

Pediatric Left Atrial Myxoma: Surgical Excision and Mitral Valve Plasty

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This report describes the case of a 12-year-old girl with a giant left atrial myxoma who presented with severe mitral regurgitation symptoms. Echocardiography demonstrated a 69×30 mm solid mass in the left atrium (LA), occupying almost the entire mitral orifice. After successful surgical excision of the tumor, concomitant with mitral valve plasty, there was no clinical or echocardiographic recurrence at 12-month follow-up. (Ann Thorac Cardiovasc Surg 2007; 13: 65–7)

Key words: left atrium, mitral valve plasty, myxoma, pediatric

Introduction

Primary heart tumor is uncommon in the pediatric population, with a reported incidence of 0.2%.¹⁾ Only 14.5% of all cardiac tumors occur in patients aged under 16 years.²⁾ Mitral valve plasty is also rare in pediatric patients with left atrial tumors. However, we report on a case of a pediatric patient who successfully underwent a mitral annuloplasty and excision of the tumor.

Case Report

A 12-year-old girl presented with complaints of dyspnea on exertion over the preceding 2 months. Physical examination revealed Levine 2/VI systolic murmur with diastolic rumbling. An electrocardiogram showed sinus rhythm with normal axis and no conduction disturbance. An echocardiograph confirmed the presence of a giant left atrial mass with severe mitral insufficiency. A pedunculated tumor advanced through and obstructed the mitral valve during diastole, and was expelled retrogradely into the left atrium (LA) during systole (Fig. 1). Mitral inflow velocity was only 1 m/s despite the pres-

ence of the huge tumor. Preoperative magnetic resonance imaging (MRI) demonstrated no evidence of brain infarction.

Through a median sternotomy incision under cardiopulmonary bypass, the right side of the LA was explored. A mass was noted to be growing within the LA. This giant mass was completely wedging on the mitral orifice leaving the endocardium intact. The “neck” of the mass was located at the fossa ovalis. A 69×30 mm soft and gelatinous mass was successfully removed with excision of a full-thickness portion of the interatrial septum, including the fossa ovalis (Fig. 2). The atrial septal defect was closed with 5-0 prolene suture. The mitral valve annulus was found to be significantly dilated, and a water-leak test confirmed massive mitral regurgitation from the center of the mitral ring. The remainder of the mitral valve, including the 2 leaflets, chordae, and papillary muscles, appeared to be intact. The mitral valve annulus was reduced from 30 mm (140% of normal value) to 21 mm using the Kay-Reed method. Histologic examination showed polygonal, stellate cells with an abundant acid mucopolysaccharide-rich matrix consistent with a benign myxoma (Fig. 3). There were no postoperative complications. At the 12-month follow-up, 2-dimensional echocardiography showed a trivial mitral insufficiency, however, the patient has been completely asymptomatic without arrhythmia or any evidence of tumor recurrence.

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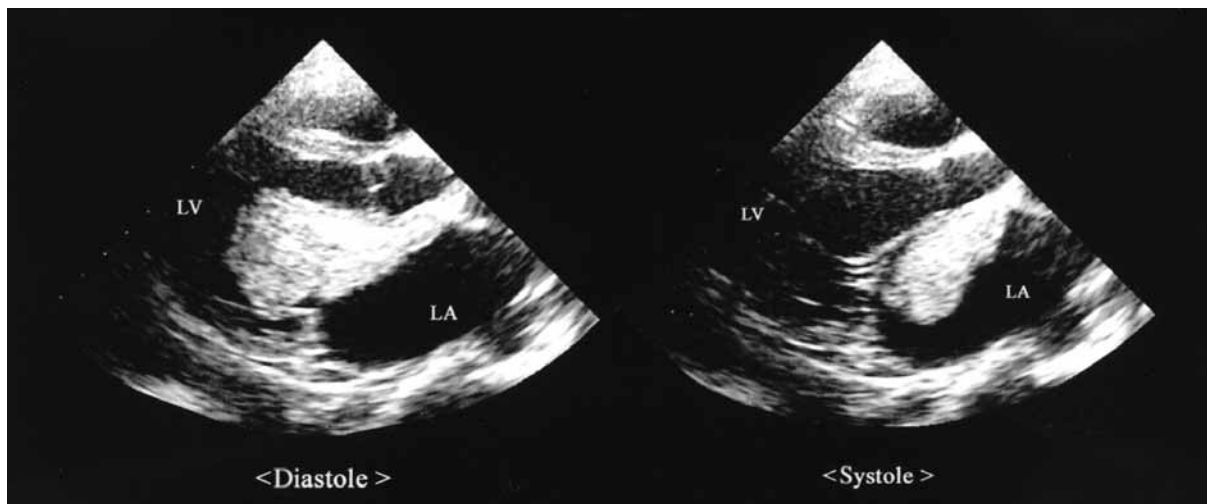


Fig. 1. A left atrial tumor advanced through and obstructed the mitral valve during diastole, and was expelled retrogradely into the left atrium during systole.
LV, left ventricular; LA, left atrium.

