CASE REPORTS

RIGHT ATRIAL MYXOMA MISTAKEN FOR CONSTRICTIVE PERICARDITIS

BY

R. W. EMANUEL and W. E. LLOYD

From the Brompton Hospital

Right atrial myxomas are rare. The diagnosis can sometimes be made clinically and confirmed radiologically if the tumour is calcified but angiocardiography is usually required (Differding, Gardner, and Roe, 1961). The diagnosis is seldom made by cardiac catheterization alone, and this is partly due to the difficulty in detecting minor degrees of obstruction at the tricuspid valve, as illustrated by the case recorded here. This case also demonstrates the importance of the differential diagnoses between right atrial myxoma and constrictive pericarditis (Bahnson and Newman, 1953; Ellis, Mankin, and Burchell, 1958; Padhi, Kelly, and Lynn, 1959).

Case Report

Mrs. A., aged 26 years, first attended the Brompton Hospital in July 1954. Her symptoms, which had been present for several months, included fatigue, loss of 10 lb. (4·5 kg.) in weight, an occasional evening temperature up to 99·5° F. (37·5° C.), slight cough with yellow sputum but no hæmoptyses. There were crepitations over the right middle lobe and the chest radiograph showed a small anterior pleural effusion displacing the heart to the left (Fig. 1 A and B). The Mantoux test was strongly positive to 1:1000 old tuberculin. Additional investigations included Hb 91 per cent, white blood cells 6000/c.mm., E.S.R. (Westergren) 5 mm. in the first hour: sputum smears, cultures, and gastric washings were negative for tuberculosis. The signs over the right middle lobe cleared rapidly and the chest radiograph returned to normal within a week (Fig. 1C). Her father was known to have pulmonary tuberculosis. During the next four years fatigue continued, and she reported an occasional evening temperature of 99° F. (37·2° C.). Physical examination, chest radiographs, and E.S.R. remained normal.

In June 1960 she developed left-sided pleuritic pain and reattended hospital on July 9, when "a loud systolic crunching noise at the lower end of the sternum" was heard for the first time. There was no comment about the venous pressure. Chest radiographs showed a small opacity in the periphery of the left mid zone, which cleared rapidly, but the right atrium and pulmonary artery were both a little more prominent than in 1954 (Fig. 1D). The electrocardiogram was normal. Fatigue, slight breathlessness, and nocturnal sweating continued. The E.S.R. in July and August was persistently raised, 40, 26, and 34 mm. in the first hour.

On October 22 she was seen by one of us (R.W.E.) because of the murmur. The venous pressure was raised, 15 cm. above the sternal angle, and the dominant wave was a sharp *y* descent and trough. Kussmaul's sign was strongly positive, and the liver firm and moderately enlarged. The arterial pulse was small and paradoxical and neither ventricle was palpable. A loud rough murmur filled the whole of systole and continued into the first third of diastole. The murmur was widely conducted, but maximal on the left at the lower end of the sternum. The radiograph confirmed slight cardiac enlargement already noted in June (Fig. 1D). There were now low voltage positive T waves in the standard and left chest leads. She was admitted to hospital with a diagnosis of active tuberculous pericarditis, and treatment with sodium para-amino salicylate (P.A.S.) and isoniazid (I.N.A.H.) was started. Within a week the E.S.R. had fallen to 10 mm. and never again exceeded 17 mm. in the first hour. The serum proteins were 6·7 g./100 ml. with albumin 4·1 g./100 ml., and there was a slight increase in the globulin fraction which returned to normal three months later. On the seventh day of antituberculous treatment she developed a high fever 102°–105° F. (38·9°–40·6° C.) which persisted until both P.A.S. and I.N.A.H. were withdrawn. Reintroduction of either of these drugs caused a further febrile reaction, and it was not until February 1961 that she was fully

