Prolapsing right atrial myxoma

Clinical and haemodynamic considerations

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A 22-year-old woman with a right atrial myxoma prolapsing to the right ventricle is described. The haemodynamic findings were similar to those of cases of prolapsing myxoma of the left atrium; a notching on the ascending limb of the right ventricular pressure curve, and an initial negative, irregular deflection on the pulmonary artery pressure curve with a pronounced rise in the mean right atrial pressure (18 mmHg) were found. On deep inspiration there was a significant deepening of the y descent from 12 mmHg to 2 mmHg, indicating a changing, dynamic obstruction of the right ventricle inflow tract. These haemodynamic features can be helpful in the diagnosis of prolapsing right atrial myxoma.

Myxomata of the atria are uncommon, especially in the right atrium (Sannerstedt *et al.*, 1962; Morrissey *et al.*, 1963). Recently, Sung *et al.* (1975) differentiated between two types of left atrial myxoma: type I, in which the tumour prolapses during diastole into the left ventricle, and type II, which is non-prolapsing. Sung described a notch in the upstroke of the left ventricular pressure curve and a rapid y descent in the pulmonary arterial wedge pressure curve as characteristic of the prolapsing type.

The purpose of this report is to present a case of a right atrial myxoma, prolapsing during diastole into the right ventricle, producing haemodynamic features similar to those described by Sung for the prolapsing left atrial myxoma.

Case report

A 22-year-old single woman of Arabian origin had been completely well until 6 months before her admission, when she began complaining of palpitation, frequent cough, and haemoptysis; one month before admission her complaints became worse with pain in the right posterior thorax, frequent nausea and vomiting, and a loss of 7 kg in weight. On admission she had a temperature of 37.9°C; there was no cyanosis, clubbing, or dyspnoea; her blood pressure was 120/80 mmHg, pulse rate 110/min, with regular rhythm. There was pronounced jugular venous engorgement with prominent a and v waves and a negative Kussmaul sign. Auscultation

of the lungs revealed poor air entry into the right lung base and a pleural friction rub at the left base. The apex beat was palpated in the left fifth inter-space in the midclavicular line; no right ventricular heave was noted. A diastolic thrill at the lower left sternal border was palpable; the first heart sound was prolonged and accentuated. The second sound was normal; a grade 4/6 mid-diastolic presystolic murmur extending into the first sound and early systole was heard at the lower left sternal border. The character of the murmur was similar to a pericardial friction rub; the intensity of the murmur was unaffected by the position of the patient but increased slightly on deep inspiration. The liver was palpable 5 cm below the costal margin and tender; the spleen was not palpable. There was mild pitting oedema in both legs.

The electrocardiogram showed sinus rhythm with a rate of 110/min, tall P waves in leads II, III, and aVF. The QRS axis was +120°, and there were no signs of right or left ventricular hypertrophy (Fig. 1a).

The sedimentation rate was 55 mm in the first hour and 85 mm in the second hour; total serum protein 84 g/l, albumin 30 g/l and globulin 54 g/l, A/G ratio 0.55 with a slight increase in β globulin and a distinct increase in γ globulin. Total bilirubin was 1.5 mg/100 ml (26.7 μ mol/l), mostly indirect. Other blood chemistry was normal and haemoglobin, white cell count, platelets, and serology were within normal limits.

The chest x-ray film showed slight enlargement

Fig. 1 (a) An electrocardiogram showing a normal sinus rhythm, peaked P waves in leads II, III, and aVF; the QRS axis is $+120^\circ$. (b) Right atrial pressure curve: the most outstanding finding is a rapid and deep y descent on deep inspiration.

of the right atrium, bilateral basal pleural effusions, and the lung scan with macroaggregated albumin labelled with ^{99}Tc was suggestive of multiple pulmonary emboli.

The phonocardiogram at the lower left sternal border showed an accentuated and prolonged first heart sound and a mid to late diastolic murmur extending into the first heart sound.

