

Accessory mitral valve causing left ventricular outflow tract obstruction

Shengli Jiang, Tao Zhang, Wei Sheng, Changqing Gao

Department of Cardiovascular Surgery, Chinese PLA General Hospital, Chinese PLA Cardiac Surgery Institute, Beijing, China

ABSTRACT

We studied the clinical characteristics and operative treatment of left ventricular outflow tract obstruction (LVOTO) caused by a congenital accessory mitral valve (AMV). Two patients were admitted to our department. Preoperatively, case 1 was diagnosed as congenital heart disease with severe LVOTO and an anterior mitral valve cleft. The patient in case 2 had a congenital atrial septal defect combined with AMV and mild LVOTO, as well as mild mitral valve regurgitation. In case 1, LVOTO was caused by a type I (fixed) AMV. In case 2, the AMV was type II (mobile type). Both AMV were resected, and the concomitant cardiac disorders were treated simultaneously. The operations were successful, and the LVOTO almost disappeared. Patients with LVOTO caused by AMV should undergo operation for removal of the accessory valve. These patients should be followed up and observed periodically by Doppler echocardiography to identify any aggravation of the LVOTO.

INTRODUCTION

Accessory mitral valve (AMV) is a rare congenital cardiac anomaly, and left ventricular outflow tract obstruction (LVOTO) caused by an AMV is even more rare [Edwards 1965]. We admitted 2 patients with such a deformity, and both patients underwent successful surgical correction.

CLINICAL DATA AND METHODS

Case 1 was of a 7-year-old boy who was admitted after experiencing shortness of breath after strenuous activities in the previous half year. A physical examination revealed a blood pressure of 90/60 mm Hg, a trembling that could be felt on the left side of the sternum, and a coarse systolic murmur that was transmitted to the precordium and the nape. An echocardiography evaluation suggested severe subaortic stenosis with a pressure gradient of 160 mm Hg and mild incompetence due to an anterior mitral valve cleft. Cardiac catheterization and left ventricular visualization showed 2 dome-like filling

defects under the aortic valve. The preoperative diagnosis was congenital LVOTO. Case 2 was of a 16-year-old boy with a diagnosis of a congenital atrial septal defect (ASD). The echocardiography examination also revealed a floating object in the left ventricle that was connected to the anterior mitral valve leaflet with chordae-like tissue. The systolic pressure gradient in the left ventricle was 25 mm Hg, which was combined with mild to moderate mitral valve incompetence. The preoperative diagnosis was congenital AMV and mitral valve incompetence.

RESULTS

Operations were performed with the aid of cardiopulmonary bypass. The ascending aorta in case 1 was incised transversely. A soft white fibrous tissue was found on the ventricular surface of the anterior mitral valve leaf. Its distal end was attached to the anterior mitral valve chordae and the interventricular septum by a chord-like structure and a small papillary muscle, respectively. To obtain a better view of the AMV and its relationship to the mitral valve and other structures, we incised the right atrium and interatrial septum. Then, the AMV was resected through this approach. The defect on the anterior mitral valve, measuring 2 × 2.5 cm, was repaired with interrupted suture, and the results of the water-flooding test of the mitral valve were satisfactory. Other procedures were performed as usual. In case 2, a right atrial incision was made. The AMV was attached to the base of the anterior mitral valve via a pedicle, which appeared as a white membrane-like structure, approximately 1 × 2 cm in size. The AMV was removed directly along the base of the pedicle. The water-flooding test showed no mitral valve incompetence, and the ASD was closed with a pericardium patch.

The procedures of the 2 operations were carried out in a satisfactory manner. Pathologic examinations of the excised AMV revealed fibrous tissue with mucoid degeneration in case 1 and fibrous tissue with inflammatory cell infiltration in case 2. A postoperative echocardiography evaluation in case 1 showed that the subaortic stenosis had been relieved, with a pressure gradient of 15 mm Hg in the LVOT. In case 2, the ASD was closed completely without a pressure gradient in the LVOT, although very mild mitral valve regurgitation was present. The patients were discharged with full recovery. They were followed up postoperatively for 5 to 6 years, and the cardiac functions of the 2 patients were in the normal range.

Received December 8, 2009; accepted February 2, 2010.

Correspondence: Shengli Jiang, MD, Department of Cardiovascular Surgery, Chinese PLA General Hospital, No. 28 Fu Xing Ave, Beijing, 100853, P. R. China; 86-10-66938236; fax: 86-10-88211280 (e-mail: jiangsl301@sina.com).