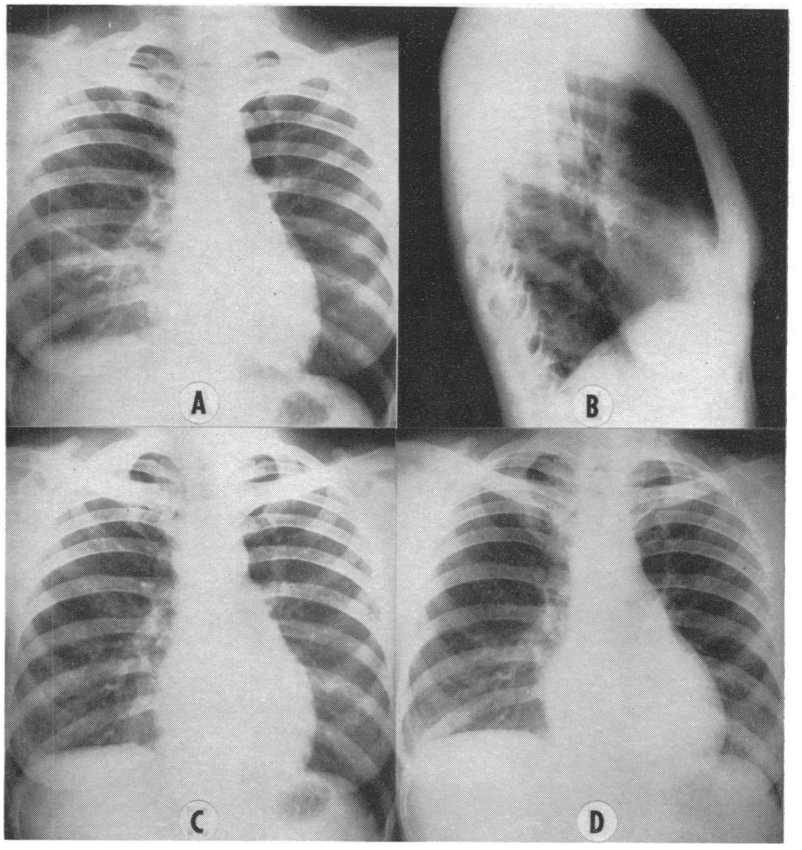

FIG. 1.—Chest radiographs. (A) and (B) July 25, 1954, small anterior pleural effusion extending into lesser fissure with displacement of the heart to the left. (C) July 30, 1954, after absorption of pleural effusion. (D) July 1960, increased size of right atrium and fullness of pulmonary artery.

desensitized. The physical signs remained unchanged except for the murmur, which varied in intensity and length, and a presystolic element was heard on several occasions. The murmurs were accepted as exocardial except by one of us (W.E.L.) who thought throughout that from their character they could well be endocardial and were reminiscent of mitral stenosis.

Cardiac Catheterization (December 29, 1960 by R.W.E.). Two catheters were introduced, one from each arm, thus allowing simultaneous pressures to be recorded from two intracardiac sites. The results are summarized in Fig. 2 A and B. Photographic superimposition of simultaneously recorded right atrial and ventricular pressure tracings (Fig. 2C) demonstrated a gradient that was not appreciated when these tracings were originally analysed (Fig. 2B). The atrial *a* wave was considerably higher than the *a* wave transmitted to the ventricle, indicating some obstruction during atrial systole. Similarly during ventricular diastole, the *y* trough in the atrial tracing was less deep than its counterpart, the early diastolic dip, in the right ventricular pressure tracing, again suggesting obstruction between atrium and ventricle at this phase in the cardiac cycle. The rapid *y* descent, however, denied significant obstruction during the period of rapid ventricular filling, but would be compatible with tricuspid regurgitation presumably due to imperfect valve closure, which would also account for the relatively high *v* wave in the atrial pressure pulse. When the simultaneous right atrial and right ventricular pressure tracings were recorded the atrial tracing was damped (Fig. 2B compared with Fig. 2A). From the initial right atrial tracing (Fig. 2A) it is apparent that the pressure difference during atrial systole is, in fact, greater than that shown in Fig. 2C. The pulmonary arterial pressure was 8 to 13/4 to 7 mm. (mean 6); right brachial arterial pressure 83 to 88/64 to 66 (mean 73) mm. Hg.

