Ann Thorac Cardiovasc Surg 2014; Supplement: 742–745
Online November 8, 2013
doi: 10.5761/atcs.cr.13-00109

Case Report

Accessory Mitral Valve: A Rare Cause of Asymptomatic Severe Left Ventricular Outflow Tract Obstruction

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Accessory mitral valve (AMV) is an extremely uncommon congenital cardiac anomaly and it is a rare cause of left ventricular outflow tract (LVOT) obstruction. Identification of asymptomatic patients with LVOT obstruction is uncommon since the symptoms usually become manifest when the gradient across the LVOT rises. We describe a rare case of asymptomatic AMV with severe LVOT obstruction in a 10-year old child with no other congenital or acquired cardiac defects who successful underwent surgery. This case emphasizes the importance of early surgical indication also in absence of symptoms when significant LVOT obstruction occurs.

Keywords: mitral valve, mitral valve surgery, accessory mitral valve, left ventricular outflow tract obstruction

Introduction

Accessory mitral valve (AMV) is an extremely rare congenital cardiac anomaly of embryologic development of the endocardial cushion. It frequently arises from the anterior mitral leaflet[1)] and it is a rare cause of left ventricular outflow tract (LVOT) obstruction.[2)]

Most of the patients are diagnosed with AMV during the first decade of life and this pathology is frequently associated with other congenital cardiac defects.[3,4)] Identification of asymptomatic patients with LVOT obstruction is uncommon since the symptoms usually become manifest when the gradient across the LVOT rises.

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Received: May 9, 2013; Accepted: September 5, 2013
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We describe a rare case of echocardiographic diagnosis of AMV with severe LVOT obstruction in an asymptomatic child with no other associated congenital or acquired heart defects.

Case Report

A grade 2/6 medium harsh systolic ejection murmur, audible on the left side of the sternum at the third and fourth intercostal space and radiating to the neck, was first detected in a 10-year-old boy at sports medical examination. He had no symptoms at rest or during exercise and performed normal child's play activities.

The electrocardiogram (ECG) showed typical left ventricular hypertrophy (high voltages with an ST-T strain pattern). On telecardiogram, the cardiothoracic ratio was in the normal range and no other pathological findings were detected. Other laboratory data were also within normal limits.

Transthoracic echocardiography (TTE) revealed mild hypertrophy and an abnormal tissue protruding into the LVOT during systole, less than 1 cm from the aortic cusps, with insertion on the membranous interventricular