DISCUSSION

AMV patients are usually asymptomatic in the beginning, with only a cardiac murmur to be heard. LVOTO begins to present when patients are approximately 10 years old [Prifti 2001]. Some patients may have severe aortic incompetence due to prolapse of the AMV into the aortic valve during the diastolic phase [Sono 1988]. A few patients with a serious LVOTO may need urgent operation in the neonatal period [Meyer-Hetling 2000]. Some patients have presented with ventricular arrhythmia, which may be related to the injury to the conductive tissue caused by the occurrence of an AMV beneath the aortic valve or by increasing tension of the ventricular wall [Sono 1988]. The LVOT stenosis is usually severe, with a pressure gradient of >50 mm Hg [Yoshimura 2000]. The patient in case 1 was in this category. Some investigators have reported that an AMV was not necessarily complicated by LVOT stenosis or that the stenosis was moderate. Case 2 resembled this condition. The effect of an AMV on the LVOT, however, may gradually become aggravated as the patient ages, and then the LVOTO might appear [Garrett 1990]. In patients who also have a ventricular septal defect or mitral valve incompetence, the degree to which the AMV affects the LVOTO may be underestimated [Kanemoto 1992].

AMV is usually misdiagnosed, because congenital AMV is uncommon and usually combined with other cardiac anomalies. An echocardiogram can detect the systolic forward movement of the anterior mitral valve or the pedicled floating tissue connected to the anterior mitral valve in the LVOT and thereby suggest the existence of this disease [Ascuitto 1986]. Cardiac catheterization or angiography may be helpful for visualizing the LVOT and measuring the pressure gradient between the left ventricle and the aorta. A differential diagnosis should be made when there is valvular vegetation or some kind of left ventricular tumor, which can resemble AMV in the image. Case 1 was not diagnosed as AMV with a preoperative echocardiogram and cardiac angiography because of rarity of this kind of disease, and case 2 was correctly diagnosed because of our previous experience with the disease.

AMV is divided into 2 pathologic types [Sono 1988; Prifti 2001]. The fixed type (type I) is characterized by a direct connection of the AMV to the mitral valve via either a nodular or membranous connection. In the mobile type (type II), the

AMV is connected to the anterior mitral valve via a fibrous rope-like tissue that is either pedunculated or leaflet-like and floating in the LVOT, producing the LVOTO. Case 1 was the fixed type of AMV, which was connected to the anterior mitral valve under the noncoronary aortic valve. Case 2 belonged to the mobile type.

The principle of the operative procedure not only involves removal of the AMV tissue to relieve the subaortic stenosis but also requires care to avoid inadvertent damage to the mitral valve that could lead to mitral valve incompetence. The surgical results of relieving the LVOTO due to an AMV are satisfactory. The operation should not be performed in patients without the outflow tract stenosis or in patients with mild stenosis without combination with other cardiac anomalies. Patients should be followed up to guard against aggravation of the LVOTO.

REFERENCES

- Ascuitto RJ, Ross-Ascuitto NT, Kopf GS, Kleinman CS, Tälner NS. 1986. Accessory mitral valve tissue causing left ventricular outflow obstruction (two-dimensional echocardiographic diagnosis and surgical approach). *Ann Thorac Surg* 42:581-4.
- Edwards JE. 1965. Pathology of left ventricular outflow tract obstruction. *Circulation* 31:586-99.
- Garrett HE Jr, Spray TL. 1990. Accessory mitral valve tissue: an increasingly recognized cause of left ventricular outflow tract obstruction. *J Cardiovasc Surg* 31:225-30.
- Kanemoto N, Shiina Y, Goto Y, et al. 1992. A case of accessory mitral valve leaflet associated with solitary mitral cleft. *Clin Cardiol* 15:699-701.
- Meyer-Hetling K, Alexi-Meskishvili VV, Dähnert I. 2000. Critical subaortic stenosis in a newborn caused by accessory mitral valve tissue. *Ann Thorac Surg* 69:1934-7.
- Prifti E, Frati G, Bonacchi M, Vanini V, Chauvaud S. 2001. Accessory mitral valve tissue causing left ventricular outflow tract obstruction: case report and literature review. *J Heart Valve Dis* 10:774-8.
- Sono J, McKay R, Arnold RM. 1988. Accessory mitral valve leaflet causing aortic regurgitation and left ventricular outflow tract obstruction. *Br Heart J* 59:491-7.
- Yoshimura N, Yamaguchi M, Oshima Y, et al. 2000. Clinical and pathological features of accessory valve tissue. *Ann Thorac Surg* 69:1205-8.