Multiple Myxomas Originating From Anterior and Posterior Mitral Leaflets in the Left Ventricle Leading to LV Outflow Tract Obstruction

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An extremely rare case of myxomas originating from the mitral leaflets was diagnosed in a 64-year-old man presented with a history of exertional dyspnea and palpitations. Two masses originating from the anterior and posterior mitral leaflets in the left ventricular (LV) cavity, causing LV outflow obstruction, were detected by echocardiography. The myxomas were successfully removed with the mitral leaflets via left atriotomy and mitral valve replacement. No embolic events occurred in the preoperative or postoperative period. In this article, we wanted to present. (Circ J 2008; 72: 1709–1711)

Key Words: Cardiopulmonary bypass; Tumor; Valvular diseases

Intracardiac myxomas are the most common benign tumors of the heart in adults. Approximately 75–80% of myxomas originate from the left atrial septum; 10–20% are localized in the right atrium; and the rest of 6–8% are bi-atrial, or in the right or left ventricle! Isolated left ventricular (LV) myxoma is extremely rare, accounting for 2.5–4% of all cases. The most common treatment strategy is surgical extirpation under cardiopulmonary bypass, with an excellent prognosis. We present an extremely rare case of 2 LV myxomas originating from the mitral anterior and posterior leaflets into the LV cavity and obstructing the outflow tract (LVOT), and discuss the current literature.

Case Report

A 64-year-old man presented with a history of exertional dyspnea and palpitations and nonspecific chest pain. New York Heart Association (NYHA) functional capacity was class II. There was a grade 2/6 systolic murmur over the left sternal edge on auscultation. There was no history of fever, syncope, orthopnea, weight loss or fatigue, or of peripheral or cerebral embolism. Clinical signs of myxoma-like clubbing, atrial fibrillation, hepatomegaly or pre-tibial edema were not seen in the physical examination. Erythrocyte sedimentation rate was 46/h.

Echocardiography revealed 2 masses in the LV cavity. The larger one originating from the mitral posterior leaflet was 41×31 mm and was attached to both the interventricular septum and the posteromedial papillary muscle. The other had a diameter of 14×13 mm and originated from the anterior mitral leaflet. It was prolapsing through the aortic valve during systole, causing a severe obstruction (LVOT gradient; peak = 65.91 mmHg, mean = 46.46 mmHg) (Table 1, Fig 1). Coronary angiography was normal and ECG showed normal sinus rhythm.

Midline sternotomy and bicaval cannulation were performed and cardiac arrest was induced by antegrade cold blood cardioplegia and moderate hypothermia. A horizontal incision of the left atrium (LA) was made and the mitral leaflets were observed to be normal. Subvalvular structures related to the posteromedial papillary muscle were short, thick and attached to the larger mass. Initially an attempt was made to extract the masses from the LA through the mitral orifice. The small mass could be extracted by pressing the apex, but all of the large mass could not be extracted by this technique. After the mitral leaflets were resected, the gelatinous and yellow-brownish big mass was extracted easily (Fig 2). It was attached to the anterior mitral leaflet, posteromedial papillary muscle and interventricular septum. Because the masses were fragile, mini aortotomy was performed. The ascending aorta, LVOT, LV and LA cavities were controlled for embolic particles, and irrigated with saline. A bileaflet mitral valve replacement (MVR) was performed with a 27 M St Jude Medics (St. Jude Medical, Inc, St.Paul, MN, USA). Cross-clamp time was 122 min. There were no postoperative complications. Normally functioning metallic mitral valve and normal LV cavity were observed on postoperative echocardiographic examination. Myxoma was confirmed by pathological examination (Fig 3).

