

A Mitral Valve Myxoma: Case Report

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Abstract

Myxoma is the most common type of primary tumors of the heart. It is a rare tumor with an estimated incidence of 0.5 per million population per year. Most cardiac myxomas originate in the atrial septum in the area of the fossa ovalis and 75% occur in the left atrium. Valvular myxomas are very rare: the tricuspid valve is the most frequently involved location, followed by the mitral, pulmonic, and aortic valves.

Our case was a 24-year-old male with mitral valve myxoma and a history of progressive dyspnea. After diagnosis was confirmed by 2-dimensional echocardiography, he underwent cardiac surgery, the tumor was successfully excised from the posterior leaflet of the mitral valve, and the mitral valve defect was repaired with direct suture (*Iranian Heart Journal 2006; 7 (2):44-47*).

Key words: myxoma ■ mitral valve ■ 2D echocardiography

Myxoma is the most common type of primary tumor of the heart.^{7,8} It is a rare tumor with an estimated incidence of 0.5 per million population per year.⁹ Most of the myxomas are located in the left atrium (75% to 80%), specifically the interatrial septum (IAS) at the fossa ovalis.^{10,12} Right ventricular myxomas are extremely rare, representing 5% of all myxomas.¹¹ Cardiac myxoma can produce a wide spectrum of systemic findings including systemic and pulmonary embolism, and intracardiac obstruction.^{13,14} Mobile and/or pedunculated left atrial myxomas may prolapse to various degrees into the mitral valve orifice, infrequently resulting in the obstruction of AV blood flow and mitral regurgitation. Therefore, the signs and symptoms of myxomas often mimic those of mitral valve disease.¹⁵ Echocardiography is a sensitive and noninvasive method for diagnosis of this tumor.¹⁶

In this report, we describe a patient who had a mobile myxoma of the mitral valve.

Case Report

A 24-year-old male patient was admitted to the hospital with a history of progressive dyspnea for the previous 6 months. He had exertional dyspnea at first, but it gradually worsened and progressed to resting dyspnea. Recently, he had paroxysmal nocturnal dyspnea and loss of appetite and weight.

On physical examination, there was a loud S1, apical tumor plop, and a grade II apical systolic murmur, followed by a diastolic rumble. In the lungs, fine crackles were detected.

On admission, his blood pressure was 130/80 mmHg.

An ECG recording showed normal sinus rhythm and nonspecific ST-T wave changes in precordial leads.

A transthoracic echocardiogram showed a mobile mass with regular contours and homogenous appearance, which measured 4×5 cm and was attached to the posterior

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leaflet of the mitral valve and chordae tendinae of the left ventricle (Fig.1).



Fig. 1. 2-Dimensional echocardiography showing a mobile mitral valve myxoma.

In M-mode echocardiography, echo-free space was not observed on the surface of the mitral valve (Fig. 2).

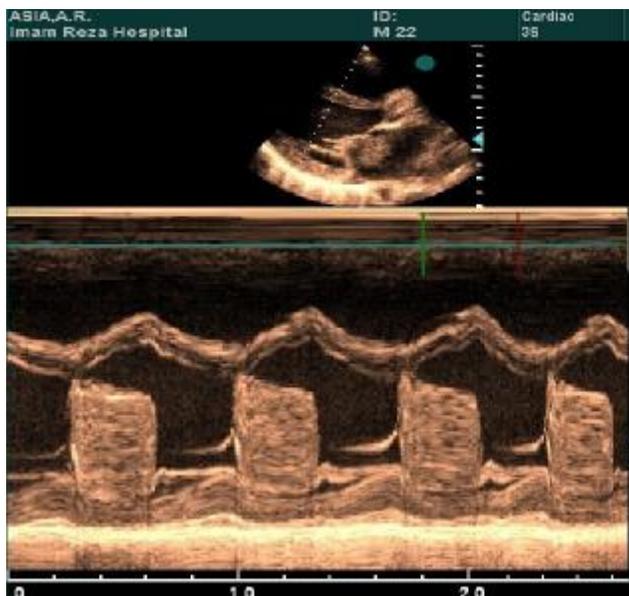


Fig. 2. M-mode echocardiography showing surface of mitral valve without echo-free space.

