Myxoma Cordis

Diagnosis Established Pre-operatively;
Surgical Removal of the Tumour

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Myxoma cordis is a rare disorder. Up to 1951, 128 cases were known in the literature (Mahaim, Prichart). It has long been a subject of controversy whether these formations were to be regarded as organized thrombi or as tumours, but now they are considered as tumours. The myxomas are the most frequently occurring primary tumours of the heart, viz., about 50 per cent. In 50 per cent of cases they are polypous neoplasms of a partly gelatinous, partly rubber-like consistency, as a rule with a pedicle, arising from the auricular septum close to the foramen ovale. In 75 per cent they are situated in the left atrium. They have been observed in all age-groups in both sexes, but mainly between the 30th and 60th year of life. The size may vary from a pea to that of a closed fist. The small tumours cause no symptom.

Before dealing with clinical symptoms of the larger-sized tumours, we will first report the case-history of a patient examined by us.

J. K. R., male, age 45. One year previous to the time of writing he suffered from palpitations and an oppressed feeling in the chest when cycling against the wind. These symptoms disappeared as soon as he rested. The complaints increased gradually, especially in the last eight months. When he had been in a stooping position and then straightened himself, he became dizzy and felt palpitations, lasting for some hours. During the last six months he became short-winded if he had only walked about 25 metres, or on slight exertion. Stooping made him short of breath, which improved as soon as he stood up again. He was also unable to sleep on his right side as he used to do, for then again he became dyspnœic. He was however able to sleep well when lying on his back or on the left side, but his head had to be elevated. Of late coughing had become worse, with sensations of vertigo. During the last year he had repeatedly suffered from attacks of bronchitis, sometimes also from fever. His appetite was good, micturition and defaecation were normal. He had not had swollen feet. In his youth he had often suffered from bronchitis with attacks of wheezing, lasting for about one week. There was no history of rheumatic affections.

He was a normally built man, in good nutritional condition, without cyanosis or oedema. Pulse: regular, equal, rate 88/min. Respiration: 18/min. Blood pressure: 130/90. The venous pressure was not increased.

The apex beat was not palpable. Percussion showed the heart to extend from the midclavicular-line to the mid-line. The first sound at the apex was not loud, but split; the second pulmonary sound was accentuated. There was no murmur, in the supine or in the lateral position. Percussion was normal over the lungs; the pulmonary boundaries did not show large excursions; expiration was prolonged, with wheezing rales.

Liver and spleen were not palpable. Urine: albumen negative; glucose negative; urobilin trace; sedimentation normal, Haemoglobin content 80 per cent; erythrocytes 4,400,000; leucocytes 8,100; eosinophils 3 per cent, polynuclears 88 per cent, lymphocytes 24 per cent, monoocytes 5 per cent. Erythrocyte sedimentation rate 19 mm. after one hour.

X-ray film of the thorax (Fig. 1): The heart was not enlarged, marked hilar shadows radiating towards the periphery, no hilar pulsations. In the right oblique position an impression of the oesophagus near the left atrium was visible (Fig. 2). On fluoroscopy the extremely strong pulsatory movements of the oesophagus at the

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Figure 1: Posteroanterior x-ray (J. K. R.).—Figure 2: Right oblique position; slight impression on esophagus.—Figure 3: Esophagram of esophagus.
level of the left auriculum formed a striking feature. The kymogram revealed (Fig. 3) that these pulsations coincided with ventricular systole. E.C.G. (Fig. 4) : sinus rhythm PR 0.2 seconds, QRS 0.08 seconds, high P-peaks in II, III, AVF, negative P in V₁, diphasic P in V₂ and V₃, right hypertrophy (QR in AVR, negative T in III, V₁-V₆, R₁ in V₅ and V₆). Circulation time 13 seconds (magnesium sulphate). Vital capacity 3,225 ml. (normal value 4,110 ml.), residual air 36 per cent, functional residual air 58.5 per cent.

