ATRIAL MYXOMA

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SYNOPSIS

Two cases of atrial myxoma, one in the right atrium and the other in the left atrium are described. The clinical, electrocardiographic, haemodynamic and angiographic features are presented and discussed. Both patients underwent surgical removal. Case 1 survived the operation while Case 2 succumbed from severe acute pulmonary oedema. The clinical and haemodynamic features of atrial myxoma are discussed.

Atrial myxomata originally described by King in 1845 are relatively rare. Strauss and Merlin (1945) could only find a rate of seventeen primary cardiac tumours per million necropsies from the years 1938 to 1942 in the United States, while Benjamin (1939) reported a prevalence rate of 0.03 per cent of primary cardiac tumours in 40,000 consecutive necropsies. Goldberg (1952) was the first to diagnose this condition during life. The diagnosis of atrial myxoma is important as this condition can be completely 'cured' by surgical removal of the tumour. Barhnson and Newman (1953) were the first to report successful surgical removal. However, there are reports of metastases and recurrence even after surgical removal (Walton, 1972). Its diagnosis, too, can sometimes be difficult as it can simulate almost any cardiovascular disorder, or produce symptoms and signs which suggest diseases of other systems, drawing attention away from a diagnosis of a cardiac lesion. Its occurrence in siblings have been described by Siltaren (1976). As far as we are aware, there have been no reported cases of atrial myxomata — either diagnosed in life or at post-mortem described in the local literature. We report two cases of atrial myxomata, one in the right atrium and in the other in the left atrium. Both were diagnosed pre-operatively and had surgical removal of their tumours. These two patients illustrate all the manifestations of atrial myxomata (Goodwin, 1968).

ILLUSTRATIVE CASE HISTORY NO. 1

Y.A.T., a 48 year old Chinese male, developed left hemiparesis with slurring of speech in January 1975. He recovered fully after three days, without any treatment. From March 1975, he experienced progressive dyspnoea, orthopnoea and episodic attacks of giddiness especially on standing up from a squatting position. There were no actual syncopal episodes. During this time, he noticed a gradual loss of twenty pounds in body weight from March 1975.

He was admitted for investigation from July to September 1975. On examination, his general condition was satisfactory but was pale and tachypnoeic at rest. Blood pressure was 95/60 mmHg and jugular venous pressure was raised to the ears. Apex beat was at the 6th left intercostal space, at the midclavicular line. The first heart sound was widely split and a 'tumour plop' was audible (Fig. 1). There were signs of pulmonary hypertension. Liver was enlarged 3 cm. While in hospital, he suddenly collapsed in the bathroom and was found to be hypotensive. Two weeks later, he complained of sudden onset of left-sided chest pain associated with haemoptypsis. The initial clinical diagnosis was pulmonary hypertension due to chronic thrombo-embolism. A lung scan showed no uptake in lower left lobe and was suggestive of pulmonary embolism, however pulmonary angiograms showed absence of emboli in the pulmonary vasculature. The pulmonary artery pressure, was however elevated. He was discharged in September with digoxin, frusemide and warfarin.

His symptoms remained unchanged and cardiac catheterisation was done on 13.1.76. Prior to cardiac catheterisation, a provisional diagnosis of left atrial myxoma was made.

INVESTIGATIONS

Hb 15.0 gm per cent, Tw 9,500, serum bilirubin 0.6 mg per cent, alkaline phosphatase 7 King's Units, total protein 8.3 gm per cent, albumin 3.7 gm per cent, SGPT 13 King's Units. Urine analysis was normal. Blood urea was 45 mg per cent. ESR 2 mm/1st hour. K^+ 4.3, Na^+ 135, Cl^- 103 meq/litre.

12-lead ECG showed normal sinus rhythm, severe right ventricular hypertrophy with strain pattern, and right axis deviation (Fig. 2).

In the chest X-ray there was cardiomegaly, (CTR 0.57), pulmonary arterial and venous hypertension and left atrial enlargement.

Echocardiogram showed square wave pattern and multiple echoes behind the anterior leaflet of the mitral valve and in the left atrium (Fig. 3).

