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## Mitral Regurgitation due to a Calcified Myxoma

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### Introduction

Myxoma is a rare lesion which occurs in 35 to 50% of the primary cardiac tumors. Many successful cases have been reported since CRAFOORD<sup>4)</sup> successfully excised a left atrial myxoma using H-L bypass in 1954.

Improvements in ultrasonic cardiography have facilitated accurate diagnosis, thus excision of myxoma in the early stages has brought about a low surgical mortality. Most cases of myxoma in left atrium (LA) have findings which are similar to mitral stenosis. But other than annular dilatation, mitral regurgitation due to trauma from the pedunculated tumor is very rare. A case of myxoma in LA, the surface of which had a calcification in the top and perforation in the posterior leaflet of the mitral valve, is reported herein.

### Case report

The patient, a 15-year-old boy, had been healthy since birth. He had neither cyanosis nor cardiac murmur. But he was prone to respiratory infection, occasionally associated with stridor, and needed long-term treatment. At the age of 14, he consulted a family physician due to dyspnea and palpitation. He was diagnosed as heart valvular disease and was advised to have an accurate examination. The patient had no past history of rheumatic fever.

On February 22, 1972, the first cardiac catheterization was performed in a local hospital, but the diagnosis was inaccurate.

On March 6, 1972, a hemorrhagic tendency appeared which was diagnosed as purpura. Later, edema, albuminuria and hematuria developed, and he was diagnosed as acute nephritis. The following day, dyspnea and heart failure developed. He was subsequently put on a regime of diuretic and digitalis, with only transient improvement.

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Key words: Calcified myxoma, Mitral regurgitation, Perforation of the posterior leaflet, Echocardiography.

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However, his condition deteriorated with increased edema and dyspnea.

On October 4, 1972, he was admitted to Tenri Hospital in order to undergo a cardiac catheterization. Upon admission, he was lean, poor in nutrition and slightly cyanotic. Neither anemia nor icterus was present. Edema was noticed in face, but not in the limbs. The liver was palpable 6 cm below the right costal margin. Signs of moderate ascites were noted. Body temperature was normal. Precordial bulging and thrust at the apex were noted. A Grade 3-4 harsh systolic murmur was heard loudest in the apex and the left sternal border.

The pulmonary component of the second heart sound was very accentuated and a third heart sound was noted. The systolic thrill was palpable in the left third intercostal space. On electrocardiogram, a remarkable right ventricular hypertrophy, ST depression of 0.2 mV in Lead VI and large P wave were noted. On chest roentgenogram, cardiac shadow was very enlarged, with a CTR of 0.61. Pulmonary vasculatures did not increase.

On October 4, 1972, a second cardiac catheterization was performed. Intracardiac pressures (mmHg) were as follows: RA 20, RV 102-112/13-19, 26-29 (58), m-PA 98-112/70-77 (87) and LA 13 (Table 1). There was an equivalent pulmonary hypertension but no shunt. Cardiac output was very low: 2.0 L/min. During cardiac catheterization, calcification shadow was noted on the image, which was considered to be around the mitral valve. This shadow moved left downwards in the diastolic phase and right upwards in the systolic phase. Retrograde LV angiogram revealed a severe mitral regurgitation, but did not clearly reveal a filling defect.

On ultrasonic cardiography, an intracavitary mass echo, which had a strong echo intensity in the partial portion, was detected. The mass echo moved rapidly across the mitral valve from the LA to the LV in the diastolic phase, and from the LV to the LA in the systolic phase (Fig. 1).

Total serum protein was 5.5 gm/dl, albumin 2.3 gm/dl and globulin 3.2 gm/dl. Therefore,

**Table 1.** Preoperative cardiac catheterization data

| Site      | Pressure             | Oxygen (Vol %) |
|-----------|----------------------|----------------|
| SVC       | (20)                 | 7.40           |
| RA middle | (20)                 | 6.33           |
| IVC       | (20)                 | 9.46           |
| RV inf.   | 102-112/13-19, 26-29 | 6.33           |
| RV out    | 102-113/ 8-14, 22-25 | 5.98           |
| m-PA      | 98-112/70-77 (87)    | 6.17           |
| l-PA      | 106-119/70 (88)      | 6.21           |
| Ao.       | 123/98 (104)         |                |
| LA        | (13)                 |                |
| LV        | 111/11, 18           | 16.13(91.1%)   |

Pulse rate 125/min., Cardiac output 2.0 L/min.

