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CARDIAC MYXOMA

BY

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Primary neoplasms of the heart are rare and only found in 0.05 per cent of autopsies. Seventy-five per cent are benign, and the majority of these are true myxomas (Lymburner, 1934).

The case to be described is an example of a myxoma of the right auricle presenting the clinical picture of chronic superior vena caval obstruction, and although this patient was under observation over a period of twenty-nine months the true diagnosis of intracardiac tumour was never made. We record one further failure to diagnose an innocent cardiac neoplasm correctly during life. Review of the literature on this subject fails to reveal that this has ever been successfully accomplished, although primary malignant growths of sarcomatous type have been correctly diagnosed in life by both Pavlovsky (quoted by Fawcett and Ward, 1939) and Shelburne (1935).

Case History

N. P., a woman aged 64 years, was first seen in October, 1940. She gave a history of breathlessness on exertion since 1939, accompanied by occasional "choking fits" on swallowing, but no dysphagia. She had observed swelling of the face and engorgement of the right external jugular vein for the previous three weeks, but there was no history of cough, pain, palpitation, or swelling of the ankles. There was no past history of rheumatic fever, chorea, syphilis, or "rheumatism."

On examination there was marked engorgement of the neck veins; they did not pulsate and were not thrombosed. There was no clinical evidence of cardiac enlargement and no murmurs were audible. The pulse rate was regular and blood pressure (B.P.) 130/80 mm. Hg. The respiratory system was normal and there was no evidence of aortic aneurysm or thyroid enlargement. A tentative diagnosis of superior vena caval obstruction, cause undetermined, was made.

The patient was admitted to hospital for further investigation. There was radiological evidence of superior vena caval dilatation. The lungs were clear and there was no retrosternal mass. There was no obstruction in or distortion of the barium-filled oesophagus. Laryngoscopy was normal. The patient was discharged from hospital in December, 1940, but was seen again one month later when she complained of a "sickening" pain, which had been present for two weeks and was situated substernally but also in the right scapula. Lassitude was marked, and cough had been troublesome for a month. On examination her clinical condition was unchanged although the engorgement of neck veins was not so pronounced as previously. One month later the pain had lessened but swelling of the ankles and legs had developed during the preceding two weeks. She was treated with intravenous mercurial diuretics and responded with a moderate diuresis, but despite this she became progressively more dyspnoeic, cyanosed, and oedematous, with persistent neck vein engorgement.

She was readmitted to hospital in March, 1941, complaining of a "continuous grinding, aching pain" localized below the right scapula. Examination showed definite cyanosis with swelling of the face. She was slightly dyspnoeic at rest. There was marked neck vein engorgement up to the angle of the jaw. There were no cardiac murmurs audible; the pulse rate was regular at 70 per minute and the B.P. was 130/80 mm. Hg. There was slight oedema of the feet and forearms. There was dullness to percussion with moist crepitations at the lung bases. Radiologically there was slight enlargement of the heart and dilatation of the superior vena cava with basal pulmonary congestion and a small left pleural effusion. An electrocardiogram (E.C.G.) showed low-voltage curves with a biphasic T_3 and a normal electrical axis. The patient was treated with regular intravenous mercurial diuretics and oral digitalis, and responded satisfactorily. She was discharged from hospital much improved on June 15, 1941, and continued in moderately good health during the next few months.

In October, 1941, she complained of a recurrence of pain in the back, which gradually increased in severity until she was readmitted on Jan. 16, 1942. Physical examination did not reveal any essential change from the previous clinical picture of cyanosis, dyspnoea, and bilateral jugular vein engorgement with no abnormal cardiac findings. The radiograph of the chest was unchanged. There was a slight polycythaemia of 6,450,000 red cells per c.mm. of blood and 106 per cent haemoglobin. An E.C.G. revealed right axis deviation with a tall upright P_2 and P_3. There was S-T sag in leads 2, 3, and 4 (digitalis effect)

PLATE I.—(*a*) Cardiograms. (*b*) Photograph of the heart containing the tumour. The greater part of the heart, seen from this angle, consists of the right auricle, which has been laid open; the tumour is the bulbous mass occupying the whole of the left upper quadrant of the Photograph.

(Plate Ia). Within a month her pain had disappeared, and she was again ambulant.

She was discharged from hospital on Feb. 18, 1942, with instructions to report within two months. However, she failed to do so and was not seen again until her final readmission to hospital on Oct. 8, 1942.