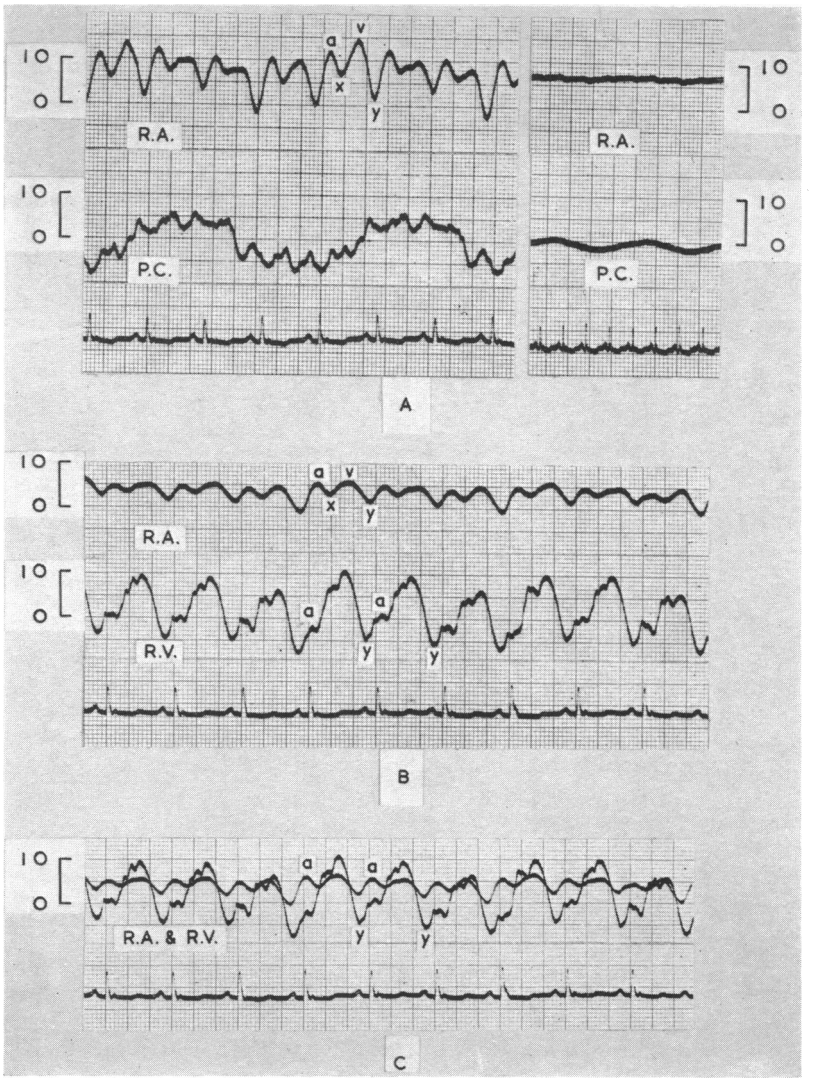

FIG. 2.—(A) Simultaneously recorded right atrial (R.A.) and indirect left atrial wedge (P.C.) pressure tracings. R.A. a=7 to 13, x=6 to 8, v=8 to 15, y=−1 to 4, mean=7 to 8 mm. Hg. P.C. a=2, x=−1, v=4, y=0, average owing to respiratory swing, mean=−1 mm. Hg. (B) Simultaneously recorded right atrial (R.A.) and right ventricular (R.V.) pressure tracings R.A. a=4 to 6, x=3 to 4, v=5 to 6, y=0 to 2 mm. Hg. R.V. a=−3 to 1, v=6 to 11, early diastolic dip −7 to −4 mm. Hg. (C) The same synchronous right atrial and right ventricular pressure tracings as in (B) superimposed to show *a* wave gradient and *y* trough gradient.