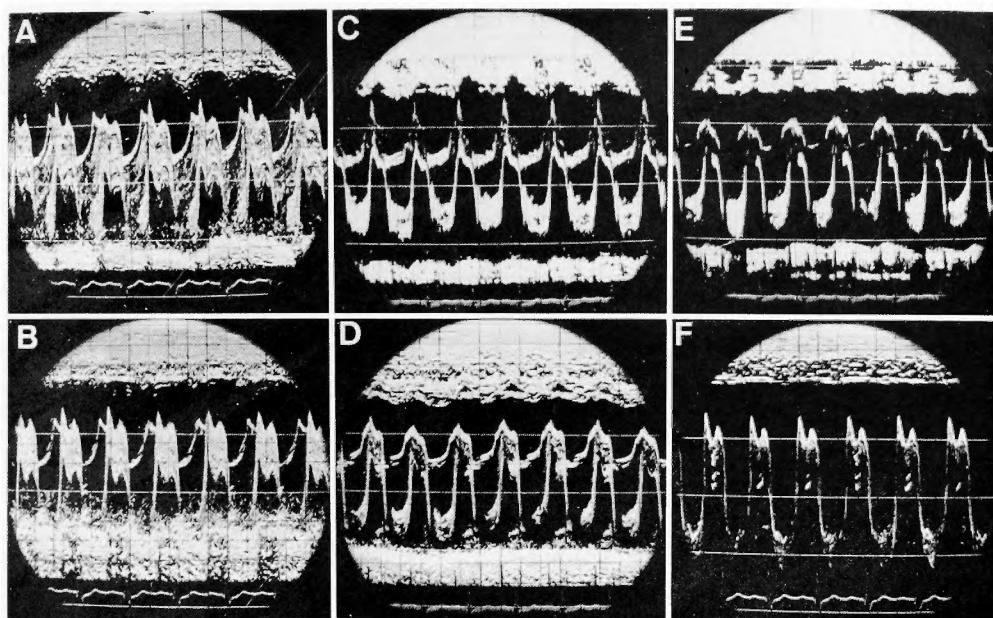
Cardiac Index 1.5 L/min./M<sup>2</sup>

Diagnosis: Severe MR (Grade 4°)

Tumor in LA (probably Myxoma)

( ): mean pressure

The intracardiac pressures of the right heart were variable due to dyspneic respiration.



**Fig. 1.** Ultrasonic cardiography (M-mode)

An intracavitary mass echo having a strong echo intensity in the partial portion is detected. The mass echo moves rapidly across the mitral valve from the left atrium to the left ventricle in the diastolic phase, and from the left ventricle to the left atrium in the systolic phase.

the A/G ratio was 0.72. The icterus index was over 10 units. Total serum bilirubin was 1.8 mg/dl with direct bilirubin of 72%. Respiratory function was moderately disturbed: %VC 52.6%, DLCO 48.5%, %MBC 61.5% and FeV 81.1%.

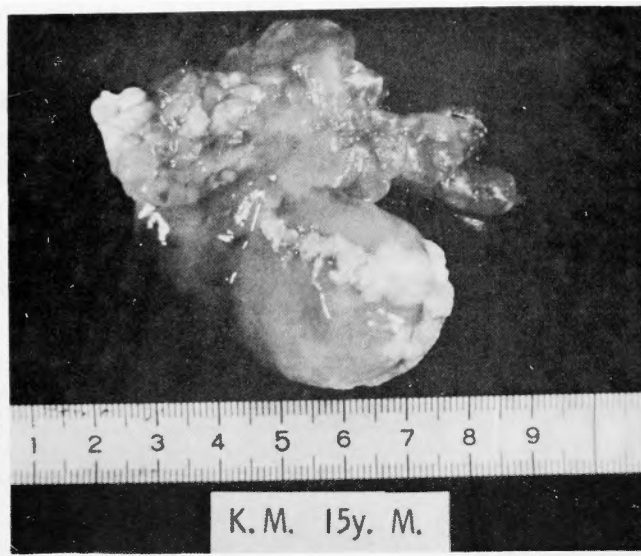
Since admission, he was under a severe regime of digitalis, diuretic and restriction of salt intake, for the treatment of heart failure.

