

## Myxoma of the mitral valve

F. A. SANDRASAGRA<sup>1</sup>, W. A. OLIVER, AND T. A. H. ENGLISH

From the Norfolk and Norwich Hospital, Norwich NR1 3SR and the Regional Cardiothoracic Unit, Papworth Hospital, Cambridge

**SUMMARY** A case of myxoma of the mitral valve is reported. The clinical features were indistinguishable from left atrial myxoma with prolapse through the mitral valve. Because of the known tendency for these tumours to recur it was treated by wide excision which necessitated replacement of the mitral valve.

Myxomas, though rare, are the most common intracavitary cardiac tumours and account for over half of all primary cardiac neoplasms (Heath, 1968). The great majority occur in the left and right atria, usually arising from the vicinity of the fossa ovalis. Myxomas may rarely be found in other regions of the heart such as the right (Gottsegen *et al.*, 1963) and left (Kay *et al.*, 1959) ventricles and Prichard's extensive review of the subject in 1951 included reference to a primary myxoma of the mitral valve.

The purpose of this report is to document an unusual case of myxoma of the mitral valve and describe how surgical management was influenced

by consideration of the malignant potential of these tumours.

### Case report

A 33-year-old housewife presented in 1975 with an 11-year history of intermittent attacks of shortness of breath. Initially these were of abrupt onset and often unrelated to effort. In 1968 a clinical diagnosis of mixed mitral valve disease had been made and this was judged to be of mild degree on the basis of a good performance on the bicycle ergometer and low pressures at right heart catheterisation. The episodic nature of her symptoms was ascribed to paroxysmal tachycardia. In 1973 she developed atrial fibrillation and there was pronounced deterioration in her exercise tolerance.

<sup>1</sup>Present address: Royal Liverpool Children's Hospital, Myrtle Street, Liverpool L7 7DG.

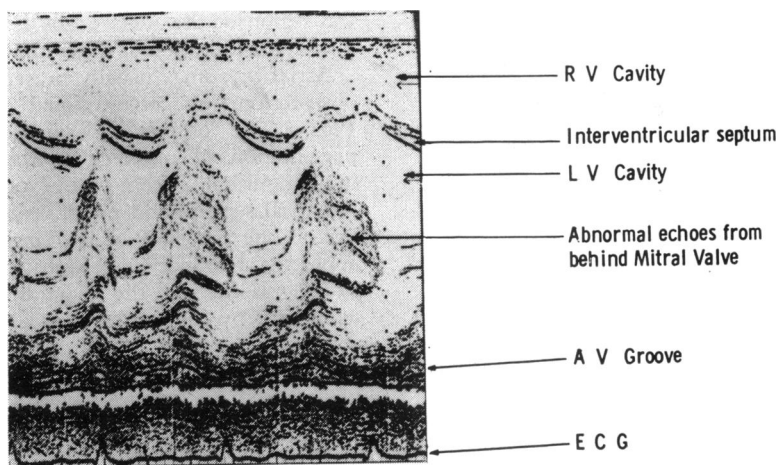


Fig. 1 Preoperative echocardiogram illustrating abnormal dense echoes behind anterior cusp of mitral valve.

On examination in July 1975 she was comfortable at rest but breathless on mild exertion. The pulse was 96 per minute and irregular and the blood pressure 120/70 mmHg. The jugular venous pressure was not raised. The apex beat was palpable in the sixth left intercostal space in the mid-clavicular line. There was a left parasternal heave and a diastolic thrill over the apex. On auscultation there was an extremely loud first sound with a late systolic murmur and a rumbling mid-diastolic murmur. The electrocardiogram confirmed the presence of atrial fibrillation and the chest radiograph showed cardiomegaly, with a cardiothoracic ratio of 14.5:27. At cardiac catheterisation the mean capillary wedge pressure was 30 mmHg. The pulmonary arterial pressure was 60/30 mmHg at rest, rising to 100/50 mmHg on exercise. The left ventriculogram showed moderate mitral reflux into a large left atrium. No filling defect was noticed in the left ventricle or left atrium and there was no calcification in the region of the mitral valve. An echocardiogram of the mitral valve (Fig. 1) showed a normal, mobile anterior cusp with multiple dense echoes posterior to it suggestive of a tumour mass in this region.

