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Case report

Mitral valve anomalies obstructing left ventricular outflow

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Abstract

This paper reports on two cases of more uncommon types of subaortic stenosis. A 2-year-old boy was found with accessory mitral valve leaflet (AMVL) attaching to the anterior leaflet, ballooning into the subaortic ventricular septum associated with a discrete subaortic membrane. The obstruction was successfully relieved by removal of the AMVL and resection of the membrane. A 19-day-old newborn with accessory tissue on the mitral valve (AMVT) causing subaortic stenosis, subaortic ventricular septal defect (VSD) and patent ductus arteriosus was operated on successfully. Accessory tissue excision through the VSD, VSD patch closure and ductus ligation were performed. © 1997 Elsevier Science B.V.

Keywords: Accessory mitral valve tissue; Accessory mitral valve leaflet; Left ventricular outflow tract obstruction; Subaortic stenosis

1. Introduction

Subvalvular aortic stenosis occurs in 8–20% of cases of left ventricular outflow tract obstruction (LVOTO), rarely presents in infancy, is more often seen in children or young adults. In 1957, Ferencz [5] reported an infant with LVOTO caused by anatomical malformation of the subvalvular apparatus of the mitral valve (MV). In 1964 Sellers et al. [9] published the different types of the abnormalities of the atrioventricular valves producing subaortic stenosis. Since then many excellent necropsy studies, but few clinical reports with accessory MV in the left ventricular outflow tract (LVOT), have been demonstrated [1–4,6–10]. We present two cases with different types of these very rare malformations of the MV.

2. Case 1

A 2-year-old boy with clinical findings of LVOTO was admitted. On echocardiography, both leaflets of the MV were visualized and thought to be normal. The aortic valve was tricuspid. The LVOT was narrowed and an accessory mitral valve leaflet (AMVL) was shown. Doppler gradient was 100 mmHg through the LVOT (Fig. 1A). Cardiac catheterization: a peak systolic gradient of 100 mmHg across the LVOT. The pressure tracing showed a pattern of subaortic stenosis. LV systolic pressure was 220 mmHg. There was no evidence of any other associated lesion. LV angiogram showed well-marked radiolucency in the LVOT on the antero-posterior and diastolic lateral projections. In systole on the lateral projections, the lucent area became filled with contrast medium.

He was operated on 15 December 1992 with cardiopulmonary bypass and cold potassium cardioplegia for myocardial protection. The aorta was opened; the aortic valve was tricuspid and not stenosed. A wide

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membrane of about 2 mm was excised from the subaortic area at the typical site. The major cause of the severe LVOTO was subvalvular, due to the abnormal AMVL. This AMVL formed a parachute-like structure, 1.5 cm long and 1 cm wide, which was attached to a concave area directly below the posterior half of the left coronary leaflet. The free inferior margin of this sac was attached by an 8-mm long chordae, which was anchored on the anterior aspect of the anterolateral papillary muscle. The parachute-like obstructing valve was then excised from the subvalvular subaortic region and from the anterolateral mitral papillary muscle. Histology of the excised tissue revealed a valvule-like structure.

The postoperative course was uneventful. Following the operation the child has grown well, and is vigorous and symptom free (6 December 1996). Recent echocardiography showed a normal-sized left atrium, normal LV contractility, and no aortic or trivial MV insufficiency. Doppler study confirmed the absence of the LVOTO (Fig. 1B).