CATHETERISATION FINDINGS

TABLE I — Haemodynamic Data of Case 1

Site	Pressure (mmHg)		Saturation
	Phasic	Mean	
Pulmonary wedge pressure	a= 25, v= 50 x= 24, y= 35	35	97%
Left pulmonary artery	74/32	48	57%
Main pulmonary artery	74/32	48	57%
Main pulmonary artery	74/32	48	57%
Right ventricle	74/12		54%
Right atrium	a= 13, x= 31 v= 6, y= 4	8	
Ascending aorta	137/100	97%	
Left ventricle	137/17		96%

Cardiac output = 3.0 L/min
Cardiac index = 4.2 L/min/m^2
Mitral mean diastolic gradient = 20 mmHg
Mitral end diastolic gradient = 15 mmHg

There was pulmonary arterial and venous hypertension with a prominent 'v' wave in the pulmonary artery wedge tracing (Table I). The left ventricular pressure curve showed a systolic notch on its upstroke — which is typical of left atrial myxoma (Fig. 4). There was a mean diastolic gradient of 20 mmHg across the mitral valve. The angiogram showed a large filling defect in the left atrium, bobbing in and out of the left ventricle (Fig. 5).

OPERATIVE FINDINGS

The patient underwent surgery on 10.3.76. A large myxoma, attached to the inter-atrial septum by a pedicle, completely filling the left atrium was found. It weighed 70 gm. Under cardiopulmonary bypass, the tumour with its pedicle was removed. Post-operatively, the intracardiac pressures were recorded as follows:- RA 5 mmHg, PA 50/25 mmHg, RV 50/15, LA 15 mmHg, aorta 100/55 mm Hg.

Microscopic examination of the tumour showed sheets of amorphous myxoid round substance in which there are interspersed, small cells, with stellate appearance as well as multinucleate cells with eosinophilic cytoplasm and small oval nuclei. There are large areas showing dilated capillaries and channels with areas of recent and old haemorrhage.

ILLUSTRATIVE CASE HISTORY NO. 2

T.L.S., a 19 year old Chinese female clerk complained of retrosternal chest pain in November

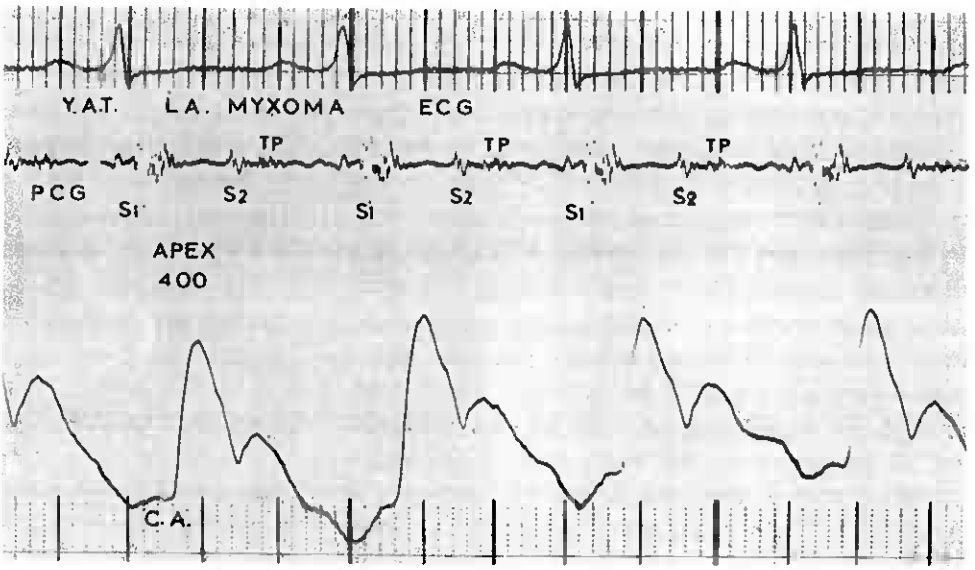

Fig. 1
Phonocardiogram of Case 1 with simultaneous carotid artery tracing. The first heart sound is widely split and a tumour plop (TP) is demonstrated.