At operation on 6 October 1975 the external appearance of the heart was consistent with chronic mitral valve disease with moderate enlargement of the cardiac chambers, a dilated pulmonary artery, and a small aorta. On cardiopulmonary bypass the

left atrial cavity was explored and a large myxoma 3.5 × 4.5 × 1.5 cm (Fig. 2) was found arising from a pedicle attached to the anterior cusp of the mitral valve close to the lateral commissure. The tumour was soft and gelatinous with a frond-like structure and was pinkish-yellow in colour. The mitral annulus was dilated but the remainder of the mitral valve and the subvalvar mechanism appeared normal. The right atrium was opened and both surfaces of the atrial septum were inspected. No other myxomas were seen. The tumour was excised with a surrounding cuff of the anterior leaflet of the mitral valve. The resulting defect was too large for a direct repair, so the valve was excised and replaced with a Björk-Shiley prosthesis. Before insertion of the prosthesis, the left atrial and ventricular cavities were carefully flushed out with cold Hartman's solution to remove any fragments of tumour that may have been dislodged during excision. The patient's recovery was satisfactory and she was discharged from hospital 11 days later. Histological examination of the tumour confirmed it to be a myxoma. There was some increase in the normal non-compact myxomatous connective tissue lamina of the mitral valve cusps and a small group of myxoma cells was found in the endocardium of the posterior papillary muscle. Two years later the patient was asymptomatic. She was in sinus rhythm and her heart size was within normal limits, with a cardiothoracic ratio of 12.5:27.

### Discussion

Intracardiac myxomas are believed to arise from remnants of myxoid tissue of embryonic myocardium. The frequent persistence of such tissue in the region of the fossa ovalis helps to explain the peculiar localisation of these tumours (Yater, 1931).

Atrial myxomas usually present with embolism, obstruction, or constitutional illness (Goodwin, 1963). The obstructive symptoms and clinical presentation of myxoma of the mitral valve are indistinguishable from that of one which arises on the atrial septum and prolapses through the mitral valve. The presence of a filling defect is usually apparent at angiocardiography, but this is sometimes missed and the diagnosis can usually be confirmed by echocardiography (Popp and Harrison, 1969).

Until relatively recently the accepted surgical management for intracardiac myxomas was simple excision of the tumour (Firor *et al.*, 1966). However, since Gerbode and colleagues (1967) reported the first case of recurrent left atrial myxoma, there have been several reports of tumour recurrence, both at and remote from the site of excision (Bahl *et al.*,

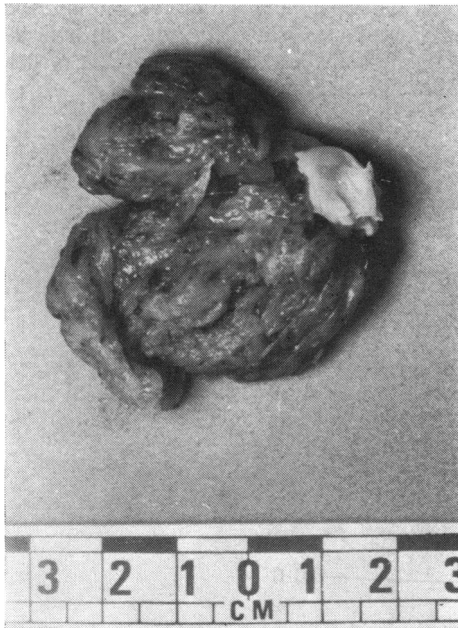


Fig. 1 Specimen of excised myxoma of the mitral valve.

