though transoesophageal echocardiography has 100% sensitivity.⁷ When an intracardiac mass is detected from echocardiography, the overwhelmingly probable diagnosis is atrial myxoma; nonetheless, cardiac metastases from other sources should also be considered. Other differential diagnoses include intracardiac thrombus, metastatic neoplasm or large vegetation. Cardiac magnetic resonance imaging shows in detail the location, insertion site and size of the tumour. Tissue characteristics and signs of neovascularisation provide important information if the differentiation between cardiac myxoma and cardiac thrombus is uncertain after echocardiography. Cardiac computed

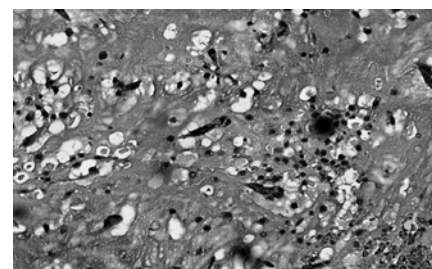


Figure 2 Histopathological examination showing abundant loose myxoid stroma with scattered round, polygonal or stellate cells with dense irregular nuclei. These features are consistent with the diagnosis of atrial myxoma.

tomography or invasive coronary angiography is the investigation of choice to rule out concomitant coronary artery disease or anomalies before surgery.

Surgical resection is the treatment of choice for atrial myxoma with a low post-operative risk. It is usually curative with an excellent long-term prognosis. However, incomplete resection resulted in a recurrence rate of 1–5%. Recurrence beyond 4 years after surgery rarely happens.⁸ However, the rate of recurrence in familial and syndromic atrial myxoma is one in five patients. Family members of patients with familial atrial myxoma and Carney's syndrome should be screened for atrial myxoma. This helps to avoid the consequences of heart failure, sudden cardiac death, arrhythmias, infection and embolisation. Our patient remained in excellent health 2 years after surgery without recurrence.

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Patient consent Obtained.

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