Myxoma of the Heart
Roentgen Diagnosis During Life in Three Cases*

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Heretofore, the presence of myxoma of the heart could only be suspected during life. Angiocardiography in three cases, one early in life; one in the fifth; and the other in the sixth decade revealed characteristic left atrial filling defects. Subsequent surgical exploration with removal of fragments of the tumor in two instances confirmed the diagnosis of myxoma. Two patients failed to survive operation and autopsy examination established the validity of the angiocardiographic findings. The fact that cardiac myxomas are benign lesions often attached to the interatrial septum by a stalk makes their surgical removal a practical possibility. Furthermore, advances in surgical techniques such as the atrial well1 and the development of extracorporeal mechanical pumps2 are expected to make surgery curative.

Case Reports

Case 1: A 53 year old white woman (N.Y.H. No. 576014) was referred from another hospital on August 15, 1950 for angiocardiography because of the finding of a right hilar mass. Five months earlier, she had had a sudden onset of crushing left chest pain radiating to the back and associated with weakness, palpitation and dyspnea. She remained in the hospital for two months with little improvement. Serial electrocardiograms showed normal sinus rhythm and a normal electrocardiographic pattern. Two months later, she was readmitted because of recurrence of sudden crushing substernal pain. No heart murmurs or electrocardiographic abnormality were detected but a right hilar mass was seen in the chest roentgenogram.

Her symptoms began at the age of 26 years with aching in neck and calf muscles. Joint pains, tonsillitis and chorea were denied. The next year, dyspnea, generalized weakness and palpitation on exertion began. The dyspnea gradually increased until six years ago when she could only climb two stairs. Constricting precordial pain radiating to both sides of the neck started five years ago and became associated with palpitation and marked weakness. Rest, particularly when lying on the right side relieved the chest pain but not the palpitation. She was in a German concentration camp for four years during World War II and managed to get to the United States in 1949. The effects of her war experiences left her markedly depressed.

On admission she was well developed and nourished although depressed. The heart rate was 90 with normal sinus rhythm and the blood pressure was 120/80. There were no cardiac murmurs and the electrocardiogram showed only left axis deviation. Chest x-ray film (Figure 1) revealed generalized heart enlargement, prominence of the pulmonary artery segment and left atrial enlargement. Angio-

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cardiography (Figure 2) showed a constant filling defect in the left atrium. This was considered evidence of an interatrial tumor. She was not improved after a month's stay in the hospital.

Weakness, palpitation and dyspnea continued and became worse. On a visit to the clinic, October 26, 1951, she was discovered to have a totally irregular pulse with a rate of 140 to 150 per minute. Electrocardiography revealed rapid auricular fibrillation with impure flutter. Digitalis was prescribed and after a night's observation she was sent home. After a brief episode of digitalis toxicity, digitalis leaf was substituted and on a daily maintenance dose she has satisfactory slowing of the heart rate. Cardiac catheterization performed with Dr. Daniel S. Lukas on November 2, 1951 revealed pulmonary arterial and pulmonary capillary (wedge) hypertension, and a low cardiac output. After one month's hospital stay, although her heart rate was slowed, she was unimproved.

For the next 15 months she was seen regularly in the out-patient department but was found to be getting worse. Finally, on February 12, 1953 she was readmitted for surgical excision of the left atrial tumor. A short, grade two, systolic murmur was heard at the apex. The heart rhythm was irregular, the ventricular rate was 96, and the blood pressure 90/60. She appeared pale, and the hemoglobin was 10 grams. Two transfusions (250 cc. each) of packed red blood cells were given. There was no enlargement of the liver or leg edema. Repeat angiocardiology (Figure 2) on February 13, 1953 showed an increase in size of the left atrium and of the intraatrial mass. On February 19, 1953, surgical exploration verified the presence of a left atrial tumor.

Post-mortem examination (N.Y.H. Autopsy No. 14947) revealed a multilocular, rubbery, dark brown gelatinous left atrial tumor 15 x 7 x 6 cm. in diameter and weighing 150 grams (Figure 3). The tumor cut with ease. The atrial wall was thickened at the origin of the tumor and there was a 3.5 cm. pedicle. Microscopically, the tumor appeared mostly cellular at its origin where there was thickened collagenous endothelial tissue with loose myxomatous tissue, abundant blood supply and mononuclear cells of various sizes and shapes. Clumps of cells with abundant acidophilic cytoplasm in syncytial masses all covered by a single thin layer of endothelium were also seen. The histologic diagnosis was myxoma.