Resting oxygen uptake 247 ml./min. Arteriovenous oxygen difference 71 ml./l. Resting cardiac output 3·5 l./min. (Fick). Pulmonary vascular resistance less than 3 units. All pressures were taken from the sternal angle.

On February 15, while on full antituberculous therapy, she developed a second attack of left-sided pleurisy with a fever of 102° F. (38·9° C.). The pain and pyrexia subsided rapidly as a left pleural effusion developed. The sputum contained coagulase negative staphylococci, *Streptococcus viridans*, and *Neisseria catarrhalis*. White blood cells 6500/c.mm., E.S.R. 9 mm. in the first hour. The radiograph, in addition

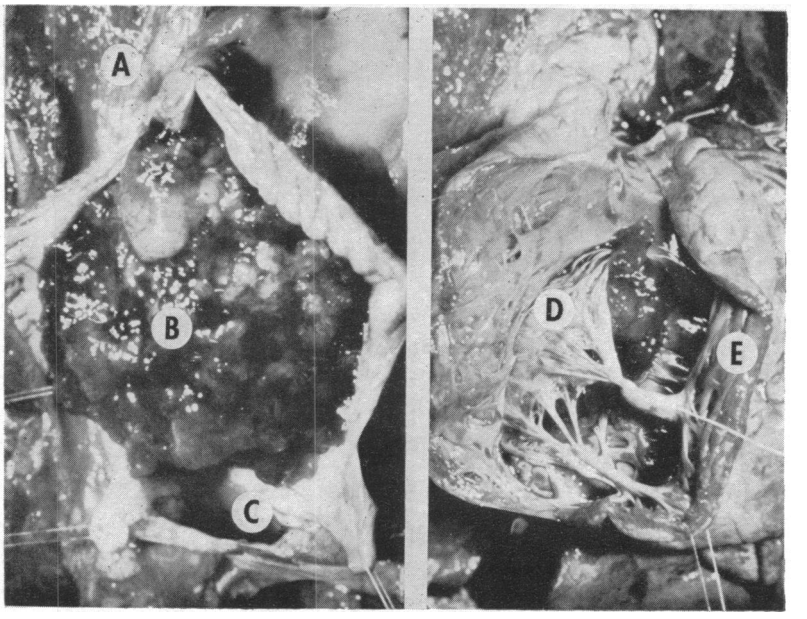

FIG. 3.—Post-mortem appearance of right atrial myxoma. (A) Superior vena cava. (B) Myxoma. (C) Inferior vena cava. (D) Tricuspid valve with myxoma protruding through the valve into the right ventricle. (E) Right ventricle.

to a left pleural effusion, showed small areas of collapse at both bases. On February 18 the cardiogram which had remained unchanged, showed T inversion in V3, V4, and V5: by March 15 these changes were less. She made a rapid recovery and she returned home on March 29. She died suddenly on April 22.

Necropsy (April 24, 1961 by Dr. Donald Teare). A large glistening gelatinous tumour occupied the whole of the right atrium. Part of the mass protruded through the tricuspid valve into the right ventricle (Fig. 3). The macroscopic and microscopic appearances were those of myxoma. Death was due to obstruction of the main pulmonary artery by portions of the myxoma which had become detached from the main atrial mass. The tumour itself was attached by a short strong pedicle to the atrial septum above and slightly anterior to the closed foramen ovale. The pericardium was normal except for a few ounces of clear straw-coloured fluid. The valves, myocardium, and coronary arteries were also normal. Each pleural cavity contained a little fluid. The lower part of the right lung was densely adherent to the chest wall and diaphragm. There were light adhesions over the lower part of the left lung and lingula. There was, however, no evidence recent pulmonary infarction, although a fibrous scar in the lingula might have been the site of a previous episode. In the right lower lobe several small arteries, 3–4 mm. diameter, were blocked by whitish material which histological examination showed to be myxoma. No tumour emboli were found in the left lung and there was no evidence of pulmonary tuberculosis.

