Case reports

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An unusual case of large right atrial myxoma

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SUMMARY A case of right atrial myxoma is described. The patient had vague and variable symptoms and it was not until five years after first presentation that she had developed clinical and electrocardiographic evidence of right atrial hypertrophy, and angiocardigraphic evidence of a fist-sized myxoma which was subsequently removed surgically.

Although 75 per cent of cardiac myxomata occur in the left atrium, there are many reports of right atrial myxoma.^{1 2} Symptoms of exertional dyspnoea, weakness, and fatigue are frequent and have been related to a reduction in cardiac output² which has been confirmed at cardiac catheterisation in some instances.^{3 4} These symptoms are, however, usually accompanied by signs of obstruction to right atrial outflow.^{2 5} We describe a patient who first presented with variable symptoms of weakness, malaise, and dizziness in the absence of physical signs or electrocardiographic abnormalities. Though several medical practitioners referred to her bizarre symptoms, the diagnosis of right atrial myxoma was not made until the development of abnormal physical signs and an abnormal electrocardiogram.

Case report

A 28-year-old woman, a typist, complained of several episodes of light-headedness, sweating, breathlessness, and weakness which lasted up to 30 minutes and were usually unrelated to exercise. She was referred to a consultant cardiologist who found no clinical, radiological, or electrocardiographic abnormalities.

The most impressive feature of the interview was her pronounced agitation and distress upon seeing a dead cockroach under the consultation couch which led to an attack similar to those she had experienced previously. A diagnosis of hysterical hyperventilation resulting from a personality disorder was made at that time, but no further investigations were performed to substantiate this.

In the following year she was referred to a gynaecology department for investigation of amenorrhoea, which was attributed to discontinuing an

oral contraceptive. The amenorrhoea persisted, but when she and her husband eventually decided after another four years that they did not want any children, she was admitted for bilateral tubal ligation. Several doctors again commented on her bizarre personality, but it was also noted that she had a systolic murmur and she was referred to the cardiology clinic, by then five years after her original appointment.

She complained of further attacks similar to those described earlier, but they were more frequent and more severe. During such an episode she would "feel like a jelly", with extreme tiredness and light-headedness, was sometimes breathless, and usually felt as though she wanted to close her eyes and sleep. Attacks occurred both on exercise and at rest and she frequently had to sit or even lie down in a shop on the way to work if she felt unwell. If they occurred at home, she would lie face down on a bed for 30 minutes after which time she improved, but sometimes she felt unwell for as long as 12 hours. Between attacks she felt well for periods of up to several weeks and she regularly took part in vigorous exercise.

By then, however, there were several abnormal features. There was a very prominent "a" wave in the jugular venous pulse and also a scratchy intermittent systolic and diastolic murmur, maximal in the fourth intercostal space at the left sternal edge and sounding like a pericardial friction rub or an exocardiac murmur. The electrocardiogram showed very tall P waves in leads II, III, and V1 to V4, indicating right atrial hypertrophy. There was also T wave inversion in V1 to V4 (Fig. 1). The chest x-ray film showed a straight heart border, but no increase in heart size.

The erythrocyte sedimentation rate was 60 mm

in the first hour and, though the total protein was normal, the albumin fraction was low at 49 per cent, with slightly raised alpha-2 and gamma globulins. The liver was not enlarged but the alkaline phosphatase was raised at 222 units/l (normal 20 to 85 units/l) with an increased liver band on electrophoresis.

At cardiac catheterisation the right atrial "a" wave was 19 mmHg, the right ventricular pressure was 27/10 mmHg, and the pulmonary artery pressure 16/7 mmHg. Cineangiography showed a large filling defect in the right atrial cavity.

Because of the auscultatory evidence which suggested pericarditis, the T wave changes in the right ventricular leads, the raised alkaline phosphatase, and a liver scan reported as showing small metastatic deposits, it was feared that there was a malignant tumour, with right ventricular and pericardial infiltration and hepatic metastases. A liver biopsy, however, showed no evidence of tumour.

At thoracotomy the right atrium was found to be almost completely filled by a large, hard, bluish-purple mass which was slightly lobulated and was attached over a distance of 2 cm to the lateral wall of the atrium. The tumour (Fig. 2) measured

9 × 8 × 6 cm and histological examination confirmed that it was a myxoma. Inspection of the liver showed that it was enlarged and congested, but there was no sign of tumour.

Postoperatively she recovered satisfactorily; the alkaline phosphatase and liver scan rapidly returned to normal. One year after operation, she was entirely free from symptoms and had a normal venous pressure, no friction rub or murmur, and a normal electrocardiogram and chest x-ray. Erythrocyte sedimentation rate and plasma proteins were also normal. She had returned to an active life, her only complaint being that her menstrual periods had restarted.

Discussion

A combination of features (namely what appeared to be a pericardial friction rub, T wave changes in the electrocardiogram, elevation of the alkaline phosphatase, and an abnormal liver scan) suggested the possibility that the filling defect noted on cineangiography was caused by a malignant tumour. Previous case reports of a right atrial myxoma have, however, recorded the presence of what seemed to be a pericardial friction rub⁶ though the cause is uncertain. The T wave abnormalities in leads V1 to V4 are unexplained, but similar changes have been observed.⁷ It is possible that the raised alkaline phosphatase and perhaps also the liver scan abnormalities were a result of hepatic congestion secondary to obstruction in the right atrium.

The tumour originated from the lateral wall of the atrium, whereas in most reported cases the origin was from the atrial septum in the vicinity of the fossa ovalis.⁸ Origin from the free inferolateral wall of the right atrium has, however, been described.^{9 10}

Fig. 1 Comparison of electrocardiograms from first attendance and five years later, showing the development of large P waves and anterior T wave inversion.

Fig. 2 The appearance of the right atrial myxoma after surgical removal.

Symptoms are said to be more variable when a tumour occurs in the right rather than the left atrium.¹¹ It was, however, a striking feature of this patient's history, that the symptoms were not only vague but also intermittent. Despite the presence of such a large tumour—one of the largest recorded^{10 12}—she could be free from symptoms for several weeks. As symptoms were variable and often relieved by lying face down on a bed, it is possible that they were related to reductions in cardiac output produced by variable right atrial outflow obstruction.

Constitutional effects of atrial myxomata are common and include fever, anaemia, weight loss, increased sedimentation rate, and raised serum globulins.^{12–14} Our patient had increased erythrocyte sedimentation rate and also increased alpha-2 and gamma globulins. It may be that her amenorrhoea, which lasted for five years and recovered after operation, was a result of a similar constitutional effect of the tumour. Though weakness and malaise have been described in other cases of right atrial myxoma, this patient's symptoms were present several years before when a consultant cardiologist had found no clinical, x-ray, or electrocardiographic abnormalitie.

To what extent her symptoms were initially caused by the tumour on the one hand or by her personality on the other, and whether her behaviour was entirely a result of a systemic effect of the tumour remain matters for speculation. Nevertheless, it seems that she now leads a normal life and her behaviour has been normal apart from initial anxiety about the possibility of recurrence of the tumour.

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