She gave a history of increasing breathlessness on exertion for the previous two months, accompanied by swelling of the ankles, legs, and abdomen. On examination she was markedly cyanosed, with gross non-pulsatile engorgement of the neck veins. The B.P. was 110/80 mm. Hg in both arms. The pulse rate was 80 per minute and the rhythm regular. There was a small left-sided pleural effusion and bilateral basal crepitations. The liver was just palpable and tender, and there was a moderate degree of ascites, with oedema of the feet, ankles, and sacrum. The urine contained no abnormal constituents. The circulation time (arm to tongue, using sodium dehydrocholate) was 47 seconds in the right and 44 seconds in the left arm.

In November, 1942, the patient's condition was practically unchanged, and she still had signs of congestive cardiac failure, despite adequate digitalization and intravenous mercurial diuretics. Her general condition was slowly deteriorating, and the most probable diagnosis appeared to be constrictive pericarditis with marked cardiac decompensation chiefly affecting the right heart. A further E.C.G., taken in November, 1942, was consistent with this clinical diagnosis, having a low voltage, large upright P waves in leads 2, 3, and 4, small QRS complexes of normal duration, flat $T_{1,2}$, and inverted T_4.

On Nov. 23, 1942, she experienced a sudden attack of acute dyspnoea, but recovered rapidly after administration of oxygen. On Dec. 6, 1942, she had a second attack of sudden acute dyspnoea, accompanied by localized pain across the lower sternum, which lasted for about ten minutes and was relieved by opiates and oxygen. An E.C.G. was taken during this attack but did not show any changes suggestive of myocardial ischaemia. Subsequently her condition showed further slow but steady deterioration, and it was decided to carry out an exploratory thoracotomy.

This was performed by Mr. N. R. Barrett on Feb. 7, 1943. The mediastinum was exposed through a left-sided anterior intercostal incision: this gave good access to the front of the left ventricle and to part of the front of the right ventricle, but the rest of the heart was not explored. When the pericardium was opened a little free fluid was found in the pericardial sac but the pericardium itself was normal and was not thickened. Nothing was found at operation to account for the patient's condition She made a fair immediate recovery from operation, but subsequently took a gradual downhill course, and she died on March 13, 1943.

Post mortem.—A post mortem examination was carried out and the following were the principal findings. There were numerous mediastinal adhesions. The left pleural cavity contained a small amount of straw-coloured fluid, while the right lung was very adherent to the chest wall. The parietal pericardium was adherent, but was not considered to be restrictive. When the heart was opened the left auricle and left ventricle were found to be normal. The right ventricle was normal. The right auricle was large and contained a mass, the size of an orange, arising from the interauricular septum by a very short pedicle. This mass was endothelialized and lobulated and, on cut section, appeared white and myxomatous. It was macroscopically a true myxoma of the heart arising from the septal wall (Plate Ib).

Discussion

The clinical features and pathology of cardiac myxomas have been fully discussed and the outstanding literature reviewed by Yater (1931) and Fawcett and Ward (1939).

In this case the tumour arose from the right interauricular septal wall. Chiari (quoted by Gorlitzer, 1934) described a case of myxoma of the right auricle, but in other reported cases the myxomas were either present in the left auricle (Yater, 1931; Gorlitzer, 1934; Jensen, 1934; Lymburner, 1934; Bien and Ch'in, 1936; Gilchrist and Millar, 1936; Bennett and others, 1938; Fawcett and Ward, 1939; Hamilton-Paterson and Castleden, 1942; Thompson, 1944; Burnett and Davidson, 1945) or on the heart valves (Abrahamer, 1931; Jaleski, 1934).

As the site of the tumour determines the clinical picture in each case, a myxoma of the left auricle presents a picture of mitral stenosis with congestive cardiac failure (Jensen, 1934; Bien and Ch'in, 1936; Hamilton-Paterson and Castleden, 1942; Thompson, 1944; Burnett and Davidson, 1945) although the classical murmurs of mitral stenosis may be variable or absent (Lymburner, 1934; Bennett and others, 1938; Fawcett and Ward, 1939). The myxoma of the right auricle in this case presented the clinical picture of chronic superior vena caval obstruction with no evidence of organic cardiac disease, as shown by abnormal cardiac murmurs or arrhythmias.

In the majority of reported myxomas of the left auricle, the duration of the illness from onset of symptoms to death has usually been short, and sudden death has been the rule (Gilchrist and Millar, 1936; Bennett and others, 1938; Fawcett and Ward, 1939). This course is dependent upon the size of the tumour and the length of the pedicle. If the pedicle is of sufficient length to permit free mobility of the myxoma, sudden death may occur as a result of complete obstruction of the mitral orifice (Houck and Bennett, 1930; Gorlitzer, 1934; Fawcett and Ward, 1939) or acute pulmonary oedema may develop because of partial

obstruction of the pulmonary veins (Gilchrist and Millar, 1936; Thompson, 1944). In this case, breathlessness on exertion was first experienced in 1939, and the patient died in 1943. Although the tumour was large, almost completely filling the auricular cavity, the pedicle was extremely short and complete obstruction of the tricuspid orifice or great veins was not possible. Partial obstruction of the tricuspid orifice may have accounted for the two attacks of acute dyspnoea, and it is interesting that these attacks occurred when the patient was leaning forward and were relieved when she fell back in bed.