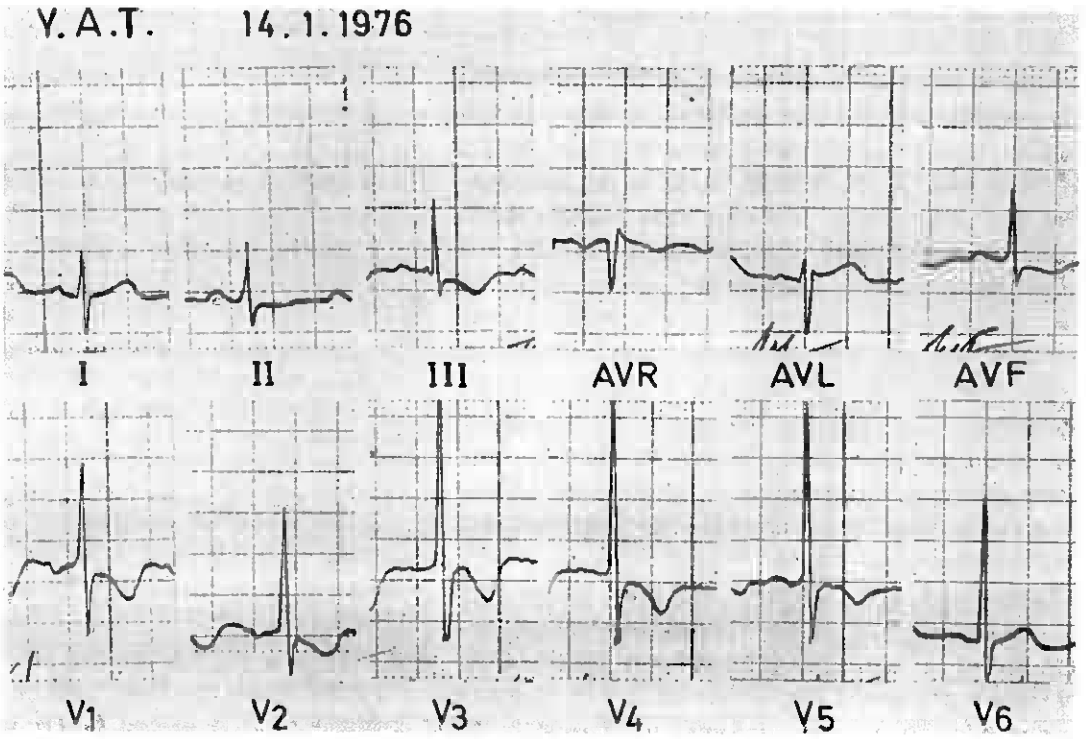

Fig 2
The 12-lead electrocardiogram of Case 1 which shows severe right ventricular hypertrophy.

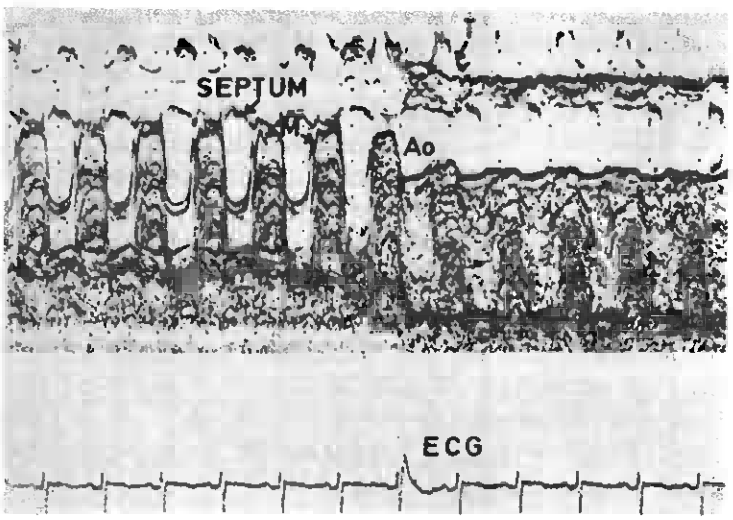

Fig. 3
Echocardiogram of Case 1 shows the square wave pattern of the mitral valve labelled M with multiple echoes behind the anterior mitral leaflet seen in diastole. The sweep towards the aorta (AO) shows multiple echoes in the left atrium (labelled LA) due to the tumour.

1975. She was free of symptoms until July 1974, when she was admitted for complaints of exertional dyspnoea, palpitation, chest pain and fever. These episodes of breathlessness were partially relieved by leaning forward and worsened by lying on her right side. Examination then, showed she was febrile, heart rate was 120 per minute. At the apex and left sternal border a fourth heart sound and an ejection systolic murmur were heard. The liver was enlarged 2 cm and the spleen was just palpable. Significant investigations were as follows:- Hb 10.2 gm per cent, Tw 14,900/cu.mm. ESR 66 mm/1st hour. Urea 22 mg per cent. CXR was normal. ECG showed low voltage, sinus tachycardia and prominent R waves in the right praecordial leads (Fig. 6). Her fever subsided spontaneously. She was thought to have a viral infection and was discharged on 6.8.74.

