Atrial myxomas: diagnosis and management

The first published report of left atrial myxoma appeared in 1845, and in 1955 the first successful excision was performed (Straus and Merliss, 1945).

Atrial myxoma can be a differential diagnosis for almost every cardiac condition. An atrial myxoma is a neoplasm of the endocardium. It is a fast-growing but benign tumour. Over 75% of primary cardiac tumours are benign and 50% of these are atrial myxomas. The remainder comprise lipomas, fibroelastomas and rhabdomyomas (Reynen, 1995).

Atrial myxomas can occur in any age group, but predominate between 30 and 60 years of age. They are more common in women. Cases of atrial myxomas are usually sporadic, but syndromic or familial cases do occur. In cases where multiple myxomas are found (multilocular tumours), other syndromic causes should be considered (McCarthy et al, 1986). The case report in this issue (p. 56) documents the case of a myxoma as a part of a cutaneous syndrome known as Carney complex (Carney et al, 1986). The authors discuss the diagnostic criteria, the chromosome defect and best management strategy.

Most patients present with one or more of the triad of embolism, intracardiac obstruction or constitutional symptoms. Most embolism is systemic as most myxomas occur in the left atrium. Cerebral and especially retinal arteries are usually affected, sometimes leading to permanent visual loss. Peripheral arterial, visceral and coronary arterial embolism have also been reported (Percell et al, 2003).

EMBOLISM

Right-sided myxomas present with less evident embolic events. Cases have been reported involving chronic tumour or thrombus embolization resulting in pulmonary hypertension and massive fatal pulmonary embolism (Peters et al, 1974; González et al, 1980).

INTRACARDIAC OBSTRUCTION

Atrial myxomas may partially obstruct filling of the left or right ventricle, mimicking the effects of mitral or tricuspid stenoses. Hence patients often complain of dyspnoea, recurrent pulmonary oedema and right heart failure. Valvular obstruction may vary with body position as demonstrated in another case report in this issue (p. 53), where the patient's dyspnoea worsened on lying flat, presumably as a result of worsening obstruction to left ventricular filling.

The opposite effect, namely valvular regurgitation, may result from excessive tumour movement back and forth across the valve.

Right atrial myxomas may mimic constrictive pericarditis with functional tricuspid stenosis and elevated right atrial pressures (Emanuel and Lloyd, 1962).

CONSTITUTIONAL SYMPTOMS

These may include fatigue, fever, an erythematous rash, arthralgia, myalgia and weight loss.

ABNORMAL RESULTS

Patients are often anaemic, which is often normochromic or hypochromic, or even haemolytic. Inflammatory markers are elevated as are globulins. Less commonly, polycythaemia, leucocytosis and thrombocytopenia may occur.

DIAGNOSTIC TESTS

The most accessible and diagnostic test is echocardiography. An electrocardiogram is usually non-specific and most commonly shows sinus rhythm. Chest X-ray may reveal an enlarged left atrium and signs of pulmonary congestion, but again is non-diagnostic.

Transthoracic echocardiography provides information on the location, size, shape, attachment and mobility of the myxoma, with further information on the insertion site, and morphological features including vegetations available from trans-oesophageal echo.

The advantage of computed tomography and magnetic resonance imaging is the availability of non-overlapping views with differentiation of tissue types.

Angiography should be performed on all patients over 40 years of age, before surgical treatment. Coronary angiography is usually adequate without the need for ventriculography. It may also provide information on tumour blood supply (see the case report on p. 53), and also emboli resulting in aneurysms, dilatation and coronary stenosis (Reynen, 1995).

TREATMENT AND RECURRENCE

The surgical removal of the tumour, its pedicle and the attached area of atrial wall is usually curative and should be performed promptly to prevent further complications. All chambers of the heart should be carefully inspected to ensure that the tumour is not multi-

KEY POINTS

- Atrial myxomas may be a differential diagnosis for almost any cardiac condition.
- They may be isolated or sporadic, or instead familial or part of a syndrome, such as the Carney complex.
- Recurrence is more common in the familial or complex-related myxomas, so screening post-removal should be regular and thorough.

focal. Operative mortality in previous series ranged between 0 and 3%. Supraventricular tachycardia and bradycardia requiring permanent pacemaker insertion are some of the surgical sequelae.

The rate of recurrence is dependent on whether the atrial myxoma is sporadic (1–3%) or familial or part of a complex (2–12%). Most recurrences are diagnosed within 4 years of surgery. Follow-up echocardiography should be performed twice yearly.

These case reports demonstrate the cardiocutaneous manifestations of atrial myxoma and also the variety of presentations associated with atrial myxoma – the well-recognized differential diagnosis. HM

Shrilla Banerjee

Specialist Registrar in Cardiology

The Heart Hospital

UCLH NHS Trust

London W1G 8PH

Carney JA, Hruska LS, Beauchamp GD, Gordon H (1986) Dominant inheritance of the complex of myxomas, spotty pigmentation, and endocrine overactivity. *Mayo Clin Proc* **61:** 165–72

Emanuel RW, Lloyd WE (1962) Right atrial myxoma mistaken for constrictive pericarditis. *Br Heart J* **24:** 796–800

González A, Altieri PI, Màrquez E, Cox RA, Castillo M (1980) Massive pulmonary embolism associated with a right ventricular myxoma. *Am J Med* **69**: 795–8

McCarthy PM, Piehler JM, Schaff HV et al (1986) The significance of multiple, recurrent, and 'complex' cardiac myxomas. *J Thorac Cardiovasc Surg* **91**: 389–96

Percell RL, Henning RJ, Siddique Patel M (2003) Atrial myxoma: case report and a review of the literature. *Heart Dis* **5**(3): 224–30

Peters MN, Hall RJ, Cooley DA, Leachman RD, Garcia E (1974) The clinical syndrome of atrial myxoma. *JAMA* **230:** 695–701

Reynen K (1995) Cardiac myxomas. *N Engl J Med* **333:** 1610–17

Straus R, Merliss R (1945) Primary tumor of the heart. *Arch Pathol* **39:** 74–8