Relentless and rapid progress of cardiac failure despite adequate rest and digitalization has been cited as one of the diagnostic criteria of intracardiac tumour (Yater, 1931; Fawcett and Ward, 1939). In this case, although response to rest, digitalis, and mercurial diuretics was not entirely satisfactory, the progress of the illness was fluctuant over a period of twenty-nine months, and the patient was sufficiently improved on three separate occasions to warrant her discharge from hospital.

An abnormal radiological outline due to the presence of a large tumour of the left auricle has been described. In Bennett and others' case a large bulge in the region of the pulmonary conus was noted, while Gilchrist and Millar observed a projection into the posterior mediastinum associated with marked enlargement of the heart to the right and left. In this case, the only abnormal radiological findings were the persistently widened superior vena caval shadow and a moderate uniform cardiac enlargement.

Electrocardiograms have been recorded in only a few cases of primary cardiac tumour, and arrhythmias which have been noted usually occurred when the tumours were malignant. Bundle-branch block was reported by Shelburne (1935) in a case of sarcoma of the left auricle and left ventricle, which was correctly diagnosed during life. Innocent tumours do not interfere with the conducting mechanism of the heart. Gilchrist and Millar recorded paroxysmal auricular tachycardia in a case of myxoma of the left auricle, and Fawcett and Ward reported an attack of paroxysmal auricular fibrillation just before death. No other instances of cardiac irregularities have been reported. In Burnett and Davidson's case of a myxoma of the left auricle, the E.C.G. showed right axis deviation with normal P waves, low-voltage T_1 and T_2 and slight depression of S-T_3, while in Thompson's case there was a normal electrical axis with general low voltage.

In this case, the electrocardiographic pattern was very similar to that found in acute cor pulmonale with tall "right auricular" P waves, a deep S_1, flat T_2, inverted T_3, and small Q_3.

The pathology of these tumours has been the subject of much debate and disagreement. The point at issue has been whether they are in fact true myxomas or merely organized thrombi. Most authorities are in agreement with Ribbert (1924), who maintains that they are true tumours arising from myxomatous rests in the interauricular septum (Ewing, 1928; Yater, 1931; Abrahamer, 1931; Bien and Ch'in, 1936; Fawcett and Ward, 1939). The evidence in support of this view has been summarized by Fawcett and Ward, who lay emphasis on the following facts. Myxomas are always found in the auricles or on the heart valves, whereas thrombi are more commonly found in the ventricles. They arise from a site in the interauricular septum where it is known that myxomatous rests may occur. In the majority of cases there is no past history of endocarditis, which would tend to encourage stasis of the blood and thrombus formation. Microscopically these tumours demonstrate no lamination and few cells—which are characteristically spindle or star-shaped. A thrombus will tend to retract and show scarring, whereas myxomas are smooth, villous, papillomatous, or polypoid—the picture of a true neoplasm—and they steadily increase in size.

Husten (1922) contended that the majority of these tumours were organized thrombi, and of seventy-one reported cases he maintained that only nine were true myxomas. In his view of the pathology of the condition he is supported by Hamilton-Paterson and Castleden, who named them "pseudo-myxomas," and based their belief on the grounds that organizing thrombi could show all the features found in myxomas.

The cardiac tumour described appeared to be a true myxoma as it did not in the least resemble a thrombus.

Conclusion

A case of a myxoma of the right auricle occurring in a woman of 64 years is described. She was observed in chronic cardiac failure for twenty-nine months. The essential clinical features were marked cyanosis, superior vena caval obstruction, peripheral oedema, and two abrupt attacks of acute dyspnoea. The electrocardiogram showed an acute cor pulmonale pattern. The diagnosis of constrictive pericarditis was entertained and proved at operation to be incorrect. At necropsy a myxoma arising from the right side of the interauricular septum was found.

The condition is very difficult to diagnose during life, and, probably because of its rarity, the correct diagnosis was not even suggested in this case. In retrospect, the diagnostic clues were prolonged right-sided heart failure without evidence of pulmonary or cardiac valvular disease, and an electrocardiogram with tall "right auricular" P waves. Fluoroscopy did not help, but it is probable that angiocardiography would have provided essential information.

It is possible that in the future such a tumour might be removed surgically, provided the diagnosis could be made in the early stages of the illness.

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