MYXOMA OF THE LEFT AURICLE

BY

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Primary tumours of the heart are comparatively rare, and myxomata of the auricles form an important section of the innocent tumours. Some 95 cases of intra-auricular myxoma have so far been reported in the world literature. The majority (77) were in the left auricle. Not one of these cases was diagnosed during life. Mahaim (1945) collected 82 cases in his extensive review of cardiac tumours, and subsequent cases have been reported by Thompson (1944), Burnett and Davidson (1945), Straus and Merliss, 2 cases (1945), Field *et al.* (1945), Rasmussen, 2 cases (1945), Brown (1946), Anderson and Dmytryk (1946), Farreras Valenti *et al.* (1947), Alvayay *et al.* (1947), Macoun (1949), and Allison and Susmann (1949).

The object of this paper is to describe and discuss three additional cases of myxoma of the left auricle and to consider the value of the electrocardiogram as an aid to diagnosis.

Case 1

A woman aged 50 had a normal exercise tolerance and no history of serious illness until the age of 41. During the next five years her capacity for exercise gradually decreased until she became breathless at rest. In May, 1945, at the age of 46, she was admitted to Queen Elizabeth Hospital, Birmingham, after an attack of acute dyspnoea. A clinical diagnosis of atypical coronary thrombosis was made. Serial electrocardiograms were taken (Fig. 1). The dyspnoea was relieved after two weeks' rest in bed. Although the patient remained breathless on effort during the four years following discharge, she was able to undertake periods of sedentary work.

On February 15, 1949, she developed sudden pain and stiffness in the back of the neck together with increasingly severe breathlessness. Twenty-four hours later she was admitted urgently to the West Middlesex Hospital.

On examination she was seen to be very dyspnoeic, but lying flat in bed and producing frothy sputum. The face and extremities were strikingly cyanosed and cold, but the peripheral arteries were palpable. There was no engorgement of the neck veins. The pulse was irregular and its rate over 150. There was slight cardiac enlargement. The heart sounds were normal and no gallop rhythm was present. A soft apical systolic murmur was heard. The blood pressure was 115/? mm. Numerous crepitations were present over both lung fields. Marked neck rigidity was found, but there were no abnormal signs in the central nervous system; Kernig's sign was negative. A lumbar puncture showed the cerebrospinal fluid to be normal.

The dominating clinical picture was that of acute pulmonary oedema, but it was remarkable that when an attempt was made to sit the patient up to relieve the dyspnoea she insisted upon remaining flat. There was no change in the physical state until she died suddenly three hours after admission.

Necropsy. Myxoma Filling the Left Auricle: Pulmonary Oedema.—The heart weighed 415 g. Filling the left auricle was a pedunculated tumour, 6 by 4 by 3 cm., arising from the auricular septum in the region of the foramen ovale. An extension of the tumour passed through the mitral orifice and expanded into the upper part of the ventricle. This extension was demarcated from the main part of the tumour by a deep groove at the level of the mitral ring (Fig. 2). The tumour was rubbery in consistence, and its cut surface gelatinous. A large recent haemorrhage occupied the upper pole. The right auricle was dilated and its wall thickened (5 mm.). There was pronounced right ventricular hypertrophy (9–10 mm.). In all other respects the heart was normal. There was no evidence of old or recent coronary thrombosis. The lungs were oedematous. The brain, meninges, and all other organs were normal in appearance.

Histology.—The tumour was partly covered by a layer of flattened cells. Its substance consisted of a faintly eosinophilic hyaline stroma containing vacuoles. Running throughout were strands of endothelial cells with ovoid and vesicular nuclei (Fig. 3). There were also some scattered plasma cells and lymphocytes. A few capillaries were seen, and in some parts these were encircled by several layers of endothelial cells. Foci of haemorrhage, haemosiderosis, and calcification were present. A large amount of reticulin and a little collagen were demonstrated in the stroma. No mucin was present. The naked-eye and microscopical appearances of this tumour were typical of a cardiac myxoma.

Case 2

A man aged 73 was admitted to the West Middlesex Hospital on June 11, 1947. No history was obtained of recent or past cardiac symptoms, and there were no abnormal findings in the cardiovascular system other than a blood pressure of 168/65 mm. Radiology of the heart showed a normal size and outline. Further examination and investigation revealed the presence of chronic lymphatic leukaemia, and the patient died from this disease two months after admission.

Necropsy. Lymphatic Leukaemia: Myxoma of the Left Auricle.—The heart weighed 435 g. A mobile ovoid tumour, 4.5 by 3 by 1.5 cm., occupied more than half of the cavity of the left auricle. It was attached by a peduncle to the septal wall at the border of the foramen ovale. There was no occlusion by the tumour of the mitral valve or pulmonary veins. All other parts of the heart were normal, and the lungs showed only the changes of senile emphysema. Leukaemic deposits were present in the liver, spleen, and lymph nodes.