She was re-admitted 4 days later on 10.8.74 for the same complaints. However, at this admission she was found to be pale and toxic looking, in low cardiac output and with small volume pulses. Blood pressure was 90/60 mmHg. Jugular venous pressure was raised 4 cm. The apex beat was at the 5th LICS, just outside the mid-clavicular line. Third and fourth heart sounds were heard at the apex. An ejection click and a ejection systolic murmur were heard at the left sternal border, third intercostal space. The liver was enlarged to 5 cm. During this admission, she had one episode of hypotension, her blood pressure was 50 mmHg systolic. Her blood pressure improved with intravenous Effortil (R) and she improved gradually with treatment and was discharged on 7.9.74.

Two weeks later (22.9.74) she was re-admitted for headache, exertional dyspnoea, and cough, and was found to be in congestive heart failure. Over the next eight weeks she remained ill with frequent episodes of chest pain, abdominal pain and hypotension. She was jaundiced and the jugular venous pressure was raised to her ears. Multiple ejection systolic clicks and a short mid-diastolic murmur at the left sternal border, second intercostal space were heard (Fig. 7). Haematological investigations showed evidence of haemolysis and thrombocytopenia. Hb was 12 gm per cent, reticulocyte count 9 per cent. Platelet count fell to 10,000/cu.mm.

She was admitted for the fourth time on 30.12.74 for fever, cough and dyspnoea. Her fever subsided spontaneously and she was discharged on 6.1.75. The fifth admission (20.4.75 to 26.4.75) was for chest pain and ascites.

She was admitted on 9.7.75 for cardiac catheterisation. Examination revealed a pale young lady in low cardiac output state. Pulses were small volume and jugular venous pressure was raised to the ears. There were prominent 'a' waves with prominent 'x' descent (Fig. 7). Ejection systolic clicks and a long early diastolic murmur were heard over the left sternal border, third intercostal space. Third and fourth heart sounds were heard at the apex. The liver was enlarged 6 cm and was pulsatile.

Hb was 14.8 gm per cent, Tw 15,300/cu.mm., platelet count 90,000/cu.mm. Chest X-ray showed cardiomegaly with normal lung fields.

CARDIAC CATHETERISATION DATA

TABLE 2 — Haemodynamic Data of Case 2

Site	Pressure (mmHg)		Saturation
	Phasic	Mean	
Main pulmonary artery	19/9	10	44%
Right ventricle	32/3		42,41%
Right atrium	a= 30, x= 20 v= 25, y= 15	24	52, 51%
Ascending aorta	105/70	80	95%
Left ventricle	105/5		97%

Cardiac output (dye curve) 1.12 litres/min Diastolic gradient across tricuspid valve = 18 mmHg

There was a systolic gradient of 13 mmHg across the pulmonary outflow tract and the right atrial pressures were markedly raised (mean = 24 mmHg) A prominent diastolic gradient across the tricuspid valve was recorded and a systolic notch was seen on the systolic upstroke of the right ventricular tracing (Fig. 8). A large ball-like filling defect occupied almost the whole of the right atrium and it could be seen moving in and out of the tricuspid valve into the right ventricle, occupying a large portion of the right ventricle and extending up into the pulmonary valve (Fig. 9). Venous laevocardiogram did not show any filling defect in the left atrium. A root aortogram revealed the absence of aortic incompetence.

She underwent cardiac surgery on 9.7.75. At operation, the right atrium was distended and moderately hypertrophied. The right ventricle was distended and contracted poorly. Both the right atrium and right ventricle were completely filled up by a greenish and haemorrhagic jelly-like tumour (Fig. 10). The tumour was 6 x 6 x 3 cm, weighed 120 gm, and had a volume of 150 ml. A small granular area was found on the septal cusp of the tricuspid valve. The tricuspid valve ring was markedly dilated and the left atrium was free of any tumour. The myxoma was removed and the whole thickness of the posterior inter-atrial septum was excised. Unfortunately, the patient developed severe acute pulmonary oedema and did not survive.