HAEMODYNAMIC STUDIES

Right heart catheterisation was performed. Cardiac output was 4.0 l/min; cardiac index 2.5 l/min per m^2 , stroke volume 36 ml. The various pressures recorded are listed in the Table. There was a pronounced rise in the mean pressures in the right atrium, inferior and superior vena cava (18 mmHg). There was a slight rise in the right ventricular

Table Haemodynamic data

Site	Pressures (mmHg) Mean	Phasic			
		a	x	v	y
Superior vena cava	18				
Inferior vena cava	18				
Right atrium	18	24	15	20	14
Right atrium in inspiration	18	28	17	22	2
Right ventricle		34/8			
Pulmonary artery	24	38/20			
Pulmonary wedge	8				

pressure (34/8 mmHg) and a notching in the ascending limb of the right ventricular pressure. Furthermore, an early deep and irregular negative deflection of the pulmonary arterial pressure curve was noted. The most significant finding was a rapid and deep y descent on the right atrial pressure tracing on deep inspiration (Fig. 1b).

Angiocardiography was performed by injecting contrast medium into the superior vena cava close to its junction with the right atrium. 70-mm films obtained in the left anterior oblique and antero-posterior position showed a large filling defect in the right atrium (Fig. 2); cineangiocardiology showed that the filling defect protruded into the right ventricle during diastole.

At operation under cardiopulmonary bypass a large tumour ($8.5 \times 5 \times 5$ cm), covered in blood

Fig. 2 70-mm film, injection of contrast medium into the superior vena cava close to its junction with the right atrium (left anterior oblique position). A large filling defect is seen in the right atrium.

clot, was found attached to the inferolateral wall of the right atrium, which protruded into the right ventricle: it was removed together with the adjacent area of the right atrial wall (Fig. 3). The histological examination confirmed the diagnosis of atrial myxoma. The postoperative course was uneventful, the patient recovering fully; there are now no audible murmurs and the previous jugular venous engorgement has disappeared. The tall peaked P waves in the electrocardiogram disappeared immediately after the removal of the tumour while the patient was still on the operating table. The patient gained 8 kg in true body weight within 3 months of the operation.

Discussion

The physical findings as described in our patient could erroneously lead to the diagnosis of organic tricuspid valve disease. Nevertheless, the short history, the absence of mitral valve disease, the absence of a right ventricular heave, the presence of normal sinus rhythm (Morrissey *et al.*, 1963), the high blood sedimentation rate, and hypergammaglobulinaemia, all supported the clinical diagnosis of right atrial myxoma. It is interesting that the high peaked P wave seen in the electro-

cardiogram in our patient and described by others in right atrial myxoma (Sannerstedt *et al.*, 1962), disappeared immediately after the removal of the tumour while the patient was still on the operating table.

Whereas in almost all reported cases of right atrial myxoma, the tumour originated from the atrial septum in the vicinity of the fossa ovalis (Nasser *et al.*, 1972 a, b), in our case it was attached to the free inferolateral wall of the right atrium. A similar location for the tumour growth has been described by Currey *et al.* (1967).

On cardiac catheterisation a notch in the upstroke of the right ventricular pressure curve was seen in our case (Fig. 2). Such a notch in the right ventricular pressure curve in right atrial myxoma has been described previously (Ramsey *et al.*, 1969) and thought to be caused by the expulsion of the protruding tumour mass during early ventricular systole across the tricuspid orifice, and back into the right atrium interfering with the early phase of ventricular contraction. A similar notch in the upstroke of the left ventricular pressure curve was also described in relation to left atrial myxoma (Pitt *et al.*, 1967). Furthermore, it was shown (Sung *et al.*, 1975) that such a notch was to be found only in those cases of left atrial myxoma

Fig. 3 The large myxomatous tumour (8.5×5×5 cm) removed from the right atrium.

which prolapsed during diastole into the left ventricle, and were identified as the prolapsing type. This notch, whether in the right or left ventricular pressure curves, is especially indicative of an atrial myxoma if the notch appears to vary in magnitude, degree, and position on the ascending limb, probably reflecting the changing location of the pedunculated prolapsing atrial myxoma, with each cardiac cycle. Such a notch has been considered not to be an artefact, because it was recorded only preoperatively and was absent after removal of a left atrial tumour (Nasser *et al.*, 1972 a, b).

In an experimental study in dogs (Pantazopoulos and Moscovitz, 1972), using a distensible balloon to simulate a pedunculated left atrial tumour, it was shown that moderate balloon distension caused a prominent notch on the ascending limb of the left ventricular pressure pulse, caused by displacement of the balloon from the left ventricle into the left atrium. Pronounced balloon distension caused even greater left ventricular notching, interrupted aortic blood flow during ejection, and in addition caused a rapid y descent in the left atrium pressure curve. In our case an early deep and irregular negative deflection of the pulmonary artery pressure curve was observed and could have been caused by a temporary interruption of the pulmonary arterial blood flow; though artefacts, recorded frequently in pulmonary arterial pressure curves, cannot definitely be excluded.

