A left atrial myxoma was surgically removed in a 58-year-old man following several embolic events. Five months later, a new myxoma was found in the right ventricular outflow tract and surgically removed. In this patient, we visualized two more recurrences 14 years later, one in the left atrium and the other in the left ventricle. A short review of the literature concerning recurrent cardiac myxomas is given.

### Introduction

Myxoma cordis is the most frequently found primary cardiac tumour. A first recurrence of cardiac myxoma has often been described. However, a second recurrence is rare. The incidence of recurrence is different for the various types of myxoma cordis. Complex and familial forms have a higher incidence of recurrence. Surgical resection is the therapy of choice. In this case report, we present a patient who experienced four cardiac myxomas diagnosed over a period of 14 years.

### Case Report

A 56-year-old man with known chronic obstructive pulmonary disease was admitted for a transient ischaemic attack (TIA) with right-sided hemiparesis in October 1985. In 1986 he underwent surgical embolectomy for acute lower limb ischaemia twice. Histological examination was performed twice and revealed an atherosclerotic plaque with thrombus formation. In December 1986 he was re-admitted for a new TIA with right-sided hemiparesis. Oral anticoagulation therapy was started. An echocardiographic study showed a tumour in the left atrium (LA), inserted on the border of the roof of the LA and the interatrial septum (IAS). The mass was surgically removed with patch closure of the discontinuity in January 1987. Histological examination revealed myxoma cordis. Resection borders were free of tumour tissue. Five months later, a routine control echocardiography was performed. A new mass in the right ventricular outflow tract of $3 \times 3$ cm inserted on the IAS was seen. The mass was surgically removed. The histological examination revealed cardiac myxoma and the resection borders were free of tumour tissue. Echocardiographic control studies without significant abnormalities were performed yearly until 1995. In December 2001, the patient returned at the age of 72 years for a preoperative check for orthopaedic surgery. He was free of complaints but clinical examination revealed mitral regurgitation murmur. The suspected mitral valve insufficiency (grades II/IV) was confirmed by echocardiography but in addition, two tumours were detected, one inserted on the atrial side of the anterior leaflet of the mitral valve with prolapse through the mitral valve and the other in the left ventricular cavity (Fig. 1). A conservative approach was chosen because of the combination of severe chronic obstructive pulmonary disease and the...
unwillingness of the patient to undergo a third operation. Treatment with oral anticoagulant therapy was continued.

Discussion

Myxoma cordis is the most frequent primary heart tumour to be found in humans. Its relative incidence in combined surgical and pathological series is around 42%. When only surgical series is considered, the proportion of myxomas increases to a relative incidence around 77%.[1]

The tumour is a neoplasm of endocardial origin. It consists of stromal cells arising from mesenchymal multipotential cells, which are capable of neural and endothelial differentiation.[2] Although cardiac myxomas are histologically benign, they may be lethal because of their strategic position.[3]

Of all myxomas 10% are familial forms. They are diagnosed more frequently in younger patients and appear to have an autosomal dominant transmission.[4] A complex myxoma occurs in families and is associated with endocrine tumours, myxomas of breast and skin, neurofibromatosis and spotty pigmentation. One of these associations is the Carney Complex. It is linked with abnormalities on chromosome 2.[5,6]

Recurrent cardiac myxoma after surgical excision is not rare. The frequency is estimated by 1–3% in sporadic forms, 12% in familial forms and 22% in complex forms.[7] Second recurrence, however, is rare. Only few cases of a second myxoma recurrence are published so far.[8,9] The re-occurrence of myxoma can be explained by several mechanisms. First, incomplete resection of the original tumour can lead to regrowth. Second, recurrence seems to have a familial predisposition. Third, intracardiac implantation of embolic fragments of the first tumour is possible. A fourth possible explanation for recurrence on the same or another location is the existence of a sort of ‘pretumoural focus’ in the myocardium. The resection borders in our patient were two times free of tumour tissue suggesting complete excision. The left and right locations make intracardiac embolization of the first tumour unlikely. None of the family members have cardiac myxomas or suspected signs of familial cardiac myxoma. Therefore, we think that this patient has a form of myocardial ‘pretumoural tissue foci’.

The location of myxomas in the general population is as follows: 75% in the LA, 23% in the right atrium and only 2% in the ventricle. In 50% of the familial forms multiple locations are present and around 10% has a ventricular myxoma.[10] Careful inspection of all heart chambers is necessary since multiple tumour locations can be present, even if one is thrilled and satisfied by a single spectacular finding.

Recidives are often asymptomatic and are mostly detected by echocardiographic follow up studies.
Semiannual echocardiographic follow-up studies are therefore indicated for all cases. The symptomatology in recurrent myxomas is the same as in general non-recurrent myxomas. Location, size and mobility are determinants of the clinical features. One or more symptoms of the typical triad (embolism, obstructed ventricular filling, general symptoms) can be present. Small tumour particles or thrombus formation on the tumour surface can explain the embolic events that occur in 30–40% of myxoma patients. No tumour fragments were found in the histological examinations following acute lower limb ischaemia in our patient. Furthermore, no more embolic events occurred after the start of anticoagulation suggesting the tumour surface to be thrombogenic. It is our opinion that long-term anticoagulation is warranted in inoperable patients to prevent thrombo-embolic events. Myxomas often give signs of obstructed ventricular filling, mimicking thereby a mitral or tricuspid valve stenosis. Production and release of interleukin 6 by the tumour cells give rise to aspecific symptoms such as fatigue, weight loss, fever and arthralgia.

The speed of tumour growth can be calculated by echocardiographic studies or by the interval between the first and the second operations. It is estimated that myxomas grow 0.15 mm in a month or 18 mm in a year. The same calculations made for tumour mass give a growth of 1.2 g in a month or 14 g in a year. The same calculations made for tumour mass growth can be estimated for the first recurrence.

The first recurrence in this case (30 mm), not visualized earlier, occurred after 5 months. Thus, in this particular patient a growth of 6 mm in a month occurred after the start of anticoagulation suggesting the tumour to be thrombogenic. It is our opinion that long-term anticoagulation is warranted in inoperable patients to prevent thrombo-embolic events. Myxomas often give signs of obstructed ventricular filling, mimicking thereby a mitral or tricuspid valve stenosis. Production and release of interleukin 6 by the tumour cells give rise to aspecific symptoms such as fatigue, weight loss, fever and arthralgia.

The standard therapy for cardiac myxomas and recurrent myxomas is surgical resection. Chemotherapy and radiation therapy are disappointing. Radical excisional therapy and total cardiac transplantation have been reported. The role of orthotopic heart transplantation remains unclear, even in malignant heart tumours. In our opinion, anticoagulation therapy is warranted to prevent thrombo-embolic events in inoperable patients.

References


