

Mitral valve myxoma: Clinical features, current diagnostic approaches, and surgical management

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Abstract

Background: *The purpose of this article is to document the clinical features of the unusual mitral valve myxoma based on the literature of recent years.*

Methods: *A literature retrieval of the mitral valve myxoma reported in recent years was made using the MEDLINE and EMBASE databases. The clinical information about this unusual disorder was collected and analyzed.*

Results: *Mitral valve myxoma showed female predilection. Their major symptoms were cardiovascular or cerebrovascular, in addition to constitutional or embolic. The diagnosis was usually made based on two- and three-dimensional transthoracic and/or transesophageal echocardiography, as well as magnetic resonance imaging in the current era. Surgical resection with mitral valve defect repair was the commonest remedy for mitral valve myxoma when mitral valve function could be preserved, and the mitral valve should be replaced when necessary. Patients showed good response to surgical treatment during the follow-up of 17.0 ± 28.4 months after the operation.*

Conclusions: *Two- and three-dimensional echocardiography and magnetic resonance imaging are the major diagnostic tools for the diagnosis of a cardiac myxoma. After diagnosis, surgery should be performed urgently, in order to prevent complications such as embolic events or obstruction of the mitral orifice. Due to the fact that myxomas can recur, regular, postoperative cardiological control is mandatory. (Cardiol J 2012; 19, 1: 105–109)*

Key words: cardiac surgical procedures, mitral valve, myxoma

Introduction

Cardiac myxomas are the commonest primary cardiac tumors, accounting for 50% of cases. Myxoma is more frequent in females than in males [1]. Most cardiac myxomas arise from the fossa ovalis, while those that originate from the heart valves are uncommon [2]. The purpose of this article is to document the clinical features of the unusual mitral valve myxoma based on the literature of recent years.

Methods

Literature about mitral valve myxomas since 2006 in the MEDLINE and EMBASE databases was retrieved, and data were analyzed in terms of its clinical manifestation, optimal diagnostic methodology, surgical approaches, and long term follow-up.

Mitral valve myxoma is defined as a myxoma arising from the mitral leaflet, annulus, commissure, junction area or subvalvular apparatus (chorda or papillary muscle) with a stalk or presenting as

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a broad-based one. Those myxomas originating from other locations of the left heart chamber, but with adherence to the mitral valve or apparatus, were excluded.

Data were expressed in mean ± standard deviation and student t test was used to evaluate intergroup differences. A p < 0.05 was considered of statistical significance.

Results

In all, there were 55 patients with 64 mitral myxomas as described in 42 articles published since 2006 [3–44]. Of the 55 patients whose sex was recorded, there were 12 males and 19 females, showing female gender predominance. There was no significant difference between male and female patients' age (32.79 ± 22.39 years, 43.75 ± 18.73 years, p = 0.1692).

The symptoms were cardiovascular in 36.7% of patients, and cerebral in 30% of patients, constituting the two main presentations of mitral valve myxoma. Some patients presented with constitutional symptoms or emboli or were asymptomatic (Table 1). One patient had a family history of myxomas [28]. Heart murmur was recorded in nine patients: eight patients had an apical systolic murmur and one patient had a diastolic murmur.

Transthoracic and/or transesophageal echocardiography has become the first choice of diagnosis for cardiac myxomas. They were used in all 33 patients whose diagnostic methods were recorded. A mitral valve myxoma can be a large mass [23], pedunculated [21], non-pedunculated [27], sessile [40], heterogeneous [6, 16], homogeneous echogenic [40], multilobulated mobile [32], well-circumscribed [41], or irregular adherent [39]. The size, exact location, and the relation of the mitral myxoma to the mitral structures could be clearly visualized, and the transvalvular pressure gradient could be measured. A cardiac myxoma may be misdiagnosed by transthoracic echocardiography [19]. However, transthoracic and transesophageal echocardiography can complement the visualization of a mitral valve myxoma. In two of the 33 patients, cardiac magnetic resonance imaging was used as a supplementary diagnostic tool for mitral valve myxoma.