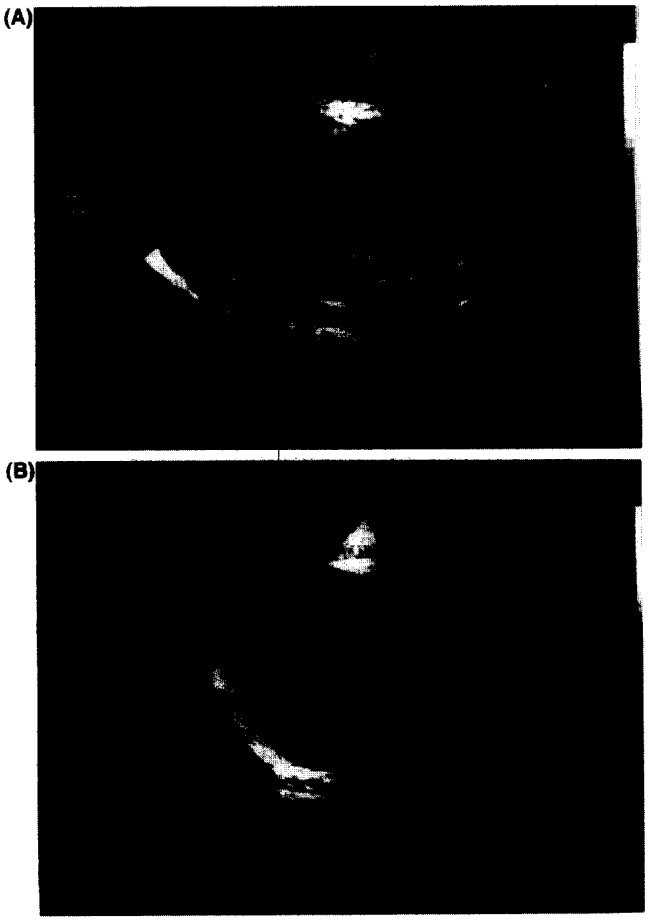

Fig. 1. (A) Preoperative echocardiographic examination: long axis view. The arrow shows the accessory mitral valve leaflet. (B) Postoperative examination: long axis view. Normal left ventricular outflow tract can be seen. LV, left ventricle; RV, right ventricle; Ao, aorta.

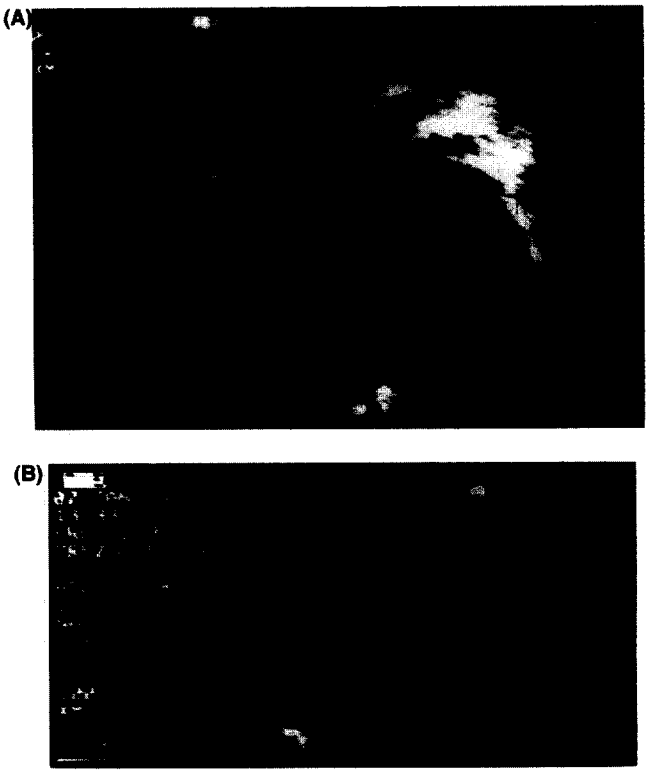

Fig. 2. (A) Preoperative echocardiographic examination: left ventricular long axis view. The arrow indicates the accessory mitral valve tissue in the subaortic area. A large membranous ventricular septal defect with muscular extension can also be seen. (B) Intraoperative epicardial echocardiography using a 7.5-Mhz transducer: an echodense patch closing the VSD can be seen. No abnormal tissue in the subaortic region was observed. Ao, Aorta; LV, left ventricle.