**FIGURE 1A**

*Figure 1A: Intra-atrial myxoma, Case 1. Conventional chest roentgenograms, 1953. A. Postero-anterior view shows moderate generalized cardiac enlargement with a localized bulge of the right heart border just below the right pulmonary artery. B. Lateral esophogram reveals posterior enlargement of the left atrium, as in mitral stenosis.*
FIGURE 2: Intra-atrial myxoma, Case 1. Angiocardiograms.
A. Left heart, frontal projection, 1950. There is a lobulated filling defect centrally located in the enlarged left atrium.—B. Tracing of A.—C. Left heart, frontal projection, 1953. The left atrium and the tumor have increased in size.—D. Tracing of C.—E. Left heart, lateral projection, 1953. Tumor mass virtually fills the entire left atrial cavity in this as well as the frontal projection.—F. Tracing of E.
The heart weighed 490 grams. The right atrium, right ventricle and left atrium were dilated approximately one and a half times their normal size. The left ventricle wall measured 15 mm. in thickness; the right ventricle 3 mm. The coronary arteries were atheromatous but patent throughout. The cardiac valves were intact and free of vegetations. The aorta was generally atheromatous but no more than that of a normal person of 55 years.

Case 2 (Previously published in Circulation, 6:762, 1952): A white male child (N.Y.H. No. 591797) three years of age, was first admitted to the New York Hospital on February 6, 1951. The past history was unremarkable except for measles at two and one-half months and chicken pox at six months. The family history revealed rheumatic fever in the paternal grandmother and a paternal aunt. He had good health until March 17, 1950, when (at the age of two years and three months) he suddenly developed a right sided hemiparesis. The paralysis lasted for only 10 minutes, but weakness remained. He was admitted to another hospital.

FIGURE 3: Gross Pathological Specimen, Case 1.
The large myxoma is in situ in the enlarged left atrium.
where physical examination of the heart was unremarkable and laboratory studies did not clarify the cause of the hemiparesis. There was no change until the next month when he complained of the sudden onset of pain in his left foot. His mother noted "red spots" along the side of the foot. However, the pain and discoloration subsided in a week and at no time were severe enough to lead to consultation with a physician. A second episode of brief paralysis of the right arm and leg occurred suddenly on September 12, 1950. Residual weakness was more marked this time. A new finding was the presence of an apical systolic murmur. The third occurrence of paralysis of the right arm and leg was noted October 22, 1950, 12 hours following a reported injury to his head. He was hospitalized again and had an uneventful course. Residual weakness was still marked. During the two months of this hospitalization several important cardiac abnormalities were noted. An apical systolic murmur was almost constantly present. An apical presystolic rumbling murmur was heard well at certain times and poorly or not at all at others. Fluoroscopy disclosed an enlarged left atrium and right ventricle.

Physical examination on transfer to the New York Hospital showed a well-developed three year old boy with weakness of the right arm, leg and face. There were increased resistance to passive extension, hyperactive deep tendon reflexes, positive Babinski and finger-stretch reflexes. The heart was not enlarged to percussion. There was an apical presystolic thrill. The first sound at the apex was accentuated and preceded by a rumbling crescendo murmur. The second pulmonic sound was not accentuated. The blood pressure was 110/70. There was no venous distention and the peripheral pulses were normal. The remainder of the examination was unremarkable. Extensive laboratory studies showed no significant abnormality. Fluoroscopy and x-ray film again showed a large left atrium and right ventricle while the left ventricle and the pulmonary arteries were normal (Fig. 4).

The course in the hospital was unremarkable. Angiocardiography demonstrated a large, irregular, polypoid filling defect in the left atrium anteriorly, immediately above the mitral orifice (Figure 5). The left atrial appendage was uninvolved. These findings were interpreted as indicative of a tumor (probably a myxoma) within the left atrium. Surgical treatment was postponed in view of the great risk involved and with the hope that a satisfactory extracorporeal circulatory mechanism might become available soon.