On cardiac catheterization the following values were found:

<table>
<thead>
<tr>
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<th>O₂-Saturation</th>
<th>Pressure</th>
<th>Mean Pressure</th>
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<tbody>
<tr>
<td>Right auricle</td>
<td>54</td>
<td>7</td>
<td>mm. Hg</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>53.5</td>
<td>120/0</td>
<td>mm. Hg</td>
</tr>
<tr>
<td>Pulmonary artery trunk</td>
<td>49</td>
<td>120/50</td>
<td>mm. Hg</td>
</tr>
<tr>
<td>Pulmonary artery periphery</td>
<td>98.5</td>
<td>40</td>
<td>mm. Hg</td>
</tr>
<tr>
<td>Radial artery</td>
<td>96.5</td>
<td>130/90</td>
<td>mm. Hg</td>
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The curve obtained from the periphery of the pulmonary artery (P.C.P. curve) showed a remarkably early start of the V-wave which was also extremely high (Fig. 5), and a missing X-dip, consequently a positive venous pulse.

The complete picture was highly reminiscent of mitral stenosis without murmurs with considerable pulmonary congestion. The high pressure in the pulmonary artery and right ventricle could likewise be explained in this way, but murmurs were never heard, and in the E.C.G. the right strain was indeed explicable. Angiocardiography, however, led to the solution.

This technique (Figs. 6a and 6b) revealed a slow passage of the contrast fluid throughout the pulmonary circulation (after eight seconds still in the pulmonary artery, after 12-14 seconds a well-filled left auricle was observed). A gap was visible in the shadow of the left auricle. This made a tumor in the left auricle plausible, a myxoma being especially suggested. The history and course of the disease also provided arguments in favour of this (see below). An operation was therefore considered. Due to the necessary preparations, this was only possible after two months.

The condition gradually deteriorated during the patient’s stay in hospital. He again suffered from fever of unknown cause for some time. Blood cultures were negative the antistreptolysin-titre was not increased (100 U./ml.). Temperature did not fall after penicillin administration, but became normal on administration of 1.5 g. pyramidon daily. Subsequently, symptoms of right decompensation developed (namely, severe oedema, increased venous pressure). One day after changing beds, he suddenly became dyspnoeic and finally comatose, but recovered the next day.

He was treated with digitalis and diamox, to which he responded favourably. The oedema disappeared and he was no longer dyspnoeic. Then it was decided that the risk of an operation was justified, even if the prognosis was not favourable in view of the period of decompensation.

Discussion

The case-history of our patient corresponds with the observations in several similar cases. Dyspnoea of effort is the prominent feature, gradually increasing and due to pulmonary congestion. Later signs of right decompensation arise. Since in some cases murmurs are heard at the apex (also pre-systolic murmur), it is understandable that the diagnosis of mitral stenosis has often been made.⁶ ¹⁰ ¹³ ¹⁷ Yet both the history and the course of the disease show peculiarities that as a rule are not encountered in a case of mitral stenosis.

In the first place the symptoms caused by changes in body position. As was demonstrated in our patient, acute dyspnoea and pre-cardial pain may suddenly occur after movement; even shock or comatose condition may
develop. Changes in position may give rise to sudden dizziness. Our patient, who had always slept on his right side, was no longer able to do so; sleeping was possible on his left side or back, but with the head elevated. The explanation of these symptoms is related with the mobility of the pedicled tumour in the left atrium, which may cause sudden serious circulatory disturbances. There is also a relationship with the variations in the physical signs that have often been described, e.g., a systolic murmur may be present in the lying position and a diastolic murmur in the sitting position, as already pointed out by Pawlowski (cf. Gottel). In our patient no murmurs were audible in the various positions.

The rapid progression of the decompensation symptoms is striking. It reacts poorly to treatment with cardiotonics\textsuperscript{11, 19} which is unusual in mitral stenosis cases. Kirkey and Leren established the diagnosis ante mortem on the strength of variation of murmurs in different positions of the patient, and the rapid progression of the decompensation.