3. Case 2

A 14-day-old newborn with congestive heart failure, and mild dyspnea was admitted on 2 January 1996. Fine rales were heard over both lung fields. Heart rate was 180/min, regular. There was a grade 2/6 protomesosystolic murmur in the third intercostal space, and a long, low-pitched grade 2/6 diastolic murmur at the apex. The liver edge was palpable 1.5 cm below the right costal margin. Thoracic X-ray showed cardiomegaly with prominent left atrial enlargement and pulmonary venous congestion. ECG demonstrated mild LV hypertrophy. On echocardiography, a large membranous ventricular septal defect (VSD) with muscular septal extension was found: accessory tissue on the mitral valve (AMVT), causing nearly total obstruction of the LVOT. Color Doppler showed turbulent flow in the LVOT: maximal aortic instantaneous gradient was 36 mmHg. VSD flow was 1.5 m/s proving pulmonary hypertension. LV function was poor, with mild functional mitral insufficiency. No aortic insufficiency was seen in the tricuspid aortic valve (Fig. 2A).

Because of the mobile mass in the LVOT, emergency operation was indicated.

On 6 January 1996 (at 19 days old, and weighing 2800 g) she underwent operation. Cardiopulmonary bypass with cold cardioplegia was established. The heart was exposed through median sternotomy. The ductus arteriosus was ligated. The right atrium was opened, and the 8-mm subaortic VSD was explored through the tricuspid valve. The 5 × 8-mm accessory tissue was pulled out and excised from the anterior leaflet of the MV through the VSD. (Histological examination demonstrated a valve-like structure.) The VSD was closed with a patch. The intraoperative epicardial echocardiography demonstrated normal LVOT (Fig. 2B).

In the postoperative period, she needed peritoneal dialysis for a short time, and on the 20th day she was transferred to another hospital for treatment of hydrocephalus internus. Ten months following the operation she is fine. Echo/Doppler studies showed no gradient detected in the LVOT, grade I aortic valve insufficiency, and no mitral regurgitation.

4. Discussion

Subvalvular aortic stenosis can be caused by several anatomic malformations, such as subvalvar fibrous membrane, diffuse stenosis on the basis of a fibromuscular tunnel, dynamic stenosis due to hypertrophy of the interventricular muscular septum and, finally, by an abnormality of the MV. The latter type could be due to accessory tissue arising from the atrioventricular (AV) valves or to restricted movement of the anterior MV leaflet because of accessory chordae, or by abnormal fusion of valve tissue to the septal wall of the outflow tract. These types are very rare forms of subaortic stenosis, especially in newborns. To the best of our knowledge only a few cases have been reported so far [1,2,7,8,10]. It may have been stated that subaortic stenosis can occur in AV septal defects as part of the intrinsic anomaly, but this fact may have been omitted, since this is a totally different type of abnormality and had very little to do with the main subject of the reports [9].

Our newborn case had a 5–8 mm diameter, balloon-like mass of accessory tissue attached to the ventricular aspect of the anterior MV leaflet, protruding into the LVOT that—when expanding—caused subaortic stenosis. The excision of this tissue could be comfortably carried out through the VSD without aortotomy. The combination of accessory MV and VSD is very rare; to our knowledge these congenital heart anomalies (AMVT and VSD together) have never been operated on successfully in the newborn period.

Our other patient exhibited a parachute-like AMVL associated with a fibrotic membrane-type subaortic stenosis. The sac became distended with blood during ventricular systole and severely obstructed the egress of blood from the LV. We have demonstrated that this accessory valve was resectable through the aortotomy without damaging the MV.

In conclusion, in our cases the preoperative echocardiographic examination demonstrated correctly the AMVL and AMVT, indicating that this is the correct diagnostic method for recognizing these malformations. We assume that intraoperative epicardial or transesophageal echocardiography should lead also to an increased recognition of AMVT causing subaortic stenosis.

Complete surgical excision of both types of the subaortic extra tissue obstruction can be successfully accomplished either via VSD, when present, or aortotomy.

These rare malformations of the MV causing LVOTO can occur in both infants and newborns.

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