The child was discharged from the hospital on March 2, 1951. Shortly afterwards there was a transient episode of swelling and redness of the left second toe. A
short course of x-ray therapy to the heart was given at another hospital during May of 1951 without any change in the cardiac findings. He was treated for bronchitis in April and pneumonia in May. During a visit to the New York Hospital Cardiac Clinic on July 25, 1951, he was noted to be in general good health. There was no change in the cardiac findings but fluoroscopy showed pulsatile dilatation of the pulmonary arteries.

On September 12, 1951, the child developed edema of the face, arms and legs and was admitted to the New York Hospital two days later. He was acutely ill with obvious dyspnea and orthopnea. The cervical veins were distended. A firm and tender liver edge was palpated 8 cm. below the costal margin. The lungs were clear; the respiratory rate was 36. The heart rate was 132 and the blood pressure 108/70. The heart was enlarged to the left. The other cardiac signs were unchanged. Edema was not noted. Venous pressure was 440 mm. saline and the fluorescein circulation time from arm to lips was 16 seconds. Electrocardiography showed right axis deviation, a notched P; and variable widening of P. Digitalization and oxygen therapy led to moderate improvement but the liver edge remained 5 cm. below the costal margin and venous pressure only fell to 255 mm. saline. Digitalis dosage was extended to early toxicity and mercurhydrin was given every day for a week. Improvement did not result however and attacks of cyanosis and dyspnea necessitated the use of oxygen therapy. With the realization that surgery offered the patient his only chance for survival this was undertaken on October 11, 1951, 19 months after the onset of symptoms. Excision of the tumor was attempted but was unsuccessful.

Postmortem examination (N.Y.H. Autopsy No. 14349) revealed that the tumor mass, portions of which remained attached to a broad flat pedicle, took origin from the interatrial septum at a point anterior to the foramen ovale (Figure 6). As had been noted at operation, the tumor had a multilobular character and was moderately gelatinous in consistency. Microscopic examination disclosed that the tumor was covered by endocardium and consisted of an evenly stained acidophilic

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**FIGURE 5A**

*Figure 5: Intra-atrial myxoma, Case 2. Angiocardiograms.*

Frontal left heart angiocardiogram.

A. The enlarged left atrium and its appendage are well filled as are the left ventricle and aorta. An irregular oval filling defect resulted from displacement of opaque substance within the left atrium. This was constant on all left heart films in frontal and lateral projections.—**B. Tracing of A.** (Previously published in "Circulation," 6:762, 1953).
myxomatous stroma. The cells, often multinucleated, were widely spaced and had stellate cytoplasm. There was no evidence of malignancy.

The histologic diagnosis was myxoma. The right ventricle was moderately hypertrophied, its wall measuring 6 mm. in thickness. The edges of the mitral valve and chordae tendineae were thickened, possibly as a result of direct trauma caused


FIGURE 7A

Figure 7: Inter-atrial tumor, Case 3. Conventional chest roentgenograms.
A. Frontal view shows slight prominence of pulmonary artery segment.
B. Lateral esophogram reveals indentation of the lower esophagus by an enlarged left atrium.
by the tumor itself. Although it is conceivable that this change in the valve chordae tendineae may have been a result of rheumatic fever, careful study of many sections of the heart revealed no Aschoff bodies. There was evidence of previous embolic infarction in the left cerebral hemisphere and the kidneys.

*Case 3:* A 46 year old housewife (N.Y.H. No. 657427) was admitted (Service of Dr. Theodore W. Oppel) on May 26, 1953. Three days previously while bending to plant seeds she had sudden onset of discomfort in the chest and palpitation. Cough, dyspnea on exertion and two pillows orthopnea followed. In the past, she had always been well and denied sore throats, chorea, rheumatic fever or heart trouble. She lived on the fourth floor of a walk-up apartment and made the climb with ease and furthermore, she had been fond of mountain climbing.

