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Case Report
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Latent Left Ventricular Outflow Tract Obstruction Due to Accessory Mitral Valve in a 12-Year-Old Boy

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Accessory mitral valve (AMV) is a rare congenital anomaly which can cause left ventricular outflow tract (LVOT) obstruction. Patients with isolated AMV usually present with exertional dyspnea, chest pain, or syncope during the first ten years of life. In patients with AMV, detection of latent LVOT obstruction can be clinically challenging. We here present a case of AMV causing latent LVOT obstruction in an adolescent, which was diagnosed by echocardiography with dobutamine provocation and finally treated by successful surgical resection.

Key Words: Congenital heart disease • Left ventricular outflow tract obstruction

Accessory mitral valve (AMV) is a rare congenital anomaly which may cause left ventricular outflow tract (LVOT) obstruction. The first surgical treatment for this lesion was described in 1963.[1] About 2/3 of those patients with AMV have associated congenital cardiovascular anomalies such as ventricular septal defects, patent ductus arteriosus and transposition of the great arteries.[2-4] Patients with isolated AMV usually present with exertional dyspnea, chest pain, or syncope during the first decade of life. However, detection of an asymptomatic patient without LVOT obstruction is uncommon.[4-6] We here present a case of AMV causing latent LVOT obstruction in adolescence, who was diagnosed by echocardiography with dobutamine provocation and successfully treated by surgical resection.

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CASE REPORT

A 12-year-old male was referred to our facility with exertional dyspnea and chest discomfort upon exertion. Physical examination revealed systolic ejection murmur of grade II/VI at the left parasternal border. Two-dimensional transthoracic echocardiography (TTE) showed a tiny, round echogenic structure attached to the basal interventricular septum in diastole (Figure 1A). This structure moved like a spreading parachute into the LVOT during systole, and was receiving several chordal insertion from the papillary muscle (Figure 1B, C). Color Doppler imaging revealed flow acceleration at the level of LVOT (Figure 1D), but peak and mean pressure gradient at rest was only 23 and 10 mmHg, respectively (Figure 1E). To determine the hemodynamic significance of the structure, dobutamine stress echocardiography was performed. At an infusion rate of 10 mcg/kg/min, peak and mean pressure gradient across the LVOT increased to 67 and 32 mmHg, respectively (Figure 1F). In the meantime, the patient complained of dyspnea and chest discomfort as usually experienced on exertion. Three-dimensional echocardiography provided images showing the mechanism of obstruction related to the displacement of the structure in LVOT and its attachment point from the interventricular septum to the lateral portion of the anterior mitral leaflet (Figure 1G, H). After re-