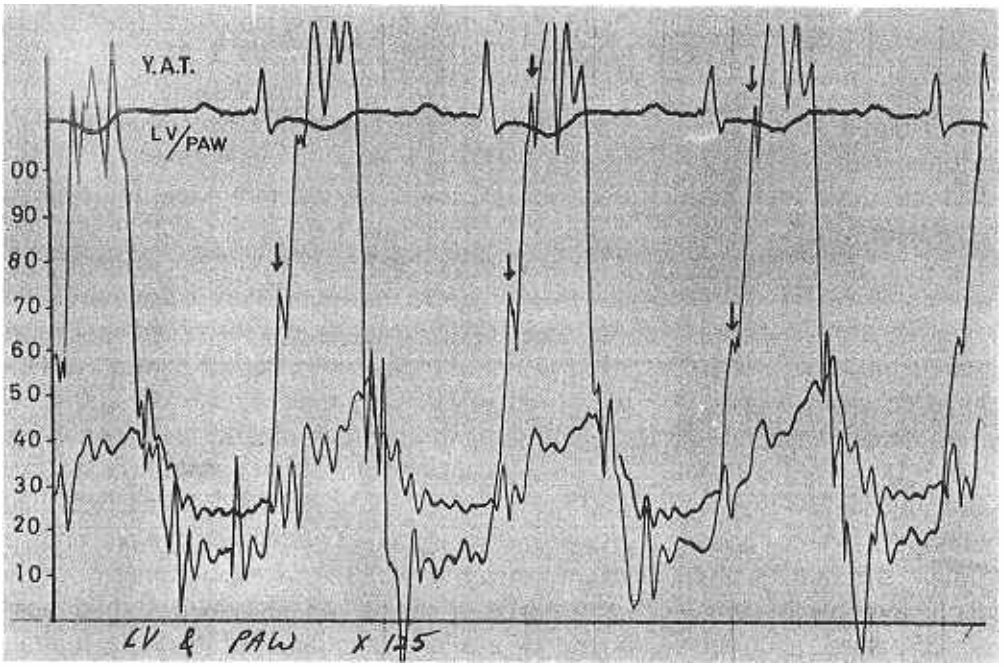

Fig. 4
The simultaneous left ventricular and pulmonary artery 'wedge' pressure tracings. This shows the mitral diastolic gradient. The arrows indicate the typical systolic notch on the upstroke of the left ventricular pressure tracing which is typical of left atrial myxomas.

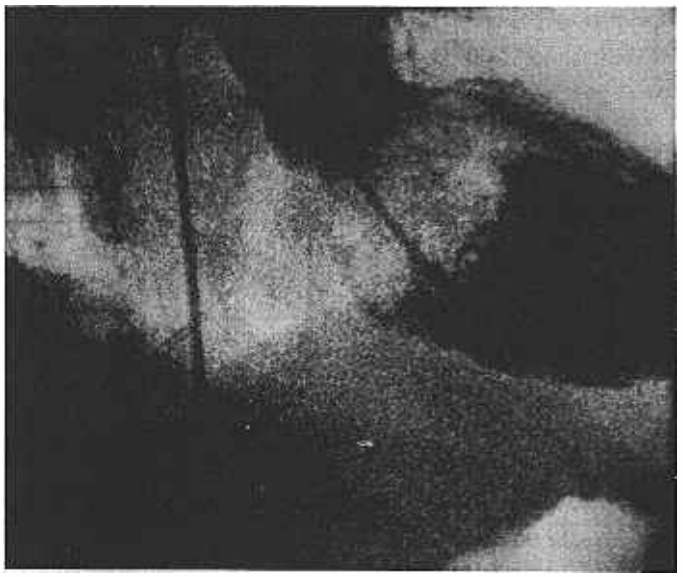

Fig. 5
The left ventricular angiogram of Case 1 in the right anterior oblique view. In this diastolic frame, the radiolucent lobulated tumour mass has prolapsed into the left ventricular cavity.