On examination, she was dyspneic but not cyanotic; the pulse was 130 and

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**FIGURE 8:** *Inter-atrial tumor, Case 3. Angiocardiograms.*

*A. Frontal.* A constant filling defect approximately 4 cm. is seen in the enlarged left atrium.—*B.* Tracing of *A.*—*C.* *Left lateral.* A left atrial tumor measuring approximately 4 cm. in diameter is seen in the posterior aspect of the left atrium.—*D.* Tracing of *C.*
regular; blood pressure 150/90. Moist rales were present in the posterior lower third of the lungs. Presystolic and systolic apical murmurs were heard. On lying, only a systolic murmur was present. The liver was not enlarged and there was no leg edema. The frontal chest x-ray film (Figure 7A) revealed slight prominence of the pulmonary artery segment while the lateral esophogram (Figure 7B) demonstrated left atrial enlargement. The electrocardiogram showed normal sinus rhythm, deviation of the electrical axis to the left but without predominance of either ventricle.

She became asymptomatic after four days treatment with digitalis, nasal oxygen, mercurials and low salt diet. Because of heart failure in the presence of mitral stenosis, preoperation evaluation for mitral valvulotomy was undertaken. Dr. Daniel S. Lukas found an hemodynamic pattern suggestive of a mitral valve obstruction but it was unlike the cardiac catheterization findings of rheumatic heart disease.\textsuperscript{3} Angiocardiography showed a filling defect measuring approximately 4 cm. in diameter in an enlarged left atrium (Figure 8). On June 27, 1953, operation verified the presence of a left atrial tumor. Excision was not attempted because it seemed unlikely that the large tumor could be delivered through the normal sized left atrial appendage, and furthermore, the tumor felt hard and seemed to have a wide attachment along the interatrial septum.\textsuperscript{4} A biopsy was believed to be inadvisable.

**Discussion**

Primary tumors of the heart are uncommon, occurring it is estimated, in 0.05 per cent of all autopsies.\textsuperscript{5} Prichard\textsuperscript{6} reviewing the subject extensively in 1951, clearly established that myxomas are the most frequent primary cardiac tumors and are truly benign tumors of embryonic mesenchymal origin. The total number of myxomata reported is approximately 204.\textsuperscript{6,7} which includes recent reports.\textsuperscript{5,8-10} The age incidence was from three months to 68 years, the majority being between 30 and 60 years. The sex distribution was equal and Prichard reiterated that cardiac myxomas had never been recognized ante-mortem.\textsuperscript{6} Before him, Yater in 1931\textsuperscript{11} and Mahaim in 1945\textsuperscript{12} reviewed the literature. Myxomas of the heart constitute about 50 per cent of the primary heart tumors. With few exceptions they have occurred in the atria; in the left atrium in 75 per cent. In almost every instance, as in two of our cases the tumor was polypoid and attached by a stalk to the foramen ovale or its rim. Microscopically, myxomas are contained within unbroken endocardium and have few cells. These are usually giant or multinuclear without mitoses and contain loose vascular tissue, hemorrhage, lymphocytes, plasmacytes, mucin staining material and collagen.

Our cases showed many of the clinical features common to the myxomas.\textsuperscript{6} Obstruction at the mitral orifice was present in cases 2 and 3 and had at times been suspected in the first case. Two of the most recently reported cases also simulated rheumatic mitral stenosis.\textsuperscript{5,9} Another clinical feature, embolic phenomenon is not uncommon and was present in one of our cases.\textsuperscript{8} Sometimes thrombi overlie the surface of the tumor or fragments of the tumor break off and may cause emboli to lodge in the brain, lungs, aorta, renal arteries or extremities.\textsuperscript{5,6,10} Sudden death due to primary artery embolus from a right atrial tumor has been described.\textsuperscript{6} Although, our second patient had four embolic episodes, autopsy study did not reveal
tumor emboli. There were, however, evidences of previous infarction in the left cerebral hemisphere and kidneys. Dexter and Work described a patient with endocardial damage which they attributed to old rheumatic fever. Similarly, case 2 had mitral leaflet with chordae tendineae thickening and shortening. However, as in Dexter and Work's case, there were no Aschoff bodies, endocarditis or pericarditis. Localized trauma of the valve leaflets by the protruding tumor is the more likely explanation for the valve leaflet and chordae tendineae changes. Case 1 developed impure flutter and auricular fibrillation. This occurrence, as in Gillcrest and Miller's case is an indication of atrial disease. Case 2 finally developed intractable heart failure which became unresponsive to digitalis and mercurials, features which have been common in cardiac myxoma.