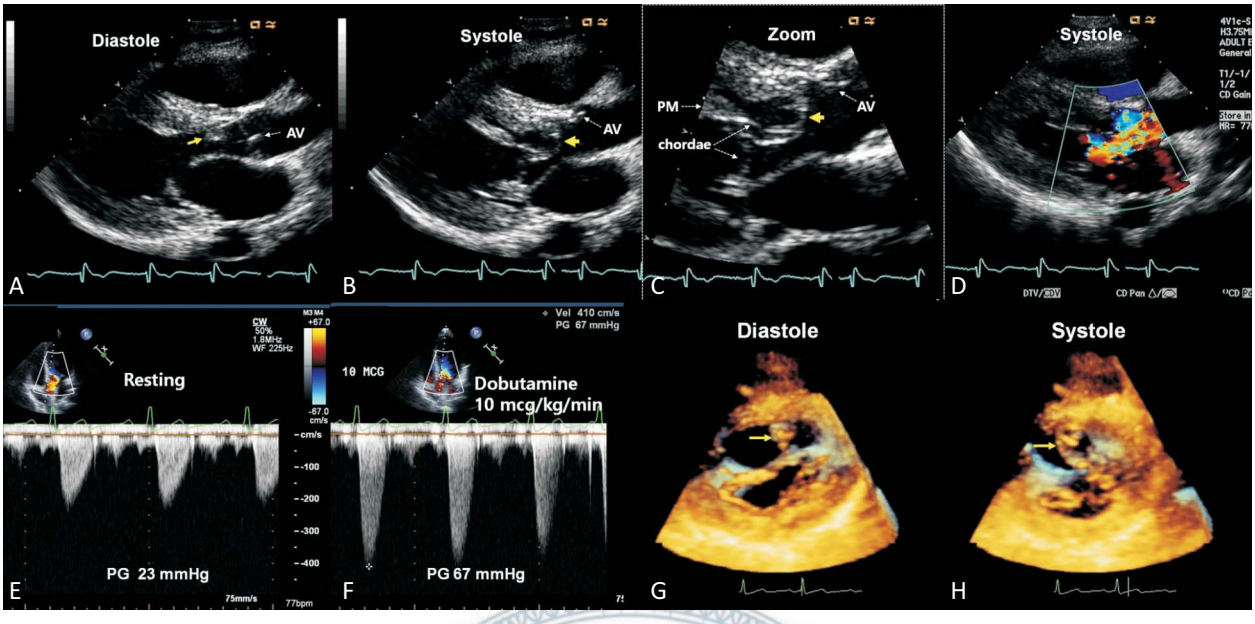

Figure 1. *(A) Single diastolic frame in parasternal long axis view of transthoracic echocardiography showing folded accessory mitral valve (single arrow) attached to anterior interventricular septum in LVOT. (B-C) Single systolic frame in parasternal long axis view of transthoracic echocardiography demonstrating accessory mitral valve spreading like a parachute into LVOT (single arrow) and several chordal insertions on it from the papillary muscle. (D) Color Doppler imaging showing accelerated flow signal at the level of LVOT. (E-F) Dobutamine stress echocardiography demonstrating an increased peak and mean pressure gradient across LVOT from 23 and 10 mm Hg at rest to 67 and 32 mmHg after dobutamine infusion at 10 mcg/kg/min, respectively. (G) Single diastolic frame in parasternal short axis view real-time 3-dimensional echocardiography showing the structure attached to the lateral portion of anterior mitral leaflet (viewed from the left ventricle side). (H) Single systolic frame in parasternal short axis view of real-time 3-dimensional echocardiography showing the structure unfolded and displaced into LVOT, which may explain the mechanism of obstruction (viewed from the left ventricle side). AV, aortic valve; LVOT, left ventricular outflow tract; PM, papillary muscle.*

viewing all the echocardiographic findings, we concluded that he had AMV causing latent LVOT obstruction. The patient was referred for surgical resection of AMV.

We examined the patient's aortic tissue after transverse aortotomy, and found a thin nature, parachute-like valvular tissue in LVOT (Figure 2A). It was attached to the anterior interventricular septum just below the level of aortic annulus and lateral portion of the anterior mitral leaflet. Therefore, several chordae were directly inserted onto the AMV instead of the lateral portion of anterior mitral leaflet as shown in TTE. The accessory mitral valve was successfully resected (Figure 2B), but intraoperative transesophageal echocardiography (TEE) revealed significant mitral regurgitation due to severe prolapse of the lateral portion of the anterior mitral leaflet. Left atriotomy was performed and new chordae were implanted to the lateral portion of the anterior mitral leaflet. Intraoperative TEE was performed after the second cardiopulmonary bypass which indicated there was no residual mitral regurgitation. On post-operative TTE, LVOT obstruction was relieved. Histopathologic examination revealed normal mitral valve tissue with degeneration (Figure 2C). Thereafter, the patient remained free of symptoms after 5 years of follow-up.

DISCUSSION

AMV causing LVOT obstruction was first described in the echocardiography literature by Cooperberg et al. in 1976.[7] Echocardiography can provide important information about the morphology, motility, attaching site and hemodynamic significance of AMV.[8,9] Though transthoracic echocardiography is usually sufficient to visualize AMV, transesophageal echocardiography and three-dimensional echocardiography may provide additional information to better delineate the associated structures with greater accuracy. Surgical excision of isolated AMV is required if there is significant LVOT obstruc-

