Myxoma of the Left Atrium

Diagnosis Made during Life with Operative and Post-mortem Findings

By Henry P. Goldberg, M.D., Frank Glenn, M.D., Charles T. Dotter, M.D., and Israel Steinberg, M.D.

A case of intra-atrial myxoma manifested clinically by evidences of mitral stenosis and peripheral emboli is presented in detail. The diagnosis was made during life and confirmed by angiocardiography. The surgical procedure, attempted as a last resort, is described, as are the postmortem findings. This case points the way to possible surgical cure in similar lesions.

To date, approximately 350 primary tumors of the heart have been reported. Prichard and Mahaim's reviews¹, ² of the subject are notable. Approximately 126 of the reported primary cardiac tumors were myxomas, while 114 were sarcomas.¹, ³ Only five (all sarcomas) have been diagnosed correctly during life,⁴⁻⁸ and in no case was angiocardiography employed. The surgical removal of an intracardiac tumor has not as yet been reported although the possibility of such a procedure has been speculated upon.⁹ The following report is concerned with the clinical, operative and pathologic findings in a 3½ year old child with a myxoma of the left atrium. In this patient the lesion was suspected on the basis of clinical findings, established with reasonable certainty by angiocardiography, and proved at surgery and subsequent postmortem examination.

Case Report

The patient (N.Y.H. 591 973), a white male child 3 years of age, was first admitted to the New York Hospital on Feb. 6, 1951.* The child's past history was unremarkable except for measles at 2½ months and chicken pox at 6 months. The family history revealed rheumatic fever in the paternal grandmother and a paternal aunt. The patient had good health until March 17, 1950, when (at the age of 2 years and 3 months) he suddenly developed a right sided hemiparesis. The paralysis lasted for only 10 minutes, but weakness remained. He was admitted to another hospital 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 Sept. 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 Oct. 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. The four embolic episodes coupled with the appearance of signs of mitral stenosis in a 3 year old boy led to the consideration of a tumor of the left atrium as the primary condition.

Physical examination on transfer to the New York Hospital showed a well-developed 3 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 again showed a

*Patient referred from the Roosevelt Hospital, New York City by Dr. Edmund Joyner.
large left atrium and right ventricle but the left ventricle appeared normal in size. The pulmonary arteries were normal.

The pulmonary orifice (figs. 1 and 2). The left atrial appendage was uninvolved. These findings were interpreted as indicative of a tumor (probably a myxoma) within the

FIG. 1. Angiocardiography. Films selected from serial studies in frontal projection made following intravenous injection of 20 cc. of Urokon sodium (70 per cent). Between 4.5 and 6.0 seconds following the injection there was good opacification of the pulmonary veins, the left atrium and ventricle and the aorta. The left atrium was enlarged and was the site of a large irregular filling defect, present on all films.

The course in the hospital was unremarkable except for a brief generalized convulsion during venepuncture. Angiocardiography demonstrated a large, irregular, polypoid filling defect in the left atrium anteriorly, immediately above the mitral left atrium. Surgical treatment was postponed in view of the great risk involved and in 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 Sept. 12, 1951, the child developed edema of the face, arms and legs; the child 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_{r} and variable widening of P_{r}. 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 Mercuhydrin 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 Oct. 11, 1951, 19 months after the onset of symptoms.

The patient, who had been receiving oxygen therapy, was taken to the operating room in his bed and anesthetized by the addition of cyclopropane to the oxygen circuit. An intratracheal tube was inserted and a slow infusion of blood and 5 per cent glucose in distilled water was started by means of intravenous plastic cannulae placed through cut-downs in each leg. The patient was positioned on his back with the left side slightly elevated and the skin was prepared and draped so as to expose the left chest. At this point there had occurred no perceptible change in the patient's condition; the color of skin and mucous membranes was normal, the pulse was 160 and the blood pressure was 90/50.

A curving incision, started at the level of the left second rib in the midsternal line, was carried below the breast area into the left axilla. The pectoral muscles were reflected from the chest wall and the third rib resected subperiostially from the costal-sternal junction to the midaxillary line. Upon entry into the left pleural space, the lung which was heavy and edematous, did not collapse. There was approximately 200 cc. of straw colored fluid in the left chest. The heart appeared moderately enlarged and the pericardium was full and tense, especially at the upper portion over the left atrium. Two cubic centimeters of 1 per cent novacaine were introduced into the pericardial sac and within two minutes the heart rate began to slow. Over the next two minutes it came to a standstill. Manual cardiac massage was instituted and continued for 20 minutes without re-establishment of cardiac action. It was then decided to attempt removal of the tumor since it was believed to be occluding the blood flow through the left heart. An incision was made in the pericardium, parallel and medial to the phrenic nerve, over the pulmonary artery and down to the midportion of the left ventricle. The left atrium was found to be prominent and a firm resistance on palpation suggested that it contained a tumor.