A remarkable observation in our patient was the strong pulsatory movements of the oesophagus at the level of the left auricle, synchronously with the ventricular action (Fig. 3); in our opinion, this fact is of diagnostic significance. This symptom is also absent in mitral stenosis. The systolic-positive excursion of the peripheral pulmonary curve without signs of mitral insufficiency can be explained by the mechanically ineffective auricular diastole, which is also of diagnostic significance. Finally, for the differential diagnosis it may be of interest that patients have no rheumatic fever in their history, but this also holds true in many cases of mitral stenosis. Moreover, some with myxoma had previously suffered from rheumatic affections\textsuperscript{15}.

In other cases of myxoma cordis the emboli in the systemic circulation (brain, lungs, kidneys, extremities and in the coronary arteries) are of great importance\textsuperscript{5, 11, 19} sometimes causing death. These emboli, combined with cardiac murmurs and occasionally also with fever, were in several cases reminiscent of bacterial endocarditis,\textsuperscript{5} but the blood cultures were always negative. In many cases the systolic blood pressure was low (less than 110 mm., even as low as 65 mm.),\textsuperscript{17} but this sign was often lacking. Cardiac catheterization in our patient revealed high pressure values in the right ventricle and pulmonary artery; but this may be explained by pulmonary congestion and may also be found in mitral stenosis. The P.C.P. curve showed the picture known in mitral insufficiency and more especially if auricular fibrillation is present.

![FIGURE 4: Electrocardiogram, right ventricular strain](image-url)
Although history and changing physical signs provide important indications, it has been the angiocardiography which during the past few years has sometimes led to the correct diagnosis (left auricle),\textsuperscript{11} (right auricle).\textsuperscript{1, 3, 15}

In the contrast shadow of the auricle a gap is visible at the site of the tumour, if this is not too small. It is quite manifest in the second oblique

FIGURE 5: Pressure curves, pulmonary-artery periphery, pulmonary-artery trunk, and right ventricle.
position (left Fig. 6). In adults one should make exposures of 12-14 seconds, since the filling of the left auricle is retarded by the pulmonary congestion. This enabled us also to establish the diagnosis.

In the differential diagnosis malignant tumours of the heart should be mentioned, which are usually sarcomas and occur less frequently than benign ones. Sarcomas are generally situated in the right heart (auricle as well as ventricle). They show a more infiltrative growth, but may also be found as polypous tumours in the cavities of the heart. Obstruction of the superior vena cava and haemorrhagic pericarditis often occur in conjunction with these tumours observed by us twice. Metastatic heart tumours occur far more frequently (incidence 20-40 times as high\textsuperscript{16}). They have already been diagnosed more often, also during life, based on occurrence of arrhythmias and haemorrhagic pericarditis in the presence of metastasizing tumour. Metastases may have originated from carcinomas of all major organs, especially of mammary glands, lungs, stomach, from skin melanomas and from lymphosarcomas. They may be situated in the right as well as in the left heart and often in both at the same time. They are rarely located in the valves.

Treatment

As operative removal of intracardiac tumours has so far been carried out in only a few cases, the results being still disappointing, it is desirable that new experiences be published. Only benign cardiac tumours can be considered for surgical removal. In the majority of cases these are the so-called myxomas or fibromyxomas, often pedicled, occurring in 25 per cent in the right atrium and in 75 per cent in the left atrium. They usually ori-
ginate from the septal wall. It further depends on the localization and size whether a benign tumour or cyst can be excised from the cardiac wall, as reported by Beck. Up to the time of writing, six cases of pre-operatively diagnosed myxoma cordis are known.1, 2, 11, 18

When operative treatment of the patient described was decided upon, we first considered which method of operation would be the best in this particular case. Study of the literature yielded the following facts:

Goldberg et al, reported two cases, one patient was operated on and died. Bahnson was no more successful with his patient. Ripstein operated on one patient under hypothermia, without result. Bailey operated on two, also with fatal issue, while Crafoord, in a personal communication to Goldberg, referred to a patient successfully operated in 1954, (July 16) with the aid of complete bypass with extracorporeal oxygenation and circulation.