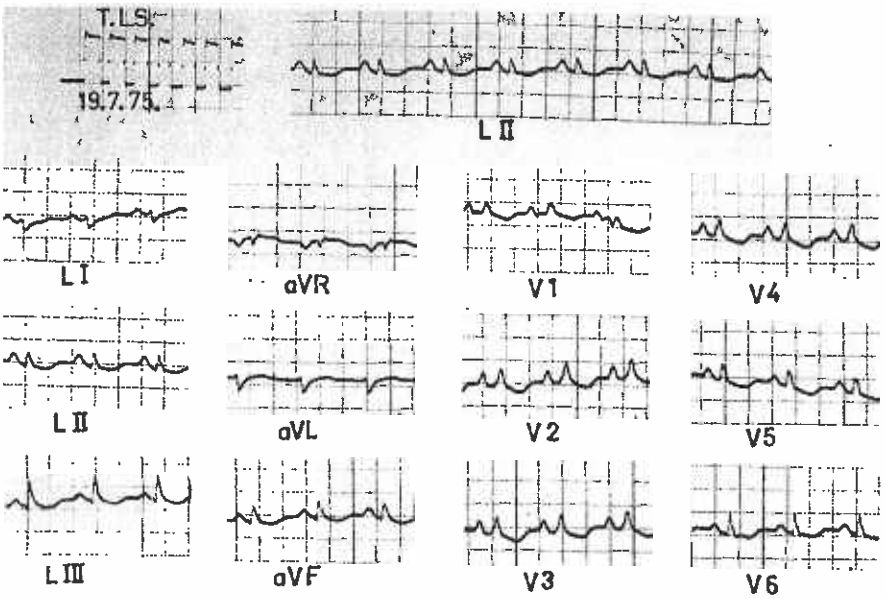

Fig. 6
12-lead electrocardiogram of Case 2 showing generalised low voltage, the prominent P waves in leads II, V1 to V4, the right bundle branch block pattern.

DISCUSSION

Atrial myxomata can present in one or more ways: (a) by obstruction to the flow of blood, (b) by embolisation or (c) with constitutional manifestation (Goodwill, 1963). Case 1 had all three features, while Case 2 presented with the latter two features.

CLINICAL FEATURES

Myxomas are the most common intra-cavitary tumours in adults between 30 to 60 years (Harvey, 1957). Left atrial myxomas are three times more common than right atrial tumours (Pritchard, 1951), while bi-atrial myxomata are uncommon (Cummings, 1961). It is more common in females compared to males, the female:male sex ratio is 3:1 (Harvey, 1957). Systemic embolisation has been reported to occur in 40 per cent of patients with left atrial myxoma (Aldridge, 1960). Case 1 developed transient left hemiparesis due to embolism of a portion of the myxoma. Multiple cystic bone lesions caused by systemic emboli have been described (Wager, 1972).

Obstruction to forward flow in both patients produce hypotensive episodes and low cardiac output. Low cardiac output in turn produces exertional dyspnoea and easy fatigibility. In Case 1, this obstruction caused pulmonary arterial hypertension, while Case 2 had signs of right heart failure with raised jugular venous pulse, peripheral oedema, ascites and hepatomegaly. Dizziness and syncope related to change in body posture are clues to the possible presence of an intra-cavitary tumour. Symptoms and signs caused by mechanical obstruction usually fail to respond satisfactorily to digitalis and diuretics and they usually demonstrate a progressively downhill course.

CONSTITUTIONAL FEATURES

The features have been ascribed to autoimmune reaction to the tumour tissue or to necrosis of the myxoma (Croney, 1967). Fever, weight loss, anaemia and raised ESR are common features and were seen in both cases. In Case 2, haemolytic anaemia was due to mechanical trauma — a wrecking ball effect on erythrocytes. Thrombocytopenia, too, is attributed to this effect (Goodwin, 1968).

AUSCULTATORY FINDINGS

The murmurs produced by myxomas are variable, and can mimic mitral or tricuspid valve disease. Right atrial myxomas, produce characteristic auscultatory findings (Morissey, 1963). These include a slow rumbling diastolic murmur along the left sternal border, tricuspid incompetence, diastolic third heart sound and less commonly a pericardial friction rub. In Case 2, the ejection click, the ejection systolic murmur and the early diastolic murmur (Fig. 7) were probably due to the encroachment of the tumour on to the pulmonary valve producing stenosis and incompetence of the pulmonary valve. What was more important, Case 2 showed variability of murmurs in the same patient on successive days. This is an important clue which may suggest the presence of myxoma. In left atrial myxoma, the first heart sound has been described as loud, and widely split — the latter simulating the presence of a fourth heart sound. The 'tumour plop (TP)' or the early diastolic sound can stimulate a third heart sound or a short rumbling and diastolic murmur (Fig. 1). It has been attributed to a sudden arrest of the forward movement of the tumour into the left ventricle.

ELECTROCARDIOGRAPHIC FEATURES

Left atrial myxomata do not produce diagnostic electrocardiographic changes (Nasser, 1972). Arrhythmias too are uncommon, although atrial fibrillation and flutter may sometimes occur. Case 1 showed sinus rhythm, and changes of severe right ventricular hypertrophy due to obstructive pulmonary arterial hypertension. ECG changes in right atrial myxoma (Fig. 6) are not specific, but they commonly show low voltage, and prominent P wave in leads II, III, V1 to V3 (Morrissey, 1963). Right bundle branch is not uncommon, while arrhythmias are not common.