Conventional roentgenographic findings in left atrial myxoma have consisted of cardiac enlargement, prominence and enlargement of the pulmonary artery and left atrium. In one instance superior vena cava widening and moderate cardiac enlargement was seen in a patient who had right atrial myxoma and died in heart failure. Left atrial enlargement was present in all our patients; in case 1 it was large enough to be evident along the right cardiac border in frontal view (Figure 1) and was mistaken for a right mediastinal tumor. The above roentgen findings, however, are all consistent with the diagnosis of mitral stenosis of rheumatic fever origin. Conventional roentgenography has not been helpful in establishing the diagnosis of atrial myxoma.

Angiocardiography, on the other hand (Figures 2, 5, 8) showed characteristic and persistent filling defects in an enlarged chamber due to an intercavitary tumor which was attached to the interatrial septum. Such changes are not seen in mitral valvular stenosis due to rheumatic fever. Mural thrombi along the right atrial wall or in the left auricular appendage have occasionally been recognized. These, however, never attain the large size of myxomas nor do they occur in the favorite location of myxomas, along the interatrial septum. A recent report describes right atrial filling defects due to the turbulent stream of regurgitating blood through an incompetent tricuspid valve. However, these are but transient defects and therefore should offer no great problems in diagnosis.

SUMMARY AND CONCLUSIONS

Three cases of left atrial intracavitary tumor were diagnosed in life by means of angiocardiography. A constant filling defect in an enlarged atrium with attachment to the interatrial wall was proved at necropsy to be due to a large myxoma in two instances. Because of the polypoid and benign nature of cardiac myxomas, surgical exploration was undertaken in each instance. Death resulted in two desperately ill patients and the autopsies confirmed the angiocardiographic findings. This, as predicted by Mahaim made possible for the first time, the diagnosis of myxoma during life. If the index of suspicion is high and every case of mitral stenosis of obscure etiology and varying murmurs is carefully evaluated, more cases of myxoma will be discovered. With earlier operation, at a time when the patient is
in good general condition, and finally, with advances in intercardiac surgery, especially the mechanical heart and lung apparatus, myxoma may become a curable disease.

RESUMEN

Por medio de la angiocardiografía se diagnosticaron tres casos de tumor del atrio izquierdo, intracavitarios. En dos casos una falta de llenado constante en un atrio crecido, con inserción a la pared interatrial fué demostrado a la autopsia que eran grandes mixomas. En vista de la naturaleza polipoide benigna de los mixomas cardiacos, se emprendió la exploración quirúrgica en cada caso. La muerte ocurrió en dos casos desesperadamente enfermos y las autopsias demostraron los hallazgos angiocardiográficos. Esto, como ha sido predicho por Mahaim20 hizo posible por primera vez el diagnóstico del mixoma durante la vida. Si se mantiene una alerta constante a este respecto y si cada caso de estenosis mitral de obscura etiología y con ruidos variables es cuidadosamente estudiado, se descubrirán más casos de mixoma. Con operaciones más tempranas, cuando el enfermo está en buenas condiciones físicas y con los adelantos de la cirugía intracardíaca, en particular el corazón y el pulmón mecánicos, el mixoma puede venir a ser una enfermedad curable.

RESUME

Grâce à l’angiocardiographie, les auteurs ont pu diagnostiquer durant la vie trois cas de tumeur de l’oreillette gauche. Deux fois, une oreillette qui paraissait augmentée de volume mais avait un emplissage défectueux, s’est montrée à l’autopsie, en rapport avec un volumineux myxome. Etant donné la nature bénigne des myxomes cardiaques, une exploration chirurgicale à été tentée dans chaque cas. Chez deux malades très atteints, la mort suivit l’intervention et les autopsies confirmèrent les constatations angiocardiographiques. Ainsi, pour la première fois se trouvait réalisé ce qu’avait prévu Mahaim20 sur la possibilité de faire le diagnostic de myxome durant la vie. Si on sait s’en méfier et si chaque cas de rétrécissement mitral sans étatologie nette, s’accompagnant de variations de signes d’auscultation, était étudié avec soin, on découvrait plus de cas de myxomes. Grâce à une opération plus précoce au moment où le malade est en bon état général et grâce aux progrès de la chirurgie intra-cardiaque, en particulier à l’existence de coeurs et de poumons artificiels, le myxome pourrait devenir une maladie curable.

REFERENCES


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