In 18 patients, the hemodynamic status associated with mitral valve myxoma was reported: three (16.7%) patients did not have any hemodynamic disorder, while the other 15 (83.3%) patients did (Table 2).

Of the 64 mitral myxomas, 44 were solitary, and the other 20, in 11 patients, were multiple cardiac

Table 1. The main symptoms of mitral valve myxoma.

Symptom	Cases (%)
Asymptomatic	3 (10)
Constitutional	4 (13.3)
Embolic	1 (3.3)
Cardiovascular	11 (36.7)
Cerebral	9 (30)
Cardiovascular and cerebral	2 (6.7)

Table 2. Hemodynamic disorders of 15 mitral myxoma patients.

Hemodynamic disorder	Cases (%)
LVOTO	5 (33.3)
Mitral regurgitation	5 (33.3)
Mitral stenosis	1 (6.7)
Mitral regurgitation + tricuspid regurgitation	2 (13.3)
LVOTO + mitral regurgitation	1 (6.7)
LVOTO + mitral stenosis	1 (6.7)

LVOTO — left ventricular outflow tract obstruction

myxomas. The atrial side of the anterior mitral leaflet was the commonest location of mitral myxoma of either multiple or solitary lesions (Table 3). The mitral myxomas were 2.96 ± 1.58 cm in size. The solitary tumors were 2.71 ± 1.38 cm, while the multiple ones measured 3.32 ± 1.78 cm. No difference was found between the sizes of the two groups (p = 0.2940). The mitral myxomas located on the atrial side of the mitral leaflet measured 2.85 ± 1.61 cm, while those on the ventricular side were 2.88 ± 1.82 cm in size. There was no significant difference in the tumor size between the atrial and ventricular side (p = 0.9783).

Of the mitral valve myxomas, 11 were pedicled and six were sessile with a pedicled-to-sessile ratio of 1.83:1.

Tumor resection was performed in 35 of 37 patients, and conservative treatment in only two patients (Table 4). Tumor resection was the commonest method of surgical treatment, followed by mitral valve replacement. Mitral valve defect repair was done in five patients, and endoscopic and minimally invasive approaches were attempted in one patient each (Table 4). Surgical approaches were mentioned in six cases: left atriotomy in three (50%), right atriotomy plus septal route in two (33.3%), and biatrial route in one (16.7%) patient, respectively.

Table 3. Sites of the mitral valve myxoma.

Site of mitral myxoma	Cases (%)
Multiple:	20 (30.3)
a.aml	11 (55)
a.pml	1 (5)
v.aml	1 (5)
?.pml	1 (5)
v.aml + papillary muscle + interventricular septum	1 (5)
annulus	1 (5)
papillary muscle	4 (20)
Solitary:	44 (66.7)
a.aml	19 (43.2)
annulus	3 (6.8)
a.aml + annulus	2 (4.5)
a.mitral valve	1 (2.3)
a.chord	1 (2.3)
?.pml	1 (2.3)
v.aml	1 (2.3)
v.aml + papillary muscle	1 (2.3)
v.pml	1 (2.3)
v.pml + chord	1 (2.3)
v.pml + annulus	1 (2.3)
mitral valve	8 (18.1)
papillary muscle	2 (4.5)
junction	2 (4.5)

? — either atrial or ventricular side of the mitral valve; a — atrial side of the mitral valve; aml — anterior mitral leaflet; pml — posterior mitral leaflet; v — ventricular side of the mitral valve

Table 4. Management of the mitral valve myxoma.