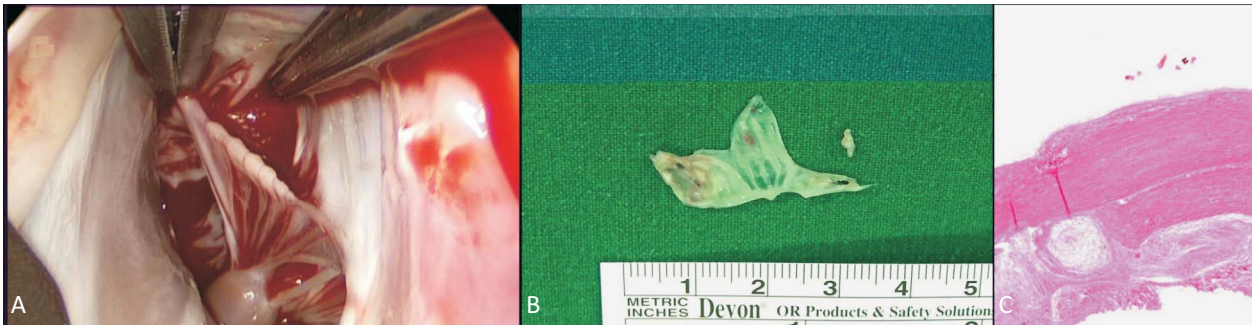

Figure 2. *(A) Inspection through transverse aortotomy showing thin nature, parachute-like accessory mitral valve with multiple chordal insertion and connections to interventricular septum and lateral portion of anterior mitral leaflet. (B) Gross pathologic specimen showing excised AMV looks like normal mitral valve tissue. (C) Hematoxylin-eosin stain of histopathologic specimen from removed mass at low power demonstrating normal mitral valve tissue with degeneration.*

tion.[10] In a symptomatic patient without obvious LVOT obstruction at rest, dobutamine or exercise stress echocardiography to provoke LVOT obstruction may facilitate the decision for surgical intervention.

Prifti et al. classified AMV into type I (fixed: A = nodular, B = membranous), and type II (mobile: A = pedunculated, B = leaflet-like).[2,3] The most common type of AMV is IIB, which may have rudimentary or well-developed chordae tendineae. In type IIB with well-developed chordae tendinae like our case, reimplantation of chordae as well as resection of AMV should be performed.

In conclusion, AMV should be considered in the differential diagnosis of LVOT obstruction, especially in young patients with dyspnea and systolic murmur. Echocardiography is very useful in both diagnosing and planning the medical management of this uncommon congenital anomaly. In addition, the use of a provocation test such as dobutamine stress echocardiography in case of latent LVOT obstruction may facilitate the process of surgical decisionmaking.

REFERENCES

1. MacLean L, Culligan J, Kane D. Subaortic stenosis due to accessory tissue on the mitral valve. *J Thorac Cardiovasc Surg* 1963;45:382-7.
2. Prifti E, Frati G, Bonacchi M, et al. Accessory mitral valve tissue causing left ventricular outflow tract obstruction: case reports and literature review. *J Heart Valve Dis* 2001;10:774-8.
3. Prifti E, Bonacchi M, Bartolozzi F, et al. Postoperative outcome in patients with accessory mitral valve tissue. *Med Sci Monit* 2003;9:RA126-33.
4. Rovner A, Thanigaraj S, Perez JE. Accessory mitral valve in an adult population: the role of echocardiography in diagnosis and management. *J Am Soc Echocardiogr* 2005;18:494-8.
5. Uysal OK, Duran M, Ozkan B, et al. Asymptomatic accessory mitral valve tissue diagnosed by echocardiography. *Korean Circ J* 2012;42:800.
6. Uslu N, Gorgulu S, Yildirim A, Eren M. Accessory mitral valve tissue: report of two asymptomatic cases. *Cardiology* 2006;105:155-7.
7. Cooperberg P, Hazell S, Ashmore PG. Parachute accessory anterior mitral valve leaflet causing left ventricular outflow tract obstruction. Report of a case with emphasis on the echocardiographic findings. *Circulation* 1976;53:908-11.
8. Ow EP, DeLeon SY, Freeman JE, et al. Recognition and management of accessory mitral tissue causing severe subaortic stenosis. *Ann Thorac Surg* 1994;57:952-5.
9. Popescu BA, Ghiorghiu I, Apetrei E, Ginghina C. Subaortic stenosis produced by an accessory mitral valve: the role of echocardiography. *Echocardiography* 2005;22:39-41.
10. Yuan SM, Shinfeld A, Mishaly D, et al. Accessory mitral valve tissue: a case report and an updated review of literature. *J Card Surg* 2008;23:769-72.