On Bailey’s advice we decided to operate under hypothermia, and were well aware of the fact that the chances of success were only slight, in view of the patient’s age (45 years), serious pulmonary hypertension and the attack of decompensation.

Cardiac fibrillation was the imminent danger. The anaesthetists, therefore, were to try and keep the pH on the alkaline side during the ether-oxygen anaesthesia, by means of hyperventilation, because this consider-

FIGURE 7: Tourniquets applied to the auricle and atrium and incisions used.
ably reduces the chances of ventricular fibrillation during operations under hypothermia.

We chose the immersion cooling technique as developed by Swan in Denver. During the operation, we were to be kept informed about the changes occurring during cooling by means of the ECG and regular determinations of the pH and alkali reserve (Brinkman et al.) and potassium determinations (Groen et al.). The patient was connected with an electrocardiograph, and transferred to the bath filled with water of 37°C. under intratracheal ether-oxygen anaesthesia after premedication with 15 mg. morphine, 0.25 mg., atropine and 100 mg. pracatal. When this was tolerated well, ice cubes were added for further cooling, and recording of the temperature and continuous ECG registration were started. The first blood sample (8:45 a.m.), taken while the patient was already in the bath and the temperature had fallen only 1°, yielded the following data: pH = 7.61 (18°C) = 7.41 (37°C). Alkali reserve = 31.4 vol. per cent. Plasma potassium 3.45 m.Eq./l. Arterial oxygen saturation 100 per cent.

Suddenly, when the patient’s temperature had fallen not more than 1°, the ECG failed completely and the patient developed a pale colour, so that, evidently, acute cardiac arrest had set in. Blocking of the mitral ostium by

FIGURE 8: Myxoma extirpated from the left atrium (6.5 x 4 x 5 cm.).
the tumour had probably been an important cause in this event.

Taking a retrospective view, the severe acidosis (31.4 vol. per cent) should, of course, have made us hesitate to continue the operation, but we did not know this until the patient was already in the bath, and the pH was still normal due to the marked hyperventilation.

In operations under hypothermia it is tried to keep the pH on the alkaline side. From the beginning of the operation, the lungs are always markedly hyperventilated, which, in our patient, became manifest in the low carbon dioxide values recorded by the carbovisor (Brinkman) and the high oxygen saturation of the blood. We failed to realize sufficiently, however, that before the operation, in the medical clinic, mercurial diuretics and diamox had been used in the treatment of oedemata resulting from the cardiac decompensation, and that the acidosis was promoted by inhibiting the catalytic action of carbon-dioxide anhydrase (renal acidosis accompanied by hypopotassaemia). We had perhaps done better not to place the patient flat on his back in the bath, because, this probably promoted blocking of the mitral ostium. The patient was taken out of the bath after the occurrence of the asystoly, and placed on the operating table in the right lateral position. Unfortunately, some precious minutes were lost during this procedure. The thorax (there was left hydrothorax) was opened, also the pericardium (there was hydropericardium), and cardiac massage started at once. We succeeded in restoring the rhythm after some minutes. The blood pressure rose to 85 mm. Hg., his colour became good and it seemed that the acute danger had passed for the time being. A blood sample, obtained by puncture from the left atrium, yielded the following

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**FIGURE 9A**

*Figure 9A: Photomicrograph of the myxoma.*

**FIGURE 9B**

*Figure 9B: None.*
particulars: pH = 7.39 (18°) = 7.19 (37°), alkali reserve: 26 vol. per cent (normal 54-55 vol. per cent), plasma potassium 2.78 m.Eq./l), arterial oxygen saturation: 96 per cent.