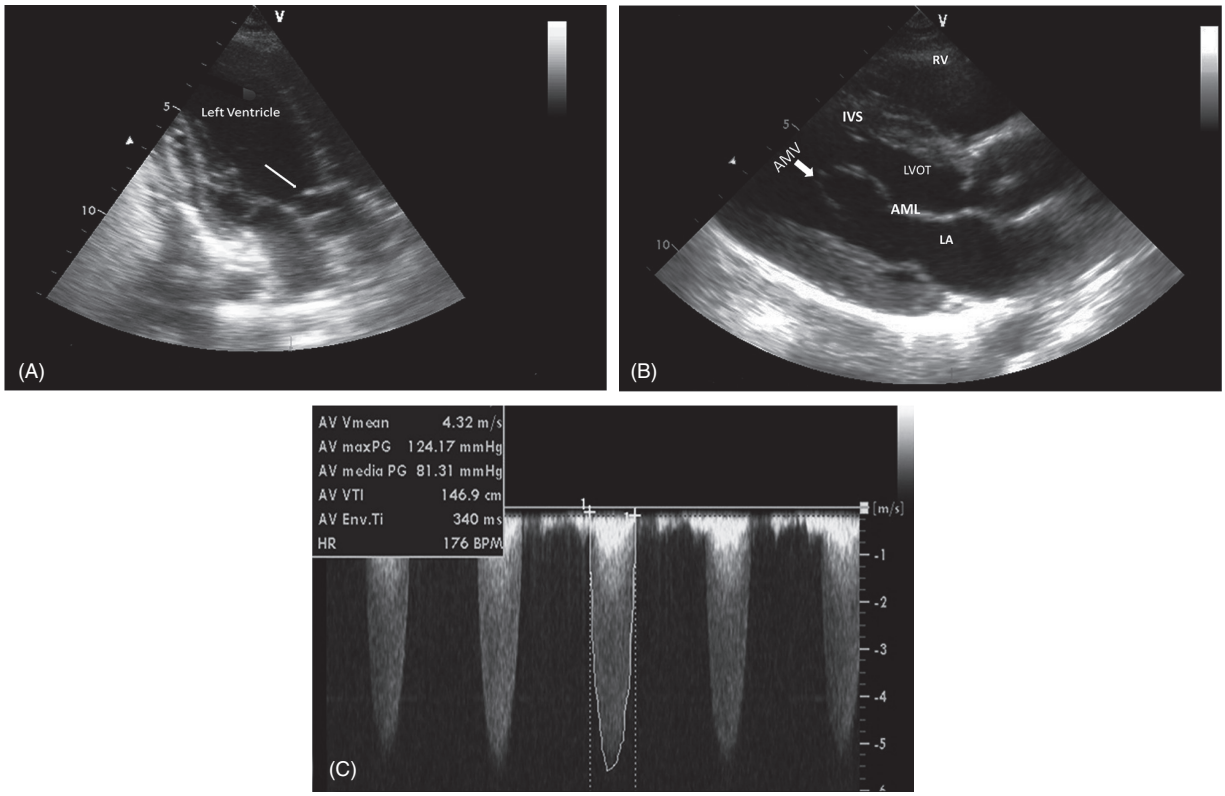

Fig. 1 (**A**) Transthoracic echocardiography, 3-chamber apical view. The arrow indicates accessory mitral valve (AMV) tissue protruding into the left ventricular outflow tract (LVOT). (**B**) Transthoracic echocardiography, parasternal long-axis view: the AMV tissue is visualized as a spherical structure attached to the ventricular surface of the anterior mitral valve leaflet. (**C**) Continuous-wave Doppler echocardiographic examination showing the presence of high pressure gradient (peak, 124 mmHg; mean gradient 81 mmHg) between left ventricle and aorta. RV: Right Ventricle; IVS: Interventricular Septum; LA: left atrium; AML: Anterior Mitral Leaflet; LVOT: left ventricular outflow tract.

septum and in direct contiguity with the anterior leaflet of the mitral valve (**Fig. 1A**).

The free margins of the LAM had also increased size and consistency for the presence of tissue accessory of approximately 2 cm in length which floated free during systole inside the ventricular cavity without compromise of the mitral valve function. This finding of saccular appearance seemed hyper reflective and was distinct from the chordae tendineae that connected the anterior mitral leaflet (**Fig. 1B**). No other cardiac anomalies were present. The peak pressure gradient (PG) across the LVOT, estimated by continuous wave Doppler measurement of peak flow velocity, was about 124 mmHg and the mean gradient was 81 mmHg (**Fig. 1C**).

Transesophageal echocardiography (TEE) was deemed unnecessary because of the young age of the patient and because TTE enabled to fully detect anatomic features and functional significance of AMV.

The patient was referred to surgery. Cardiopulmonary bypass was established by standard aortic cannulation and two-stage venous cannulation of the right atrium. The ascending aorta was opened, the aortic valve inspected and the AMV was observed beneath the non-coronary cusp. The AMV had three separate chordae that were all connected to the anterolateral papillary muscle. Part of the AMV was also attached to the non-coronary cusp as well as the ventricular septum below the non-coronary cusp and the aorto-mitral continuity.

A left atriotomy was carried out and the mitral valve explored. No clefts were found in the anterior mitral leaflet which showed the AMV tissue attached to its left ventricular surface.

Because the AMV did not contribute to mitral valve function, it was excised completely without any destruction of the valve (**Fig. 2**). The accessory chordae were left intact to avoid mitral insufficiency. The hypertrophic

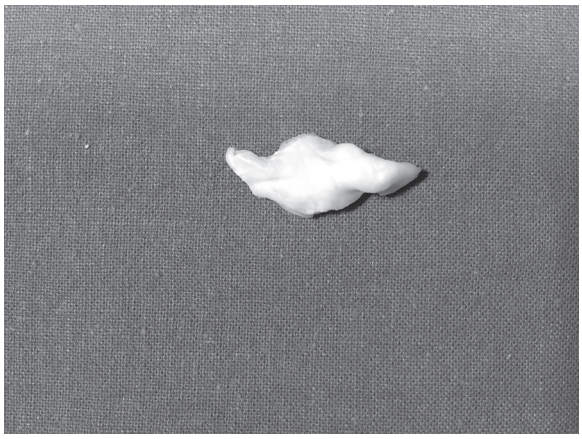

Fig. 2 Gross pathologic specimen of excised accessory tissue that looks like normal mitral valve tissue.

septal tissue was sharply excised. The operation was routinely completed.

The histopathologic examination revealed that this accessory valve was composed of valve tissue with myxoid degeneration.

The patient was discharged after eight days with an uneventful postoperative course. Postoperative echocardiography showed minimal acceleration of flow in the left ventricular outflow tract with maximum pressure gradient of 20 mmHg and medium 10 mmHg.

On 6-month postoperative echocardiographic examination no residual gradient was detected between the left ventricle and the aorta.