Histology.—Sections of the cardiac tumour were similar to those in Case 1, except that the endothelial cells were arranged in groups rather than in strands.

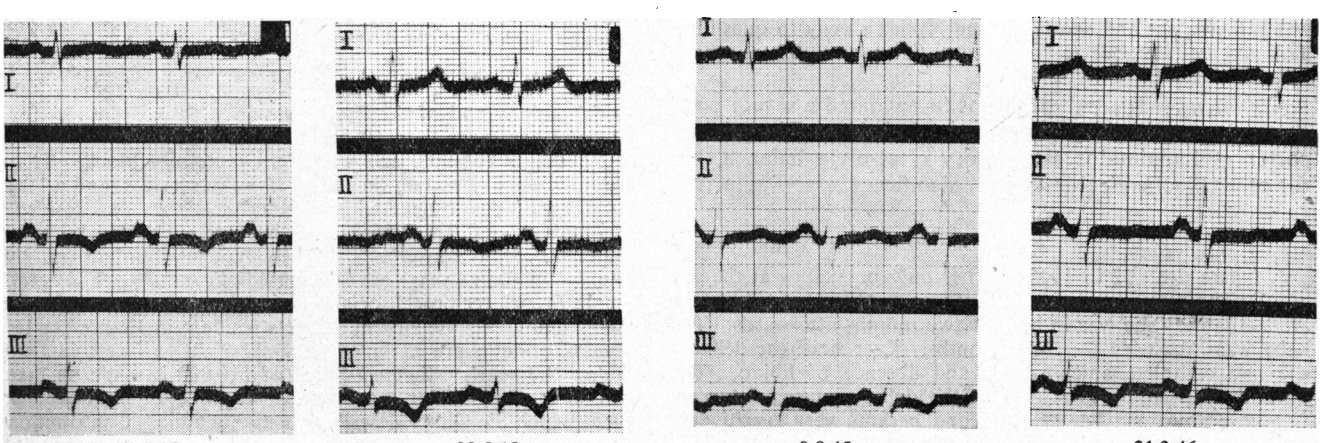

FIG. 1.—Serial electrocardiograms taken during 1945 and 1946.

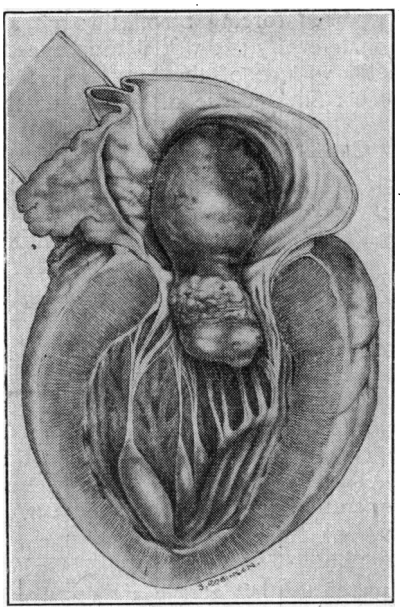

FIG. 2.—Case 1. Drawing of the heart, showing tumour filling the left auricle and expanding into the ventricle. A groove in the tumour can be seen at the level of the mitral ring.

Case 3

A woman aged 72 was admitted to the West Middlesex Hospital on March 16, 1946, in an advanced cachectic state. There was no previous history of breathlessness, but attacks of palpitation on exertion had limited her activities for some years. Examination revealed widespread pelvic carcinomatosis. Death occurred five days later.

Necropsy. Carcinoma of the Body of the Uterus with Secondary Deposits: Myxoma of the Left Auricle. — The heart weighed 380 g. A firm spherical tumour 5.5 cm. in diameter filled more than three-quarters of the left auricle. It was attached by a short pedicle to the interauricular septum near the foramen ovale and did not occlude any orifice of the auricle. There was a slight increase in the thickness of the right ventricular wall (5–6 mm.), but no other abnormality in the heart and great vessels. A carcinoma of the body of the uterus was present, with numerous secondary deposits in the pelvis and in both lungs.

Histology.—Sections from the auricular tumour were essentially similar to those described in Case 1.

Pathology

Myxomata of the auricles are generally accepted as being true neoplasms which are derived from embryonic mucoid tissue persisting usually in the rim of the foramen ovale (Ribbert, 1924; Yater, 1931). Some writers, however, consider that they are organized thrombi which have undergone myxomatous degeneration (Husten, 1922; Hamilton-Paterson and Castelden, 1942). Whatever their origin, these tumours have a characteristic macroscopic and microscopical appearance, which is described fully by Yater. The three tumours detailed above conform to this description.