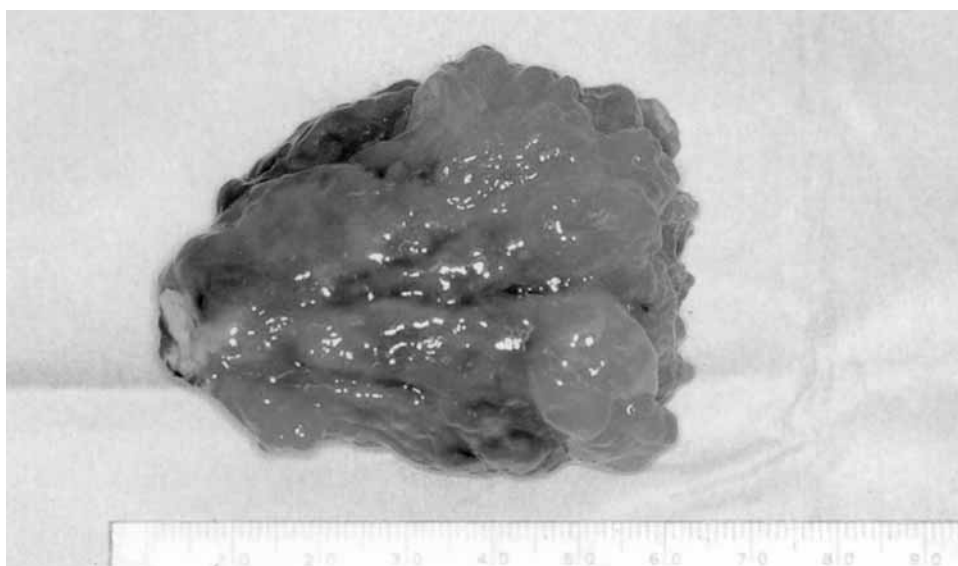


Fig. 2. A 69×30 mm soft and gelatinous mass was successfully removed.

Comment

Myxomas constitute approximately 50% of primary cardiac tumors in patients of all ages.³⁾ However, in pediatric patients, they are less common than rhabdomyomas and fibromas.⁴⁾ Myxomas occur in all regions of the heart, and may result in compression of cardiac structures, valvular insufficiency, outflow tract obstruction, coronary emboli, and occasionally sudden death. Although mitral insufficiency is common in cases with left

atrial myxoma, it can be managed without surgical repair in many instances in adult patients.⁵⁾ Conversely, in children, little has been reported about myxoma-related mitral surgery. As all of the valvular apparatus was normal except for annular dilatation, an annuloplasty was the only available procedure for treating our patient's mitral insufficiency. According to a previous report that mitral insufficiency due to annular dilatation is reversible on long-term follow-up.⁵⁾ Thus, although our most recent echocardiography showed "trivial" regurgitation,

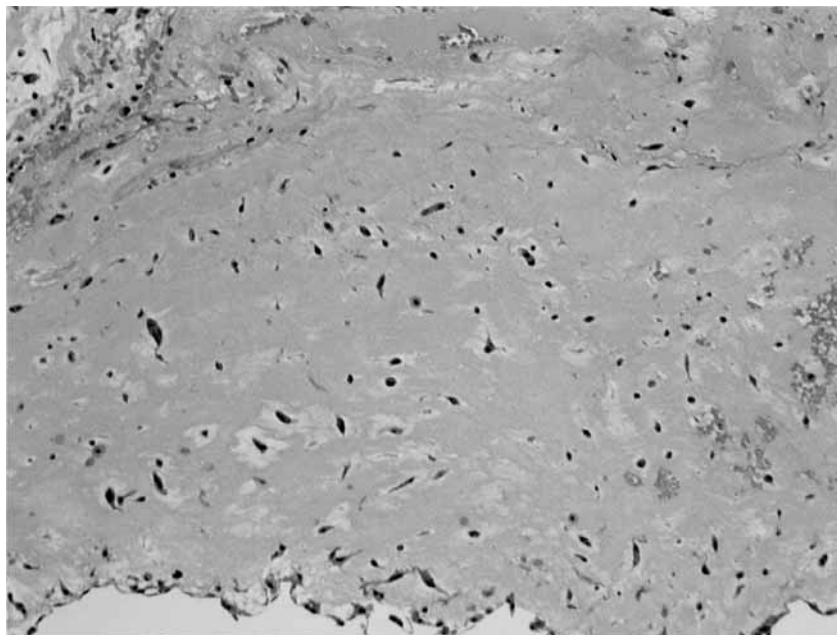


Fig. 3. Histologic examination showed polygonal, stellate cells with an abundant acid mucopolysaccharide-rich matrix consistent with a benign myxoma.

further follow-up is mandatory.

Myxoma can recur in the atrium despite radical resection of the septum.⁶⁾ It is generally believed that the multigrowth potential of the tumor seems to be more important than inadequate surgical resection, as a determinant of recurrence.⁷⁾ Fortunately in this patient, careful inspection during surgery confirmed no other tumor presence. Because the reported time interval between the initial excision and reoperation ranges from 6 months to 12 years,⁸⁾ periodic echocardiographic examinations will be necessary to monitor for recurring myxoma after resection.

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