All the patient's blood test results were normal. He did not have any symptoms or signs of the familial type of myxomas. Chest X-ray showed normal cardiothoracic ratio, and cardiac silhouette was normal with bilateral hilar engorgement.

At surgery, there was a large, white and non-pedunculated tumor measuring 5×6cm, which was attached to the posterior portion of the mitral valve ring and leaflet. The mass was successfully excised from the posterior leaflet of the mitral valve, and the defect was closed with direct suture. Histological study of the mass confirmed a benign myxoma with lax connective tissue with myxomatous cells together with endothelial cells and macrophages. The patient made an uneventful recovery in the postoperative period, and he is currently well after surgery.

Discussion

Primary tumors of the heart are rare. Of these, myxomas are the most common.^{7, 8} Most myxomas are sessile, and the weight of resected specimens ranges from 5 to 125g (mean 40g).²⁰

Cardiac myxomas usually originate in the atrial septum in the area of the fossa ovalis, and approximately 75% occur in the left atrium.²⁴ Valvular myxomas are very rare. The tricuspid valve is the most frequently involved location, followed by the mitral, pulmonic, and aortic valves.^{6, 21}

This is an exceedingly rare localization of myxomas. Kulshrestha et al.¹ reported a 50-year-old woman with a history of shortness of breath and angina. Leude et al.² published two cases of mitral valve myxoma.

Toda et al.³ reported myxoma of the mitral valve in a 20-year-old man who was hospitalized for persistent fever, embolism, and syncopal attacks.

Murphy et al.⁴ described the case of an asymptomatic 49-year-old woman who was found to have a 3.6 x 4.0cm myxoma originating from the atrial side of the anterior mitral leaflet. Handke et al.⁵ reported a 39-

year-old patient who had left-sided hemiparesis, and Chakfe⁶ reported the case of a 44-year-old man with mitral valve myxoma, which is the 21st case of mitral myxoma reported in the western literature.

Among the cases reported in the literature, the diagnosis was made at the time of autopsy in 6 cases, with premortem heart failure in 1 case. A clinical diagnosis was made in 15 cases on the basis of the following symptoms: peripheral embolism in 9 cases, cardiac signs in 4 cases, and constitutional manifestations in 2 cases. The clinical presentation of mitral valve myxoma differs slightly from that of other cardiac myxomas in that it has a lower incidence of constitutional manifestations.⁶

The majority of cardiac myxomas may produce symptoms and signs of heart disease. The clinical manifestations depend on their size and location; dyspnea is the most common manifestation in up to 80% of patients. Patients can also present with other cardiac symptoms such as chest pain, palpitation, and syncope.^{13,14}

The most frequent clinical manifestations of myxoma are unspecific systemic symptoms resulting from the tumoral production of interleukin-6, which occur in 65% of the cases.²³

Cardiac myxoma may be a source of emboli to the central nervous system and elsewhere in the vascular tree. Embolization, one of the critical complications of myxoma, occurs in about 30% to 40% of the patients. The site of embolism is dependent upon the location and the presence of an intracardiac shunt.¹⁵ In most cases, cardiac tumors have thrombi on their surfaces, and both thrombi and necrotic tumoral particles may cause systemic and pulmonary embolization. The neurological signs or symptoms may occasionally be the first or only clinical manifestations of a cardiac tumor.¹⁴ An embolic stroke without evidence of cerebrovascular disease, particularly in the presence of sinus rhythm, should raise the suspicion of cardiac myxoma. Two-dimensional echocardiography can be used to determine the location, size, shape,

attachment, and mobility of the myxoma. Transesophageal echocardiography can be particularly helpful in detecting the site of insertion and morphologic features of ventricular myxomas.¹⁸

Once the diagnosis of a cardiac myxoma has been made, surgical excision should be performed without delay due to the constant threat of embolic episodes or occlusion of the valve orifice.²¹ Removal of adequate margins has been advocated to eliminate the risk of recurrence.

Recurrence of sporadic cases of myxoma (not familiar) is uncommon and occurs in 1 to 4.7% of the cases.²² Close follow-up of the patients following excision with two-dimensional echocardiography is recommended unanimously.¹⁷ Most of the studies have reported that excision of such tumors is curative and long-term out-come is excellent. In this case, follow-up of the patient at two to 15 months after surgery did not show any signs of recurrence.

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