On October 7, 1972, pulse decreased from 110 to 84 beats/min and appetite increased. Also urination increased, 1300–2000 ml/day, and ascites decreased. It was decided that an operation would be performed when the patient's condition improved. Though the patient's condition became relatively stable, on October 15, 1972, tachycardia, nausea, dyspnea, oliguria and cyanosis occurred. Oliguria was not affected by diuretic. The BUN value was 41.6 mg/dl, serum bilirubin 7.3 mg/dl and direct bilirubin 78%.

On October 22, 1972, orthopnea occurred, with bloody sputa, due to lung edema.

On October 26, 1972, an emergency operation was performed based on a diagnosis of tumor in LA, mitral regurgitation and pulmonary hypertension. Subcutaneous tissue was edematous and there was a massive pericardial effusion. Shortly after a pericardial incision was made, there was a rise in systemic blood pressure. The right and left ventricles were remarkably dilated and hypertrophied. The right atrium and the pulmonary artery were also dilated. The diameter of the aorta and the main pulmonary artery were 18 and 36 mm, respectively.

A Dubost's incision was made, reaching the left atrium. There was a myxoma in the LA, which was attached to the atrial septum by a pedicle. The myxoma consisted of a globar portion



**Fig. 2.** The excised specimen  
The excised myxoma consisted of a globar portion and a jelly-like portion. Both portions are connected by a stalk. In the top of the globar portion, an exposed calcification is found. The size of the myxoma is  $5.5 \times 5.0 \times 3.1$  cm; the weight is 30 gm.

and a jelly-like portion (Fig. 2). Both portions were connected by a stalk. A portion of the atrial septum was removed at the point where the stalk of the myxoma was attached. The size of the myxoma was  $5.5 \times 5.0 \times 3.1$  cm; the weight was 30 gm. A calcified degeneration in the top of the myxoma was found, which was touching the center of the posterior leaflet of the



**Fig. 3.** Roentgenogram of the excised myxoma  
In the top of the globar portion, a heavy calcification is found.

mitral valve (Fig. 3). A perforation, 9.5 mm in diameter, was found in the center of the posterior mitral leaflet, resulting in regurgitation. This perforation was closed by direct suture. Then mitral annuloplasty in the posterior commissure was performed with 2 mattress sutures. Thereby the mitral valvular orifice (5 cm in diameter) decreased to 3.3 cm. Evaluation of mitral regurgitation during operation was performed by means of injecting saline solution from LV vent. Slight mitral regurgitation was noted.

Weaning from H-L bypass was without difficulty. A slight systolic thrill was palpable on the left atrial wall.

The intracardiac pressures after surgical procedures were as follows: m-PA 60/34 (44), RV 52/-4,8, LV 104/0.7, FA 104/52 (68) and LA v: 20, m: 16.

Sudden cardiac arrest occurred postoperatively and the heart was resuscitated by 15 minutes of external cardiac massage, followed by re sternotomy in order to prevent cardiac tamponade. The heart was dilated more than before the resuscitation. Bleeding was stopped, and the pericardium remained opened. But the patient was comatose. On November 2, 1972, the patient died of cerebral edema and renal failure. The duration of the occlusion of the cerebral circulation may have been too long and thus the patient did not regain consciousness. Autopsy was denied.

### Discussion

Myxomata constitute 35 to 50% of all primary cardiac tumors<sup>2)</sup> and are the most common intracavitary tumors. They occur at all ages, although they are rare in childhood, and are three times more common in women.<sup>6,7)</sup> Although myxoma has been considered a rare lesion, the number of surgical cases has been increasing. Preoperative diagnosis of this lesion is no longer unusual, and now a successful operation can be expected. The etiology of myxoma, like that of most neoplasm, is not understood.

The size, location, and pedunculated character of the myxoma results in a ball valve type blockage of the mitral valve, simulating mitral stenosis. The tumor may interfere with closure of a normal mitral valve, or alternatively "wrecking ball" trauma from the pedunculated tumor, especially when there is calcification; this may result in valve leaflet scarring or even chordal rupture. Usually, heavy calcification of atrial myxoma is uncommon and appears to be more frequent when the tumor is in the right atrium. There have been a number of reports on calcified right atrial myxoma<sup>1,3,5,7,8,9,10,11,14,19)</sup>, but only a few on calcified left atrial myxoma<sup>9,15,19)</sup>. A left atrial myxoma damages not only the mitral valve<sup>13)</sup> but also the aortic valve<sup>16)</sup>, especially when there is calcification.