When the atrial appendage was opened, a mass consisting of many small, glistening, semitranslucent, globular bodies bulged up into it from the atrium (fig. 3). Although poor cardiac contractions occurred at this time, no blood escaped into the atrial appendage from around the tumor. By using a pituitary rongeur, projections of the tumor were readily removed. As each pull with the instrument brought away 1 or 2 cc. of tumor, more of the same material appeared at the orifice of the appendage. After 10 or 12 cc. had been removed with the rongeur, a pituitary spoon was introduced into the atrium, and further large globs or segments of tumor were freed from its base and delivered through the appendage. The small finger of the left hand was then introduced into the left atrium through the appendage. The base or stalk of the tumor could be palpated and seemed to be about 3 cm. in diameter,
Fig. 3. Surgical Procedure (artist's reconstruction). When the atrial appendage was opened, the polypoid tumor mass bulged out (above). Small portions of the projecting mass were removed with a pituitary rongeur (middle), reducing the volume of the tumor and leaving the left atrium relatively free for the passage of blood (below).

Fig. 4. Gross Pathology. The tumor arises from the anterior portion of the interatrial septum just above the mitral valve orifice. The chordae tendineae of the mitral valve are shortened and thickened.
took origin from the interatrial septum at a point anterior to the foramen ovale (fig. 4). As had been noted at operation, the tumor had a multilobular character and was moderately gelatinous in consistency. Microscopic examination (fig. 5) disclosed that the tumor was covered by endocardium and consisted of an evenly stained acidophilic 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 the chordae tendineae were thickened, possibly as a result of direct trauma caused by the tumor itself. Although it is conceivable that this change in the valve and chordae tendineae may have been a result of rheumatic fever, careful study of many sections of the heart revealed no Anchoff bodies. There was evidence of previous embolic infarction in the left cerebral hemisphere and the kidneys.

**DISCUSSION**

*Diagnostic Considerations*

Although the antemortem diagnosis of primary tumor of the heart has been made five times previously, the present case is believed to represent the first recorded instance where the diagnosis of an intracardiac myxoma was made during life. The lesion was suspected for the following reasons:

Changing auscultatory signs suggested the presence of obstruction of the mitral valve not present at birth and therefore presumably not of congenital origin. The development of rheumatic mitral stenosis prior to the age of 3 in a child who had shown no evidence of rheumatic fever was considered to be unlikely. Repeated embolic episodes are frequently associated with intracardiac tumor masses, especially myxomas. The most likely diagnosis was thought to be that of an intracardiac tumor causing obstruction to the mitral orifice although it was considered possible that valvular stenosis coupled with intra-atrial thrombus formation could have caused the clinical picture. It is of interest in this connection that auricular fibrillation was never noted.

Angiocardiography in frontal and lateral projections established beyond doubt that a polypoid mass, approximately 5 by 3 by 3 cm. in size was located in the anterior portion of an enlarged left atrium, apparently attached to the interatrial septum and in a position capable of occluding the mitral orifice. No pedicle could be detected. This angiocardiographic examination, performed eight months prior to surgery, is believed to constitute the first visualization of a proved intracardiac tumor in the living patient.* Mahaim² correctly suggested that angiocardiography might afford a means for the identification of such lesions during life. It is apparent that many of the reported cases of intracardiac tumor would have been strikingly demonstrated had angiocardiography been available or employed.

*Surgical Considerations*

At the time operation was undertaken on the patient reported here the tumor virtually

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* A 52 year old female with similar angiocardiographic findings is being followed currently in the clinics of the New York Hospital. Cardiac catheterization indicated the presence of physiologic mitral stenosis of a mild degree probably due to mechanical interference with blood flow through the mitral valve caused by the large tumor mass.
filled the left atrium, markedly impeding the flow of blood. Progressive deterioration of the patient's status had occurred during the period prior to surgery and thorough digitalization did not reverse decompensation. Although the induction of anesthesia and the prone position were well tolerated, opening the chest and introduction of 2 cc. of procaine into the tense pericardial cavity was followed by cardiac arrest. It seems logical to assume that a change in intra-atrial pressure or a shift in the position of the tumor mass cut off blood flow and resulted in catastrophe.

In retrospect, it is felt that in the face of cardiac arrest it might have been more logical to proceed immediately with removal of the tumor, postponing cardiac resuscitation until this had been accomplished. Had this been done, the period of anoxia might have been six or seven instead of 40 minutes. Such tumors, if they are to be removed successfully, should be operated upon before they have grown so large as to fill the left atrium, impede the blood flow and give rise to cardiac failure. Since intracardiac myxomas are usually benign lesions, surgery would appear to be indicated in certain instances, and, it is predicted, will eventually prove to be a curative measure. The development of apparatus for maintenance of the extracorporeal circulation would be of obvious value.

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HENRY P. GOLDBERG, FRANK GLENN, CHARLES T. DOTTER and ISRAEL STEINBERG

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