Clinical Features

Examination of the 77 recorded cases of myxoma of the left auricle shows that the most common age at death is between 40 and 60 years in both sexes. The range is wide—from 4 years (Jacobsthal, 1900) to 83 years (Curtis, 1872). Females are affected nearly three times more often than males.

The presence of a left auricular myxoma has not been recognized during life, and there is no known characteristic clinical picture. Nevertheless, certain cardiovascular symptoms and signs often occur owing to obstruction of the circulation by the tumour. More than one or two of these effects are seldom found in any one case. These cardiovascular features have been discussed fully by Fawcett and Ward (1939) and Mahaim (1945), and may be summarized as follows:

1. A history usually of less than two years' duration, the chief symptom being breathlessness on effort.

2. Congestive cardiac failure which in spite of every treatment pursues a relentless course to death. This apparently is due to gradual obliteration of the auricle by the growing tumour.

3. An apical diastolic murmur identical with that of mitral stenosis. This is often strikingly variable, and is thought to be due to changing degrees of obstruction of the mitral valve by the tumour.

4. Attacks of acute dyspnoea which are sometimes remarkable in that the patient prefers to lie flat.

5. Attacks of abrupt loss of consciousness usually associated with the assumption of the upright posture. These attacks of syncope and dyspnoea may be accounted for by temporary obstruction to the mitral valve or pulmonary veins by the mobile tumour.

6. An abnormal *x*-ray outline due to enlargement of the left auricle.

7. Embolic phenomena caused by fragments of myxomatous tissue becoming detached from the tumour. If an accessible artery is involved then biopsy of the embolus should establish the diagnosis.

8. Sudden death. This is thought in some cases to be caused by the tumour falling into and completely occluding the mitral valve or pulmonary veins.

In Case 1 the myxoma was evidently the sole cause of the symptoms and subsequent death. This case presented few of the features mentioned above. One striking feature which might have suggested the correct diagnosis was that the patient, while gasping for breath in an unexplained attack of acute pulmonary oedema, refused to be raised to the sitting position. Similar in this respect is the case described by Field *et al.* (1945). Unexpected effects of posture on symptoms are said to be characteristic of mobile intracardiac tumours.

It is difficult to account for the presence in Case 1 of the severe persistent pain in the back of the neck. No explanation for this was found in the brain and meninges at necropsy. It is, however, possible that it was a referred pain of myocardial ischaemia due to the tumour in the auricle and mitral orifice reducing coronary flow. Miller (1942) indicates that referred cardiac pain may occasionally be localized to the back of the neck. A similar pain was described without explanation in the case of Field *et al.* (1945).

In Case 2, although the tumour was large enough to occupy half the auricular cavity, it did not produce any

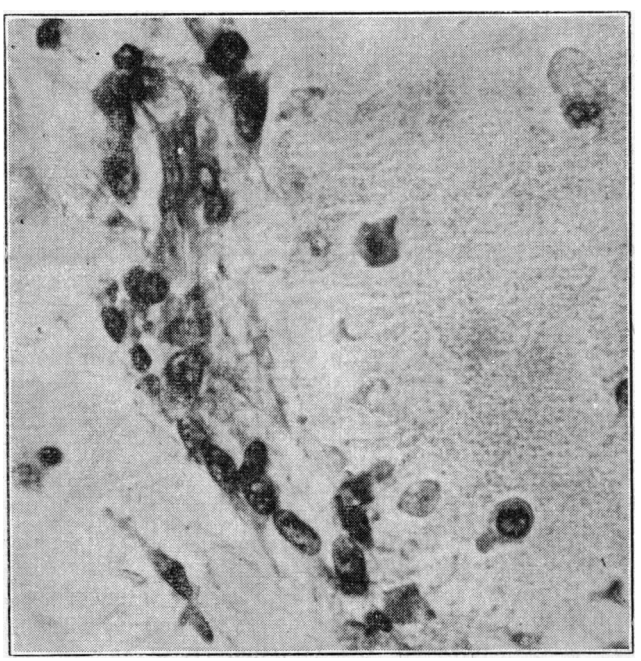

FIG. 3.—Case 1. Section from tumour showing a cord of endothelial cells. Stained with H. and E. (× 500.)

symptoms or signs, nor was it the cause of death. In Case 3 the tumour was even larger than in Case 2, yet, with the possible exception of the attacks of palpitation on effort, there were again no effects on the patient. It thus seems that the auricle is able to accommodate tumours of a large size without any great handicap to the function of the heart.