Discussion

Myxomas are the most common primary cardiac tumors, although LV myxoma is extremely rare? The best of our knowledge this is the first case of severe LVOT obstruction because of a myxoma originating from both mitral leaflets. Natale et al reported a case of severe LVOT obstruction caused by a myxoma, but the mass originated from the interventricular septum! In another case reported by Darwazah et al, multiple myxomas of the LV originating from the in-
Table 1  Echocardiography Findings

<table>
<thead>
<tr>
<th>Mitral valve</th>
<th>Aortic valve</th>
<th>Left atrium</th>
<th>Left ventricle</th>
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<tbody>
<tr>
<td>MVA: 4.0 cm², MR: 1⁺</td>
<td>AR: 1⁺, MG: 46.46 mmHg, PG: 65.91 mmHg</td>
<td>LAD: 43.0 mm</td>
<td>LVDD: 59.0 mm, LVSD: 42.0 mm, IVS: 10.0 mm, LVEF: 0.60, Large mass: 41×31 mm, Small mass: 14×13 mm</td>
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MVA, mitral valve area; MR, mitral regurgitation; AR, aortic regurgitation; MG, mean gradient; PG, peak gradient; LAD, left atrial diameter; LVDD, left ventricular diastolic diameter; LVSD, left ventricular systolic diameter; LVEF, left ventricular ejection fraction.

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**Fig 1.** Echocardiography of the ventricular myxomas.

**Fig 2.** Extracted myxoid masses and anterior mitral leaflet.

**Fig 3.** Pathological examination shows spindle-shaped cells, stellate cells and multinuclear primitive cells in a myxoid stroma (hematoxylin and eosin stain; original magnification, (a) ×10, (b) ×20).
terventricular septum and ventricular trabeculae presented clinically as an occlusion of the left axillary artery. Some cases of myxomas originating from the mitral leaflets have been documented, but the actual site of origin on the leaflets differs. Kelling et al reported a case of LVOT obstruction caused by a myxoma originating from the ventricular side of the mitral leaflet. In the present case, multcentric myxoma originating from the ventricular side of the anterior and posterior mitral leaflets caused severe LVOT obstruction (the peak and mean systolic pressure gradients of the aortic valve were 65.91 and 46.46 mmHg, respectively).

Systemic embolization occurs more frequently during LV systole in the case of myxomas and the size and localization of the myxoma are important factors because of the motion of the valve leaflets. A high rate of systemic embolization has been reported in 30–45% of the patients with LA myxoma, and the rate for myxoma localized in the LV cavity is 64%. In the present case it is interesting to note that although the multiple huge myxomas were localized in the LV cavity, there was no history of embolic events.

The recurrence of cardiac myxoma may be as early as 6 months and as late as 11 years after excision. Bossert et al reported no recurrence or late death in 59 patients, but others report a recurrence rate between 1.58% and 6.00%. In the present case, follow-up echocardiography in the 6 months postoperatively has not revealed a recurrence and the patient will continue to be examined at regular intervals.

The surgical approach to LV myxomas can be through a left ventriculotomy, left atriotomy or aortotomy, but it is generally through a left horizontal atriotomy and direct incision of the ventricular cavity. The mass with its peduncle is extracted and the adhesions are detached from the ventricular wall. If possible, large resection of the mitral leaflet is extracted and the adhesions are detached from the ventricular cavity. The mass with its peduncle presented firmly to the anterior mitral leaflet, and MVR should not be performed during extirpation of a myxoma. However, in the present case, the bigger of the 2 masses was attached firmly to the anterior mitral leaflet, posteromedial papillary muscle and interventricular septum. In addition, the subvalvular structures related to the posteromedial papillary muscle were short, thick and attached to the mass. Thus, the entire mass was resected from the interventricular septum with the anterior mitral leaflet because of concerns about residual tumor mass. We believe that by using this approach we also prevented a possible injury to the posterior wall and interventricular septum. However, MVR became inevitable, to avoid both complications and recurrence in this case.

It is well known that the LV approach increases surgical mortality rates in comparison with the LA approach. We also prefer left horizontal atriotomy as the surgical approach to avoid the possible complication of left ventriculotomy. Nevertheless, Thongcharoen et al presented a case report in which LV myxoma was removed successfully through a left ventriculotomy. Tisma et al also reported a case of LV myxoma removed successfully through an aortotomy. In the present case, mini aortotomy was performed only to prevent any embolic complicatoma.

In summary, in a case of multiple myxoma originating from the ventricular side of the mitral leaflets, MVR via a left atriotomy was successful.

References