HAEMODYNAMIC FINDINGS

Mechanical obstruction cause a rise in pressure in the compartments downstream and explain the raised pulmonary wedge and pulmonary artery pressure in Case 1. In left atrial myxomas a diastolic gradient across the mitral valve is produced (Fig. 4), but unlike mitral stenosis where there is a dominant 'a' wave seen, a dominant 'v' wave is seen instead even though mitral incompetence is absent (Nasser, 1972). In right atrial myxoma, there is a diastolic gradient across the tricuspid valve (Morrissey, 1963) A characteristic fining in both right and left atrial tumour, is the presence of a notch on the upstroke of the ventricular pressure pulse (Ramsey, 1969; Pitt, 1967). This notch is due to the movement of

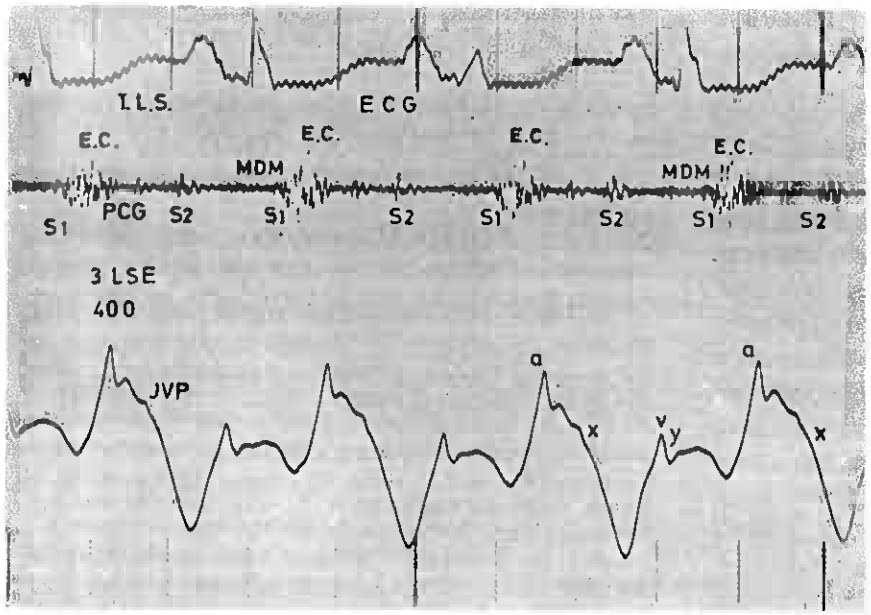

Fig. 7
The phonocardiogram and simultaneous jugular venous pressure tracing of Case 2 illustrating the ejection systolic murmur with multiple ejection clicks at the left sternal border. A mid-diastolic murmur is demonstrated.

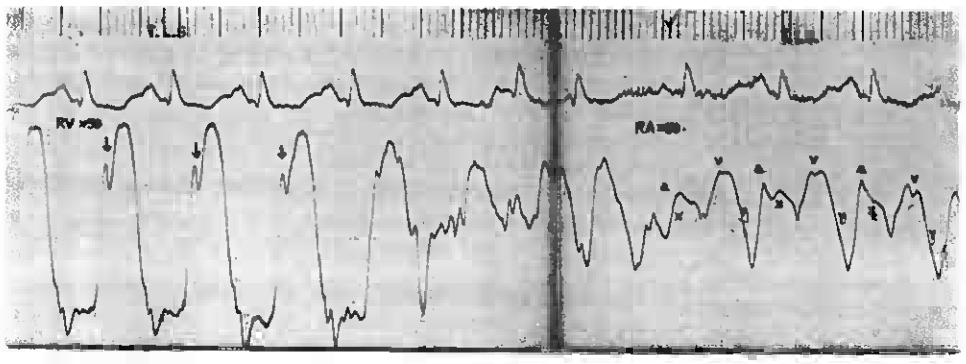

Fig. 8
The withdrawal pressure tracing of Case 2 which clearly demonstrates the gradient across the tricuspid stimulating tricuspid stenosis. The arrows indicate the systolic notch on the upstroke of the right ventricular waveform and is typical of right atrial myxomas.

Fig. 9
The right atrial angiograms of Case 2 showing a large lobulated tumour filling the whole of the right atrium and the right ventricle.

