— Case Report —

A Case of Myxoma in the Left Atrium

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Primary tumor of the heart is comparatively rare. Recently, as the results of development of the cardiac catheterization, cardiovascular angiography and artificial heart-lung, both pre-operative diagnosis and successful extirpation of the tumor seem to be made more frequently. The case reported by SAKAKIBARA (1960) seems to be first one which operation succeeded in Japan. Concerning the nature of myxoma there are two theories, one is neoplastic, and the other non-neoplastic. And the latter has considered it to be a organized thrombus. With the diagnosis of mitral stenosis, operation was performed on a 28 years female to find a walnut-sized myxoma in the atrium.

Because the artificial heart was not ready for use, the operation was terminated as exploratory one.

PRIMARY TUMOR of the heart is comparatively rare. But it has been known since early, and already in 1855 an anatomist MALPIGHI reported it, and up-to-date about one hundred sixty cases have been reported.

According to SAKAKIBARA¹²⁾ et al., myxoma constitutes the largest proportion of cardiac tumors, and according to MANHAIM, the rate is about 1/3. In the literature are found 37 operated cases of myxoma³⁾¹²⁾¹³⁾¹⁵⁾, and operation were successful in 18 of them including of the one operated by CRAFT in 1954. Recently, as the result of development of the cardiac catheterization, cardiovascular angiography and artificial heart-lung, both pre-operative diagnosis and successful extirpation of the tumor seem to be made more frequently. In Japan, autopsy reports constitute greater proportion, and cases reported by IMAI (1934), YAMASHITA, TANABE (1954) KISHIMOTO (1959), TAKATAMA, KISHI and OONEDA

(1958) numbered as many as 10. However, the case reported by SAKAKIBARA (1960) seems to be the first one which operation succeeded. In our clinic a patient was recently admitted with the diagnosis of mitral stenosis, and on operation she was found to have myxoma in her left atrium. The particulars about this patient are as follows;

Patient: 38 years old, housewife.

Chief complaints: Dysnea and expectoration.

Past history: Non-contributory. Naturally healthy, having neither rheumatism nor tuberculosis.

Family history: Non-contributory.

Present illness: About January 1962, she abruptly felt dyspnea when lying on bed, and passed into the state of orthopnea.

About July of the same year, she felt dyspnea when walking. She visited Nakao Department of Internal Medicine, in our University Hospital where diagnosis of mitral stenosis was

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made. About January 1963, lassitude, anorexia and dyspnea during the walking became severer, coughing and expectoration developed, and the sputum assumed rusty color. In January of this year, she was hospitalized into the Adult Diseases Center Maebashi, and treated with diagnosis of mitral stenosis. On April 18, she was admitted into this department for operation.

Photo 1.

Physical examination on admission: The patient had moderate build and nutrition, ordinary development of musculature and subcutaneous fat and normal physiognomy; body temperature, 36.5°C; no anemia in conjunctiva; blood pressure 102/68 on left and 102/70 on right; pulse rate 90, normal sinus rhythm with satisfactory tension. No abdominal masses palpable in the abdomen. No ascite, No distension; the spleen

Photo 2.

TABLE I RESULTS OF CLINICAL EXAMINATIONS

<i>Blood</i>		<i>Liver function</i>
<i>Hb (Sahli)</i>	10.5%	<i>Meulengracht</i> 3
<i>Erythrocyts</i>	429×10^4	<i>B. S. P. (45)</i> 0%
<i>Leukocyte</i>	8000	<i>Kunkel</i> 2v
<i>Serum Protein</i>	7.7 g/dl	<i>Thymol</i> 1u
<i>GB</i>	1058	<i>Serum Electrolyte</i>
<i>GP</i>	1028	<i>Na</i> 13 mEq/l
<i>Ht</i>	47%	<i>K</i> 5.8 mEq/l
<i>Bleeding Time</i>	2'30"	<i>Ca</i> 3.9 mEq/l
<i>E. S. R.</i>	1/2 hour 7 mm	<i>Cholesterol</i> 190 mg/dl
	1 hour 18 mm	<i>Test of P. S. P.</i>
<i>C. R. P.</i>	(-)	15' 48% 60' 85%
<i>Wassermann</i>	(-)	30' 62% 120' 100%
<i>Urine and Stool</i>	normal	<i>Adrenal function</i>
		17 KS (urine) 4.8 mg/day
		17 OHCS (urine) 5.4 mg/day

and the kidney not palpable, and the liver is soft, and palpable 2 fingers wide at the right costal margin; and no abnormalities in the extremities.