Discussion

Accessory mitral valve (AMV) is an extremely rare congenital anomaly which causes subaortic stenosis.[1] This very rare condition is thought to result from abnormal development of endocardial cushion tissue and it is associated with other congenital cardiac defects, such as transposition of the great vessels, ventricular septal defects, bicuspid aortic valve and coarctation of the aorta.[5] Usually AMV is diagnosed in the first decade of life and the most common presentation is an asymptomatic heart murmur, typically seen in patients who do not have significant LVOT obstruction.[6] In contrast, those who do have significant obstruction become symptomatic when the mean gradient across the LVOT is more than 50 mmHg[7] with exertional dyspnea, chest pain, syncope and heart failure.[1]

We describe the uncommon presentation of AMV in an asymptomatic child with evidence of a significant degree of LVOT obstruction. Cases of isolated AMV, such as ours—in an asymptomatic young with a high LVOT gradient and no associated congenital lesions—are rare. Surgery is currently indicated in symptomatic patients and in patients undergoing corrections of other congenital malformations, in those with severe valvular regurgitation, in subjects showing any complication associated with this accessory tissue or undergoing exploration of an intracardiac mass.[2]

In contrast, it is unclear as to whether intervention should be performed in patients with mild or no symptoms, because the outcome of minimally symptomatic patients with AMV without any other complex congenital/acquired cardiac anomaly and with significant LVOT obstruction has not been well delineated.

Our approach is to operate on patients with AMV tissue if a significant LVOT obstruction is present even without associated symptoms. Recent analyses confirm that obstruction is associated with poorer survival and high LVOT gradient is a well-recognized cause of sudden death. Indeed, AMV, in particular before the widespread use of echocardiography, was frequently an occasional finding at post mortem examination. It must be also taken into consideration that the pressure gradient might increase with aging. Indeed, it has been demonstrated that the pressure gradient across the LVOT may rise by 65 mmHg over a 5-year period[8] and this can be explained by the continuing turbulence produced by AMV into the LVOT which can result in permanent deposits of fibrous tissue and fixed subaortic obstruction.[7] In addition, in asymptomatic cases with LVOT obstruction, surgery should be considered, in our opinion, also because of the likelihood of embolic phenomena which may occur at any time.[3] These embolic events are thought to be related either with AMV tissue itself or to accumulation, within the accessory tissue, of platelets and other debris that may intermittently embolize. Finally, prompt surgery avoids progressive aortic regurgitation which may develop due to persistently turbulent flow patterns in the LVOT that may cause valve damage.[9]

Resection of the accessory tissue and removal of membranous or muscular substrata of obstruction is the standard surgical therapy which allowed, in our experience, for essentially complete relief of LVOT obstruction with no evidence of resultant mitral valve dysfunction.

The greatest problem during surgery is recognizing the anatomical relationship between the accessory tissue and the mitral valve. Therefore, left atriotomy associated to aortotomy is recommended to localize and fully

examine AMV tissue and to ensure that accessory tissue is removed without damaging the mitral valve structures. In addition, complete resection of the accessory tissue is mandatory since the incomplete removal of AMV and the presence of residual LVOT appear to be the main factors affecting mortality.[2] In the presented case, part of the AMV was also attached to the non-coronary cusp as well as the ventricular septum below the non-coronary cusp and the aorto-mitral continuity. However, because the AMV did not contribute to the mitral valve function, it could be excised completely without any destruction of the valve. In addition, AMV had three separate chordae that were connected to the anterolateral papillary muscle. We left intact the accessory chordae because there was a concern that disruption of the native subvalvular mitral apparatus could result in mitral insufficiency.

Echocardiography confirmed to be a reliable and indispensable diagnostic tool for clarifying the nature, morphology, and attachment points of the AMV.[10] Indeed, the intraoperative recognition of the AMV is rarely possible since the location of the attachment of the accessory tissue is usually on the ventricular side and the thin structure is commonly collapsed in the empty and arrested left ventricle on bypass.

Finally, because other types of left ventricular masses, such as vegetations or tumors (myxomas and papillary fibroelastomas) may produce similar echocardiographic findings, AMV should be considered in the differential diagnosis of a cardiac mass. Of note, the origin of these masses can be used for differentiation; tumors often arise from cardiac muscle, and vegetations tend to originate from the build-up in the low-pressure side of the heart valve.

Conclusions

In conclusion, for patients with accessory mitral valve tissue, prompt surgery should be recommended if there is significant obstruction in the left ventricular outflow tract even in the absence of symptoms. Doppler echocardiography may lead to the identification of an AMV before the development of symptoms or important fixed subaortic obstruction.

Acknowledgement

We gratefully acknowledge Dr. Judith Wilson for the English revision of the manuscript.

Disclosure Statement

None to be declared.

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