The finding of a slow y descent in the right atrial pressure curve suggests the presence of a static unchanging obstruction to right atrial emptying, i.e. tricuspid stenosis. A very rapid y descent on the other hand is considered strong evidence against the presence of an organic tricuspid stenosis. The right atrial pressure curve in our case showed a predominant a wave of 24 mmHg, v wave of 20 mmHg, and a y descent to 14 mmHg. On deep inspiration a distinct deepening and steepening of the y descent down to 2 mmHg occurred (Fig. 1b). A steep y descent is seen in severe tricuspid regurgitation but the absence in our case of a tall dominant v wave rules out this possibility.

Sannerstedt *et al.* (1962) pointed out that the most significant haemodynamic findings in right atrial myxoma are to be sought during the phase of rapid filling, consisting of irregularities of the right atrial pressure curve during this phase. They suggested that these changes could be the result of intermittent occlusion of the tricuspid orifice by the tumour mass.

The steep y descent seen in our case during deep inspiration indicates a prolapsing type of right atrial myxoma. In this prolapsing type of myxoma part of the tumour moves during early diastole

through the tricuspid valve and away from the right atrium, causing thereby a diminution in the right atrial pressure curve. This change in the right atrial pressure curve became especially accentuated during deep inspiration, causing a very steep y descent. Changing character of the y descent indicates that obstruction to the right ventricular inflow is not the result of a static and unchanging stenosis but of a changing dynamic type of obstruction. This changing configuration of the y descent should arouse suspicion of a pedunculated type of right atrial myxoma prolapsing into the right ventricle.

The haemodynamic features described here might be helpful in differentiating right atrial myxoma from 'false positive' angiographic filling defects caused by streaming and also from other space-occupying lesions of the right atrium, such as large thrombi or filling defects caused by compression of the right atrial wall from without.

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References

- Currey, H. L. F., Mathews, J. A., and Robinson, J. (1967). Right atrial myxoma mimicking a rheumatic disorder. *British Medical Journal*, **1**, 547.
- Morrissey, J. F., Campeti, F. L., Mahoney, E. B., and Yu, P. N. (1963). Right atrial myxoma: report of two cases and review of the literature. *American Heart Journal*, **66**, 4.
- Nasser, W. K., Davis, R. H., Dillon, J. C., Tavel, M. E., Helmen, C. H., Feigenbaum, H., and Fisch, C. (1972a). Atrial myxoma. I. Clinical and pathologic features in nine cases. *American Heart Journal*, **83**, 694.
- Nasser, W. K., Davis, R. H., Dillon, J. C., Tavel, M. E., Helmen, C. H., Feigenbaum, H., and Fisch, C. (1972b). Atrial myxoma. II. Phonocardiographic, echocardiographic, hemodynamic, and angiographic features in nine cases. *American Heart Journal*, **83**, 810.
- Pantazopoulos, J., and Moscovitz, H. L. (1972). The hemodynamics of experimental left atrial tumor. *Circulation*, **45-46**, Suppl. II, 201.
- Pitt, A., Pitt, B., Schaefer, J., and Criley, J. M. (1967). Myxoma of the left atrium: hemodynamic and phonocardiographic consequence of sudden tumor movement. *Circulation*, **36**, 408.
- Ramsey, H. W., Elliott, L. P., Bartley, T. D., Mantini, E., and Eliot, R. S. (1969). Right atrial myxomata. *Archives of Internal Medicine*, **124**, 206.
- Sannerstedt, R., Varnauskas, E., Paulin, S., Linder, E., Ljunggren, H., and Werkö, L. (1962). Right atrial myxoma: report of a case and review of the literature. *American Heart Journal*, **64**, 243.
- Sung, R. J., Ghahramani, A. R., Mallon, S. M., Richter, S. E., Sommer, L. S., Gottlieb, S., and Myerburg, R. J. (1975). Hemodynamic features of prolapsing and non-prolapsing left atrial myxoma. *Circulation*, **51**, 342.

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