1969; Hattler *et al.*, 1970; Walton *et al.*, 1972; Jugdutt *et al.*, 1975). The cause of recurrence is not clear and has been attributed to local implantation of cells by the primary tumour (Kabbani and Cooley, 1973) and the systemic embolisation of myxoma cells (Read *et al.*, 1974). Several reports have drawn attention to an unusually rapid growth rate of the recurrent tumour, and Walton and colleagues (1972) described an interesting case in which recurrence occurred within 22 months at the site of removal of the previous tumour, despite excision of a surrounding cuff of atrial septum. Careful histological examination of the piece of septum removed at the initial operation failed to show any tumour cells and they suggested that recurrence might have been from growth from 'pre-tumour' cells in this area. The malignant potential of atrial myxoma was stressed in a review of recurrent tumours by Read and colleagues (1974), and they described one patient who developed metastatic myxomas in the sternum and pelvis 6 years after excision of an atrial myxoma. The metastatic tumours appeared histologically more cellular, pleomorphic, and malignant, but were otherwise similar to the original atrial tumour. These authors also reported the first patient on record to develop a recurrent myxoma on his mitral valve, for which valve replacement was required.

Because of the prevailing uncertainty with regard to the prognosis of these tumours, we agree with Kabbani and Cooley (1973) that a radical approach is indicated at the primary operation. In addition, precautions should be taken to prevent tumour embolism during operation and both atria should always be explored to exclude the presence of multicentric tumours. Application of these principles in the case presented led to excision of the mitral valve, whereas a conservative operation might otherwise have been performed. The subsequent histological demonstration of a small group of myxoma cells in the endocardium of the posterior papillary muscle was an unexpected finding and could represent either a multicentric focus or implantation of cells by the primary tumour.

We are grateful to Dr P. G. I. Stovin for interpretation of the histological findings.

#### References

- Bahl, O. P., Oliver, G. C., Ferguson, T. B., Schad, N., and Parker, B. M. (1969). Recurrent left atrial myxoma. *Circulation*, **40**, 673-676.
- Firor, W. B., Aldridge, H. E., and Bigelow, W. G. (1966). A follow-up study of three patients after removal of left atrial myxoma five to ten years previously. *Journal of Thoracic and Cardiovascular Surgery*, **51**, 515-520.
- Gerbode, F., Kerth, J. W., and Hill, D. J. (1967). Surgical management of tumours of the heart. *Surgery*, **61**, 94-101.
- Goodwin, J. F. (1963). Diagnosis of left atrial myxoma. *Lancet*, **1**, 464-468.
- Gottsegen, G., Wessely, J., Arvay, A., and Temesvari, A. (1963). Right ventricular myxoma simulating pulmonic stenosis. *Circulation*, **27**, 95-97.
- Hattler, B. G., Fuchs, J. C., Cosson, R., and Sabiston, D. C. (1970). Atrial myxoma. *Annals of Thoracic Surgery*, **10**, 65-74.
- Heath, D. (1968). Pathology of cardiac tumours. *American Journal of Cardiology*, **21**, 315-327.
- Jugdutt, B. I., Rossall, R. E., and Sterns, L. P. (1975). An unusual case of recurrent left atrial myxoma. *Canadian Medical Association Journal*, **112**, 1099-1100.
- Kabbani, S. S., and Cooley, D. A. (1973). Atrial myxoma. Surgical considerations. *Journal of Thoracic and Cardiovascular Surgery*, **65**, 731-737.
- Kay, J. H., Anderson, R. M., Meihaus, J., Lewis, R., Magidson, O., Bernstein, S., and Griffith, G. C. (1959). Surgical removal of an intracavitary left ventricular myxoma. *Circulation*, **20**, 881-886.
- Popp, R. L., and Harrison, D. C. (1969). Ultrasound for the diagnosis of atrial tumour. *Annals of Internal Medicine*, **71**, 785-787.
- Prichard, R. W. (1951). Tumours of the heart. *Archives of Pathology*, **51**, 98-128.
- Read, R. C., White, H. J., Murphy, M. L., Williams, D., Sun, C. N., and Flanagan, W. H. (1974). The malignant potentiality of left atrial myxoma. *Journal of Thoracic and Cardiovascular Surgery*, **68**, 857-868.
- Walton, J. A., jun., Kahn, D. R., and Willis, P. W. (1972). Recurrence of a left atrial myxoma. *American Journal of Cardiology*, **29**, 872-876.
- Yater, W. M. (1931). Tumours of the heart and pericardium. *Archives of Internal Medicine*, **48**, 627-666.

Requests for reprints to T. A. H. English, Esq., F.R.C.S., Papworth Hospital, Cambridge CB3 8RE.