This means that now there existed marked acidosis, no longer to be compensated by hyperventilation. There was also serious hypopotassaemia. We were advised to inject intravenously 400 ml. 6 per cent NaHCO₃ and 10 ml. 1 per cent KCl. Due to difficulties with the intravenous drip, this dose was not taken up completely.

After some deliberation it was decided to continue the operation, because stopping would involve too great risks due to the imminent blocking of the mitral ostium by the tumour.

A tobacco bag suture was laid around the base of the left auricle, and taken in a Rümmel tourniquet. Another tobacco bag suture in a Rümmel tourniquet was applied to a sound part of the left atrium where the incision was to be made (Fig. 7.). The auricle was opened and the atrium was palpated with the bare right index finger. A polypous short-pedicled tumour was found, lying with its lower pole on the mitral ostium. The pedicle was easily detached from the septum, the tumour was kept floating and pressed against the site where the left atrium was to be incised. The latter procedure was performed with the left hand, and the tumour was pushed through the opening without loss of blood, following which the tourniquet was tightened.

The course of the operation was excellent. The atrial wound was closed rapidly, as was the left auricle. We refrained from placing a clamp on the base of the aorta and pulmonary artery, because the operation proper seemed to become a successful and rapid procedure. Initially the heartbeat was good; it was slow and regular, but suddenly ventricular fibrillation arose. Electric defibrillation (0.1 sec., 240 volt, 1.5 amp.) brought the heart to a standstill, following which cardiac massage was again applied. We succeeded in getting the heart beating again, in which it struck us that the myocardium was rather lax. In the meantime the patient had been injected noradrenaline via the infusion (3 mg.). This procedure was repeated when ventricular fibrillation occurred anew. Success was again obtained, but it was of shorter duration. The blood pressure was finally no longer to be measured, and death followed after 10 ml. CaCl₂ had been injected intravenicularly in vain.

Looking backwards, we should not have started the hypothermia before the pH. and alkali reserve had been determined once more on the morning of the operation. If we had known of the acidosis and hypopotassaemia, we could have tried to combat the acidosis by administration of NaHCO₃, in which, of course, it is questionable whether thus the fatal issue could have been prevented with certainty. Probably the risk would have remained great indeed.

Taking everything into account, we were well aware that the patient would be exposed to great risks during an operation, but his future was also hopeless with conservative treatment. In the future, if the diagnosis
can be established early, and if the tumour exerts a less serious influence on heart and circulation, the issue will probably be more satisfactory in such cases.

Based on the pathological-anatomical findings, and in view of the fact that these “polypous” tumours are so often pedicled, operative treatment was already suggested earlier. If the operative removal is to be successful, however, it will be necessary, that the patient be operated on before decompensation had advanced too far. The operative technique in itself was satisfactory.

At necropsy of our patient (VOS) marked hypertrophy of the right heart with pulmonary congestion was found. There was no sign of air embolism. The site of the myxoma was only indicated, and was located near the foramen ovale. Marked hydrothorax and hydropericardium had already been observed during the operation. The description of the operative specimen (VOS) is as follows:

Macroscopeally (Fig. 2). The tumour, extirpated from the left atrium of patient J. K. R., where it had been attached to the septum atriorium with a broad pedicle, measured 6.5 x 4 x 5 cm. The surface was smooth, shining, partly yellowish-grey and partly light reddish-brown. The tumour was soft and elastic, and proved to consist of fine, light yellowish-grey stripes on the cut surface.

Microscopically (Fig. 9). The greater part of the tumour tissue was composed of markedly oedematosly swollen connective tissue, poor in cells and fibres, containing at some places narrow and scattered, and at other sites wide and closely packed parallel running connective tissue bundles with many capillaries. Some of these cell-rich parts were surrounded by areas poor in cells, somewhat reminiscent of mucinous tissue, but which did not give a mucin reaction, however, as is usual in such tumours.

In the present case no reason was found to doubt the blastomatous character, and to take this affection—as done by some investigators—for an organized thrombus.