Results of Laboratory examination: As seen in Table I, there were no abnormalities in blood and in hepatic, renal and adrenal functions, and CRP test and Wassermann test were negative. Lung capacity 2,400 cc. No abnormality in urine and stool.

Thoracic findings: Physically, thrill was palpable over the apex of heart at diastole, and the first sound was accelerated, and opening snap-like murmur was heard. In the chest roentgenogram, the lung field was relatively dark, and as to the shadow of the heart, contraction was shown at the first left arch, elevation at the second and third left, and distension at the first left arch, elevation at the second and third left, and distension at the second right (photos 1, 2, 3,). Electrocardiogram (photos 4, 5) indicated right axis deviation but not small auricular deflections. Phonocardiogram showed weak pre-

systolic murmur and diastolic murmur at the apex of the heart, and also opening snap-like murmur (photo 6). From these data, diagnosis of mitral stenosis was clinically made. And operation was performed without cardiac catheterization.

Findings on operation: According to the routine procedure, Opistan and Atropine Sulfate were used for basal anesthesia, and operation performed under fluothane intratracheal anesthesia. The chest was opened by anterolateral incision in IV intercostal region, and the pericardium was cut longitudinally along phrenic nerve. After suturing the pericardium to skin and elevating the heart, purse-string suture was made in the left auricular appendage, the right index finger was inserted into it. And a goose-egg-sized tumor was found filling the left atrium. The state of the mitral opening could not be elucidated. Diagnosis of left auricular myxoma was made, but because the artificial heart-lung was not ready for use, the radical operation was postponed. Immediately the finger was pulled

Photo 3. Roentgenogram of the chest on admission to the hospital.

Photo 4. Electrocardiogram

out, the atrium was closed, and the operation was terminated as exploratory one.

Post-operative course: During the operation, blood pressure tended to fall to 80mmHg at systole. Immediately after operation, cyanosis of the extremities developed, and dyspnea and a fall in blood pressure took place, and the patient fell into acute cardiac insufficiency. She did not respond to cardiant, and died 2 hours after operation.

Autopsy findings: In the left atrium was found a walnut-sized (5 cm in dia.) nearly spherically formed yellow tumor with gelatinous appearance and of soft consistency (photo 7). This was stalked tumor, grown from the septum about 2.0 cm below the fossa ovalis. The stalk was extremely short and slender (0.8 cm in dia.). The surface of the tumor was fairly smooth, and white millet-sized fibrin-like substances and dark red hemorrhagic part were

scatteredly seen (photo 8). The cut surface, nearly like exterior surface, was yellowish and gelatinous, presenting scattered hemorrhages. The tumor was considerably large, occupying the major space of the left atrium, and the base seemed to block the mitral orifice. The left atrial endocardium was fibrously thickened, especially endocardium which was in contact with the tumor was flatly elevated, assuming yellowish color. No thickening, shortening and deformities were observed in the mitral valve, and the other valves were also normal. Fossa ovalis showed a valvular-competent. In the left lung was observed remarkable congestion, which was considered to have resulted abruptly from closure of the openings of the pulmonary vein and mitral orifice owing to some unknown reason.

However, whether this was the cause of death or not could not be decided from autopsy findings.

Histological finding: The tumor was fairly myxomatous, poor in cellular elements, and rich in stroma (photo 9.) Tumor cells were comprised of spindle-shaped or stellate-shaped mononuclear cells and polynuclear giant cells, the latter being remarkably numerous. The cyto-

Photo 5. Electrocardiogram

Photo 6. Electrophonogram

Photo 7. In the left atrium was found a walnut-sized tumor (5 cm in dia).

Photo 8. The surface of the tumor was fairly smooth, and white millet-sized fibrin-like substance and dark red hemorrhagic part were scatteredly seen.

Photo 9. The tumor was myxomatous, poor in cellular elements, and rich in ground substance. Hematoxylin and eosin stain.

plasm was basophilic, and in some cells PTAH-positive fibrils were found running along the long axis. Mitosis was not observed, but pleomorphism of the nucleus was remarkable (photo 10). The tumor tissue was supplied by numerous capillaries, and scatteredly contained lymphocytes, plasmocytes, mast cells and histiocytes. It also showed hemorrhage in some parts. There

Photo 10. Tumor cells were comprized of spindle-shaped or stellateshaped mononuclear cells and polynuclear giant cells, the latter being remarkably numerous. Hematoxylin and eosin stain.

were observed some histiocytes taking hemosiderin, which, however, were generally confined to the surrounding of the hemorrhagic foci. The stroma of tumor was rich in acid mucopolysaccharide, and contained small amount of collagenous fibers, reticular fibers and elastic fibers. Staining with oil red O disclosed infiltration of a large amount of plasma lipoprotein in the stroma. The exterior surface of the tumor was mostly covered with endothelial cells excepting a part covered with white thrombi. The protoplasm of the endothelial cell was considerably basophilic, and the proliferation of endothelial cells was partly noticed. These are characteristic findings of the so-called cardiac myxoma.