Discussion

This case illustrates the difficulty of detecting slight obstruction at the tricuspid valve and the problem of differentiating between active constrictive pericarditis and right atrial myxoma.

Clinically the differential diagnosis appeared to be active constrictive pericarditis or cardiomyopathy. The form of the venous pulse, its rapid *y* descent and *y* trough, with similar findings in the right atrial and ventricular pressure tracings, were thought to exclude the possibility of an obstructive tricuspid lesion, a diagnosis that was considered several times at the bed side. The similar behaviour of right and left atrial (indirect) pressures during Valsalva's manœuvre, passive leg raising, and exercise, which was demonstrated at cardiac catheterization, excluded the possibility of a

cardiomyopathy (Wood, 1961). The hæmodynamic findings, therefore, appeared to confirm the diagnosis of active constrictive pericarditis, although the difference between the mean atrial pressures was obviously unusual (Fig. 2A). Wood (1961) found that the normal atrial pressure relationship was retained in 18 of the 20 cases of constrictive pericarditis investigated, and in the remaining 2 the mean right atrial pressure was only 3 mm. higher than the left. In our case the resting mean right atrial pressure was 8 to 9 mm. higher than the mean left atrial pressure (indirect). These findings were discussed with Dr. Paul Wood, and it was decided that in the absence of any evidence, as we then thought, of tricuspid valve obstruction, the pressure difference of 8 to 9 mm. Hg was acceptable, although outside the range previously encountered in constrictive pericarditis.

As this case was incorrectly diagnosed as constrictive pericarditis the simultaneously recorded right atrial and right ventricular pressure pulses have been compared with those from five proven cases of constrictive pericarditis (three of these cases were investigated by the two catheter technique already described, and two by differential manometry). In the cases of constrictive pericarditis there was an insignificant gradient (plus or minus 1·0 mm. Hg) during atrial systole, whereas in the case with the myxoma the right atrial *a* wave was 5·4 mm. Hg higher than that recorded in the ventricle. Similarly in the cases of constrictive pericarditis the right atrial *y* trough was only slightly higher than the ventricular pressure (2·2 mm. Hg average, range 1·3–3·0 mm. Hg), but in the case of the myxoma the pressure gradient at this point in the cardiac cycle was increased to 6·3 mm. Hg. This analysis emphasizes the difference in the hæmodynamics of constrictive pericarditis and conditions in which there is partial intermittent obstruction at the tricuspid valve.

Summary

A case of right atrial myxoma is described in which partial intermittent obstruction of the tricuspid valve caused clinical and hæmodynamic findings which closely resembled those of active constrictive pericarditis.

We are grateful to Dr. Paul Wood for his help with this case and for permission to publish the catheter data from five of his cases of constrictive pericarditis. Cardiac catheterization in four of these was carried out by one of us (R.W.E.); and in the remaining case by Dr. Arnold Katz at the National Heart Hospital. We are indebted to our colleagues in the radiological department at the Brompton Hospital and to Dr. Peter Kerley; also to Dr. K. F. Hinson and his colleagues in the pathological department. We should also like to thank Dr. S. W. Kuper, Mr. A. Curd, and Miss N. Lampen for the photographs.

References

Bahnson, H. T., and Newman, E. V. (1953). *Bull. Johns Hopk. Hosp.*, **93**, 150.
Differding, J. T., Gardner, R. E., and Roe, B. B. (1961). *Circulation*, **23**, 929.
Ellis, F. H., Mankin, H. T., and Burchell, H. B. (1958). *Med. Clin. N. Amer.*, **42**, 1087.
Padhi, R. K., Kelly, H. G., and Lynn, R. B. (1959). *Canad. J. Surg.*, **2**, 414.
Wood, P. (1961). *Amer. J. Cardiol.*, **7**, 48.