Anatomical diagnosis: Myxoma cordis septi interatriati cordis in atrio sinistro.

SUMMARY

A case of pedicled myxoma cordis, originating from the septum of the left atrium in a 45-year-old man, in whom the diagnosis had been established pre-operatively is recorded. Operation was decided on because the patient had already suffered a severe attack of cardiac decompensation. The tumour, measured 6.5 x 4 x 5 cm., was removed under hypothermia. After the temperature had fallen 1° (immersion cooling) cardiac arrest arose, most probably due to obstruction of the mitral ostium by the lower pole of the tumour. Cardiac rhythm was restored by means of heart massage, and the operation was terminated transatrially without any loss of blood or air embolism. Unfortunately, ventricular fibrillation arose shortly afterwards; although this was overcome in the beginning, death was unavoidable due to repeated ventricular fibrillation.
RESUMEN

Se relata un caso de mixoma cardíaco pediculado originado en el septum atrial izquierdo en un hombre de 45 años en quien el diagnóstico se hizo antes de la operación. La operación se llevó a cabo porque el enfermo había sufrido ya un ataque severo de descompensación cardíaca. El tumor media 6.5 x 4 x 5 cms. y fue extraído bajo hipotermia. Después de que la temperatura había bajado 1° (enfriamiento por inmersión) hubo paro cardíaco, lo más probable debido a obstrucción del orificio mitral por el polo inferior del tumor. Se recuperó el ritmo cardíaco por el masaje y la operación se terminó por vía transatrial sin pérdida alguna de sangre y sin embolia gaseosa.

Desgraciadamente se presentó la fibrilación ventricular poco después y aunque ésta fue dominada al principio, la muerte se presentó inevitablemente debida a la fibrilación ventricular reiterada.

RESUME

Les auteurs rapportent un cas de myxome cardiaque pédisul provenant du septum de l'oreillette gauche, chez un homme âgé de 45 ans, chez lequel le diagnostic a été établi avant l'intervention. Celle-ci fut décidée parce que le malade avait déjà subi une attaque sérieuse de décompensation cardiaque. La tumeur, mesurant 6,5 x 4 x 5 cm. fut enlevée sous hypothermie. Après que la température eut tombée de 1° (refroidissement par immersion) un arrêt cardiaque survint, imputable le plus vraisemblablement à l'obstruction de l'orifice mitral par le pôle inférieur de la tumeur. Le rythme cardiaque fut rétabli grâce au massage du cœur et l'opération se termina à travers l'oreillette sans aucune perte de sang ou embolie gazeuse. Malheureusement, une fibrillation ventriculaire survint peu de temps après, et bien qu'elle ait été surmontée au début, sa répétition entraîna la mort.

ZUSAMMENFASSUNG

Wiedergabe eines Falles eines gestielten Myxoms des Herzens, ausgehend vom Septum des linken Vorhofes bei einem 45 Jahre alten Mann, bei dem die Diagnose vor der Operation gestellt worden war. Zu der Operation hatte man sich entschieden, weil der Patient bereits einen schweren Anfall cardialer Dekompensation erlitten hatte. Der Tumor in den Ausmassen von 6,5 x 4 x 5 cm. wurde unter Hypothermie entfernt. Nachdem die Temperatur 1 Grad gefallen war, (Kühlung mit Immersion), kam es zu einem Herzstillstand, höchstwahrscheinlich infolge Verstopfung der Mitralöffnung durch den unteren Pol des Tumors. Die Herztätigkeit kam wieder zustande mittels Herzmassage, und die Operation wurde zu Ende geführt vom Vorhof aus ohne irgend welchen Blutverlust oder Luftembolie.

Unglücklicherweise trat kurz danach Kammerflimmern auf; obwohl dies sich zu Anfang überwinden, liess, war der Tod unvermeidlich infolge wiederholten Kammerflimmern.
REFERENCES