Photo 11. A part of endocardium to which the tumor is attached. The endocardium is fibrously thickened, and subendocardial elastic membrane shows reduplication and fragmentation. In the interspace between the reduplicated elastic membranes, fibrous node, rich in acid mucopolysaccharide, is seen. Elastica van Gieson stain.

There are reduplication and splitting of sub-endocardial elastic membrane at the root of the stalk of the tumor in the left atrium, and in the interspace of reduplicated elastic membranes, a few nodules with considerable fibrosis are seen. These nodules contain pleomorphic fibroblast-like cells resembling mononuclear cells which constitute myxoma, and in their stroma a large amount of acid mucopolysaccharide was noted. This phenomenon can be seen in myxoma. A part of the nodules is continuous with tissue of the tumor (photo 11).

DISCUSSION

Among the various primary tumors of the heart, myxoma has the highest frequency, followed by sarcoma, rhabdomyoma and fibroma. Myxoma is most frequent in the atrium, especially in the left atrium. Concerning the nature of myxoma there are two theories, one is neoplastic, and the other non-neoplastic. And the latter has considered it to be a organised thrombus.

At the junction of the tumor in the atrial septum, in the present case there was no aggregation of flattened cells as described by PRICHARD¹¹⁾, but a few acid mucopolysaccharide rich nodules, having fairly close resemblance to the tumor tissue. This is assumed to be myxoid tissue of embryonal endocardium (RIBBERT)¹⁶⁾. From the facts that these nodules were continuous with the tumor tissue, and that organization of thrombus was seen only partially, the tumor was assumed not to be derived from organization of thrombus, but to be a true tumor. As to its symptoms, murmur resembling that of mitral stenosis is heard, changing its character according to the position of the body. Cyanosis develops paroxysmally, and sometimes syncope takes place. These symptoms seem to be ascribed to closure of the mitral opening by the stalked tumor. Since the history of disease revealed abrupt dyspnea and orthopnea on lying on bed and during walking, it was later considered that more precise examination should have been made by cardiac catheterization or angiocardiography. Even by these methods, however, it seems considerably difficult to make preoperative diagnosis. SAKAKIBARA¹²⁾ reported that in a

24 years female, enlargement of the right ventricle was shown both by roentgenogram and by electrocardiogram, remarkable systolic murmur in the left second intercostal region by phonocardiogram, and elevation of right ventricular pressure by catheterization, and that on the basis of these clinical findings, diagnosis of pulmonary valve stenosis was made, but that on operation a tumor was found in the left ventricle, which produced stenosis by pressing the opening of the pulmonary artery. Moreover myxoma is apt to occur in the left auricle, and for this reason its extirpation is difficult, and survival rate after operation was 18 out of 37 reported cases. The principal cause of death after operation was massive bleeding. According to the statistics up to 1958 by COOLEY³⁾ the rate of successful operation is 50%, and operation not under direct observation succeeded only in 2 of 15 cases, that is, the death rate of 86%. On account of this, it is considered that the operation of myxoma should be made at low body temperature or using the artificial heart-lung. ANDERSON (1961)¹⁾ performed 5 operations of myxoma in the left atrium at Toronto general hospital in the past 5 years, the first 3 of which were unsuccessfully made with the clinical diagnosis of mitral stenosis, but the 4th, which was performed after correcting diagnosis by catheterization, was successful. He reported the following as clinical characteristics of the tumor of the left atrium; (1) Having relatively short history of disease without arrhythmia and carditis; (2) Symptoms suspicious of intermittent stenosis; (3) The loud first sound without opening snap; (4) irregular diastolic murmur, heard at abnormal place; (5) in the case of sinus rhythm, absence accelerated diastolic murmur preceding systole (6) no complication denying mitral stenosis. If these findings are obtained, he said, left auricular tumor can in general be suspected, but in this case collaboration by cardioangiography is indispensable. In the present case, the symptoms so closely resembled those of mitral stenosis, though with some doubts, that we performed operation without cardioangiography and cardiac catheterization.

SUMMARY

With the diagnosis of mitral stenosis, operation was performed on a 38-years female to find a walnut-sized myxoma in the left atrium. Because the artificial heart was not ready for use, the operation was terminated as exploratory one.

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