ROSE et al.<sup>16)</sup> reported a case of left atrial myxoma, in which the aortic regurgitation was caused by the rubbing of the myxoma against the aortic valve resulting in direct damage to the left and non-coronary cusps. Until this present case, there been no reports in which a calcified myxoma resulted in the perforation of the posterior mitral leaflet.

NASSER et al.<sup>13)</sup> reported a case in which four large perforations were in the anterior mitral leaflet with a marked scarring. When the mitral leaflet and chordae tendineae are damaged, mitral surgical procedures, i.e. valvuloplasty, annuloplasty<sup>12)</sup> and mitral valve replacement<sup>13)</sup>

may be needed, following excision of the atrial myxoma.

MACVAUGH et al.<sup>12)</sup> reported a case of calcified myxoma in which the successful mitral annuloplasty was performed by plicating the annulus along the posterior leaflet with a heavy Dacron suture backed with cloth pledgets. When an exposed calcification of myxoma is present, scarring and/or perforation of the mitral leaflet, or rupturing of the chordae tendineae may occur. Thus mitral valve replacement may be more advisable than valvuloplasty.

SHARRATT et al.<sup>18)</sup> reported an operative case of left atrial myxoma, in which mitral valve prolapse appeared to be the cause of the mitral regurgitation; as the regurgitation was mild, the mitral valve was not replaced, but a moderate mitral regurgitation still remained.

In our case, there was a perforation (9.5 mm in diameter) in the center of the posterior leaflet. This was considered to be the result of rubbing by the exposed calcification. This perforation was closed by direct sutures. In addition, mitral annuloplasty plicating the posterior commissure using 2 mattress sutures was performed because of dilatation of mitral annulus and shortening of the posterior leaflet following the direct closure of the perforation. A mitral regurgitation test performed by injecting saline solution from a LV vent revealed mild regurgitation. In our case, sudden cardiac arrest occurred shortly after the closing of the chest. And we thought that residual regurgitation and pericardial closure were the main causes of this cardiac tamponade. Retrospectively, mitral valve replacement and the opening of pericardium should have been performed.

Calcification of myxoma can be easily detected by ultrasonic cardiography<sup>17,20)</sup>, compared to chest roentgenography<sup>3,14,17)</sup>. In ultrasonic cardiogram, in addition to an intracavitary mass echo, a strong echo caused by calcification was easily detected. An increase in amplitude of the anterior leaflet and excursion of interventricular septum were noted. LV angiocardigram does not always detect the "filling defect" of myxoma, moreover with angiocardigram there is a greater possibility of embolism. Immediately when a calcified myxoma in LA is diagnosed, emergency surgery should be performed, if necessary, using a mitral prosthesis.

Ultrasonic cardiography is the most useful and safest examination in detecting whether a myxoma is calcified or not.

Recurrences of resected myxoma, although unusual, have been documented with increased frequency in recent years. Not only is ultrasonic cardiography useful in detecting myxoma and differentiating it from mitral stenosis but it is essential for examination in the follow-up study.

### Summary

The patient, a 15-year-old boy, had been suffering from left and right heart failure for one year. He underwent an operation based on preoperative diagnosis of calcified myxoma in the left atrium and mitral regurgitation. A perforation, 9.5 mm in diameter, in the center of the posterior leaflet of the mitral valve was found. The myxoma in LA had an exposed calcification in the top, which may have rubbed the posterior leaflet, resulting in the perforation. Severe brain damage was brought about following cardiac tamponade, which occurred shortly after the operation. Autopsy was denied.

In a calcified myxoma, ultrasonic cardiography is the most useful and safest examination.

When a calcified myxoma is diagnosed, emergency operation should be performed, considering the possible damage to the mitral leaflet and/or chordae tendineae.

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## 和文抄録

## 石灰化粘液腫による僧帽弁閉鎖不全症

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15才男子, 術前検査で石灰化粘液腫, 重症僧帽弁閉鎖不全, 肺高血圧と診断された. 両心不全が高度となり, 緊急に手術を施行. 粘液腫は,  $5.5 \times 5.0 \times 3.1$  cm, 30 gr. 先端部に石灰化を認めた. それが接する僧帽弁後尖中央部に 9.5 mm の穿孔を認めた. 粘液腫摘出お

よび穿孔閉鎖, 弁輪縫縮術を施行したが, 術后, 心タンポナーデから心停止を来たした. 蘇生に成功したが, 脳症を合併し, 術后 6 日目死亡した.

石灰化左房粘液腫の場合, 僧帽弁の機械的損傷による逆流を考慮すべきである.