Electrocardiograms

In cases of left auricular myxoma in which the tumour is large enough to produce chronic obstruction to the circulation, myocardial strain will occur in the left auricle, right ventricle, and right auricle. Certain electrocardiographic changes may thus be expected in theory: the auricular strain may produce P waves of increased height or duration, and the right ventricular strain may result in combined S–T segment depression and T-wave inversion in Lead II and Lead III (Goldberger, 1947).

In a number of reports on cases of myxoma of the left auricle electrocardiograms are described, but in only a few of these are the tracings presented (Houck and Bennett, 1930; Gilchrist and Millar, 1936; Thompson, 1944; Rasmussen, 1945; Field *et al.*, 1945). On examination of these, only the case of Gilchrist and Millar shows abnormal P waves, the voltage being higher than 0.25 mv. Possible S–T segment and T-wave changes are obscured by digitalis therapy in all cases except that of Field *et al.* In their case, although right ventricular hypertrophy was present at necropsy, there is no clear evidence of right ventricular strain in the electrocardiogram.

In Case 1 four electrocardiograms were taken during 1945 and 1946 (Fig. 1). In these it can be seen that the P wave in Lead II is abnormal. It is bifid and prolonged in duration (0.11 sec. to 0.12 sec.) and resembles that which occurs with mitral stenosis. The S–T segment is depressed and the T-wave inverted in Lead III and usually in Lead II. No digitalis therapy complicates the records. It would therefore seem that the electrocardiographic changes indicate combined auricular and right ventricular strain present four years before death. This interpretation is supported by the necropsy findings.

The value of the electrocardiogram as an aid to diagnosis is difficult to assess. However, it is likely that evidence in the tracings of auricular and right ventricular strain without any clear clinical explanation may indicate the presence of a tumour in the left auricle.

Sudden Death

The usual explanation given for sudden death occurring in a case of left-sided auricular myxoma is that the tumour falls into and permanently obstructs the mitral orifice or pulmonary vein openings. In Case 1 there was every appearance at necropsy that the tumour had extended through the mitral valve long before death: there was the marked groove encircling the tumour shaped closely to the mitral ring and also the expansion of the tumour beyond it which had a diameter greater than that of the mitral orifice (Fig. 2). Death in this case was therefore not caused by the tumour suddenly falling into the mitral valve. Further, owing to the fixation of the tumour in the mitral orifice it could not have caused death by falling into the pulmonary vein openings.

In Case 1 and in four other reported cases in which rapid death occurred (Yater, 1931; Clerc *et al.*, 1937; Fawcett and Ward, 1939; Field *et al.*, 1945) it was found at necropsy that the myxoma almost completely filled the auricle concerned, and, further, that recent haemorrhage involved a large part of the tumour. In each of these five cases the heart had been forcing blood through an almost obliterated auricular cavity; it would thus appear that the rapid increase in volume of the tumour due to haemorrhage was the most likely cause of sudden death.

Comment

The diagnosis of left auricular myxoma is an extremely difficult problem. If, however, this tumour is suspected clinically, then angiocardiography may possibly confirm the presence of an intra-auricular mass.

The recognition of this condition during life may cease to be of purely academic interest, as surgical removal of the tumour may shortly become practicable. Björk (1948) described the successful use of a cardio-pulmonary machine to perfuse the brain of animals when the blood flow through the heart was stopped for over half an hour. He quoted Crafoord as considering that it should be possible in the future, with the use of such an apparatus, to carry out bloodless intracardiac operations on the human subject.

Summary

Three cases of myxoma of the left auricle are described. One case presented an interesting postural symptom and also an unusual pain which may have been due to myocardial ischaemia. The other two cases were remarkable in that a large tumour was present in each without any obvious cardiac dysfunction.

Abnormalities in the electrocardiogram are considered. It is suggested that graphic evidence of auricular and right ventricular strain, without any clear clinical explanation, may indicate the presence of a tumour in the left auricle.

The causation of sudden death is discussed. It is possible that haemorrhage into the tumour may produce sudden death in some cases.

It is noted that surgical removal of such tumours may become practicable in the near future.

I wish to thank Dr. N. F. Coghill and Mr. J. Scholefield for permission to publish their cases. I am greatly indebted to Professor K. D. Wilkinson for the use of the clinical records and electrocardiograms from Queen Elizabeth Hospital, Birmingham, where the first case was previously under his care. I also wish to thank Dr. A. C. Counsell for his assistance and advice over the pathological specimens, Miss S. Robinson for the drawing of the heart, and Mr. D. A. Vinter for the photomicrograph.

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