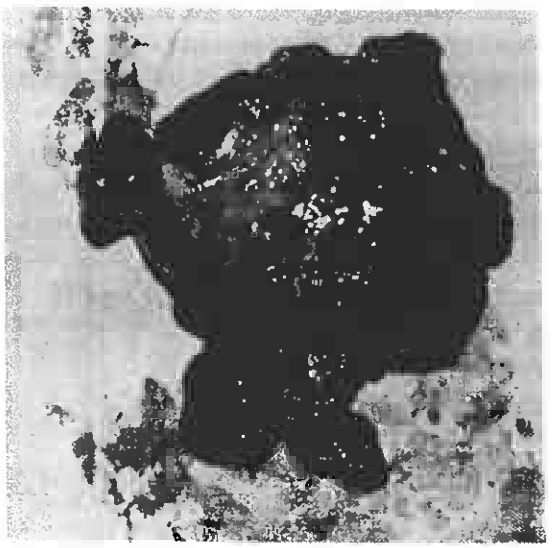

Fig. 10
The right atrial tumour of Case 2 was a haemorrhagic and greenish jelly like tumour weighing 120 gm and measured 6 x 6 x 3 cm.

the tumour in early systole back into the atrium causing loss of volume and tension in the ventricle, resulting in a sudden fall in pressure.

ECHOCARDIOGRAPHY (Fig. 3)

Echocardiography is an excellent, simple and non-invasive bedside technique for the detection of atrial myxomata. Its use for this purpose has been described as early as 1959 (Edler, 1961). In left atrial myxoma, the anterior mitral valve leaflet has a square wave pattern and clouds of echoes are seen behidn this valve and in the left atrium (Wolfe, 1969). However, echocardiography is less useful for right atrial myxoma (Harhold, 1973).

CONCLUSION

The severe pulmonary oedema in Case 2 was due to the sudden removal of mechanical obstruction to system venous return. A more aggressive approach to haemodynamic monitoring and early treatment of increasing left atrial pressure could have prevented this complication. Both these cases which were diagnosed during life illustrate that, although myxomata are uncommon, they are not all that rare, provided there is a high index of suspicion. Non-invasive tools, like echocardiography can be used as a screening procedure to diagnose this 'curable tumour'.

REFERENCES

Barhnson HT, Newman EG: Diagnosis and removal of intra-cavitary tumours of the right atrium. Bull Johns Hopkins Hosp, 39: 150-163, 1953.

Benjamin HS: Primary fibromyxoma of the heart. Arch Path, 27: 950, 1939.

Crawford C: Discussion on late results of mitral commissurotomy. Henry Ford International Symposium on Cardiovascular Surgery; Studies in Physiology. Diagnosis and techniques (Lam CR, ed). Philadelphia and London, WB Saunders; 1955, page 202-203.

Cumming GR, Finkel K: Intracardiac myxoma involving the right and left atria in a young patient. J Paed, 58: 559-567, 1961.

Currey HFL, Mathews JA, Robinson J: Right atrial myxoma mimicking a rheumatic disorder. Brit Med J, 1: 547-548, 1962.

Goldberg HP, Glenn F, Dotter CT, et al: Myxoma of the left atrium. Diagnosis made during life with operative and post-mortem findings. Circulation, 6: 762-767, 1952.

Goodwin JE: Diagnosis of left atrial myxoma. Lancet, 1: 464-468, 1963.

Harhold NB Jr, Gau GT: Echocardiographic diagnosis of right atrial myxoma. Mayo Clinic Proc, 48: 284-286, 1973.

Harvey WP: Clinical aspects of cardiac tumours. Am J Cardiol, 21: 328-343, 1968.

King TW: On simple vascular growths in the left auricle of the heart. Lancet 2: 428-430, 1845.

Morrissey JF* Campeti FL, Mahoney EB, Yu P: Right atrial myxoma Am Heart J, 66: 4-14, 1963.

Nasser WK, Davis RH, Dillon JC et al: Atrial myxoma. Am Heart J, 83: 694-704, 810-824, 1972.

Strauss R, Merlin R. Primary tumour of the heart. Arch Pathol, 39: 74-78, 1945.

Walton JA, Kahn DR, Willis PW: Recurrence of left atrial myxoma. Am J Cardiol, 29: 872-876, 1972.

Wolfe SB, Popp RL, Feigenbaum H: Diagnosis of atrial tumours by ultrasound. Circulation, 39: 615-62, 1969.