Management	Cases (%)
Tumor resection	15 (40.5)
Tumor shaved	2 (5.4)
Tumor resection + mitral valve preservation	1 (2.7)
Tumor resection + mitral valve defect repair	5 (13.5)
Tumor resection + mitral valve defect repair (endoscopic)	1 (2.7)
Tumor resection + tricuspid and mitral repair	1 (2.7)
Minimally invasive tumor resection	1 (2.7)
Mitral valve replacement	8 (21.6)
Tumor shaved + artificial chord + mitral ring insertion	1 (2.7)
Conservative treatment for transient ischemic attack	1 (2.7)
Follow-up without surgery was decided because of her young age, unaffected mitral valve functions and possible risk of early recurrent thoracotomy	1 (2.7)

The patients were observed for 17.0 ± 28.4 months after the operation. Only three complications were found in the patient setting: (1) a 40 year-old male patient had right retinal artery occlusion three weeks after the operation: he retained mild neurological deficit three months later [12]; (2) a saccular aneurysm of abdominal aorta in a 12 year-old girl three weeks after mitral myxoma resection: she underwent an emergent surgery, and was doing well at six month follow-up [16]; and (3) a 25 year-old female patient developed thrombotic mitral valvular obstruction one year later: a re-replacement of the mitral valve was performed and she was well at ten year follow-up [40].

Discussion

Heart myxomas are usually located in the atrial septum near the fossa ovalis or the heart walls and tend to be larger in size, thereby obstructing the valvular orifice, while those originating from the heart valves are uncommon and small in size [2]. Like the patients with usual cardiac myxomas, the clinical symptoms of mitral valve myxomas were predominantly cardiovascular (chest discomfort, nonspecific chest pain, shortness of breath, exertional dyspnea, orthopnoea, and palpitation) and cerebral (numbness, dysesthesia, hemiplegia, focal seizure, coma, transient ischemic attack, and stroke), and could be constitutional (fever, weight loss, night sweat, cachexia, weakness, and fatigue), or embolic. Some patients may have elevated erythrocyte sedimentation rate, rheumatoid factor, and C-reactive protein [16]. Apical systolic murmur could be audible in some patients. Patients with mitral myxoma may be associated with infective endocarditis [14]. Due to the influence of the mitral myxomas on the mitral orifice or other adjacent heart structures, hemodynamic disorders including left ventricular outflow tract obstruction, mitral valve stenosis or mitral valve regurgitation may occur.

In the very early days, the diagnoses of heart valve myxomas were always made incidentally at autopsy. Later, the clinical diagnosis mainly relied on surgical exploration or ventriculogram due to the limits of contemporary diagnostic techniques [2]. Nowadays, with the development of less and non-invasive diagnostic tools, myxomas, including valvular myxomas, can be easily visualized by two- and three-dimensional echocardiography or magnetic resonance imaging [45]. Transthoracic and transesophageal echocardiography can be insufficient in non-invasive imaging of a myxoma. Other methods, such as real-time three-dimensional

echocardiography or magnetic resonance imaging, should be used as complementary techniques to evaluate intracardiac tumors with congenital cardiac abnormalities before any cardiac surgery treatment [46].

Recurrence rates reported for cardiac myxomas are 4–7% for sporadic cases and 10–21% for familial cases. Although recurrence rates are high, second recurrences are rare [28].

After diagnosis, surgery should be performed urgently, in order to prevent complications such as embolic events or obstruction of the mitral orifice. Follow-up examination, including echocardiography, should be performed regularly [1].

Tumor resection is the commonest surgical treatment of mitral valve myxoma. Mitral valve defect repair or annuloplasty are sometimes necessary in patients whose mitral functions are normal. In those patients whose mitral leaflet cannot be preserved, the mitral valve should be replaced. With the development of minimally invasive surgical techniques, mitral valve myxoma can be resected endoscopically with small incisions.

Conclusions

In conclusion, two- and three-dimensional echocardiography and magnetic resonance imaging are the major diagnostic tools for the diagnosis of a cardiac myxoma. After diagnosis, surgery should be performed urgently, in order to prevent complications such as embolic events or obstruction of the mitral orifice. Due to the fact that myxomas can recur, regular, postoperative cardiological control is mandatory.

Conflict of interest: none declared

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