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Case report

Accessory mitral valve tissue in association with bicuspid aortic valve and aortic coarctation – Case report

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ABSTRACT

Accessory mitral valve tissue (AMVT) is a rare congenital malformation of the heart. The first case of AMVT was described in 1842. The first surgical treatment of this lesion was published in 1963 and the first echocardiography (ECHO) diagnosis of AMVT was performed in 1985.

A 50-year-old male with medical history of surgery of aortic coarctation 39 years ago was accepted to the hospital because of dyspnoea and repeated pain in the left thorax. ECHO revealed bicuspid aortic valve including progressive dilatation of an aortic root and ascending aorta with mild aortic regurgitation. Dilatation of mitral annulus with moderate regurgitation (without stenosis) and accessory mitral valve tissue combined with trivial obstruction of left ventricle outflow tract (LVOT) was also present. AMVT with chordae was excised and mitral annuloplasty has been accomplished. Aortic root remodelling (Yacoub procedure) combined with an aortic valve repair and extra-aortic ring implantation has been performed subsequently. Control ECHO one year following surgery showed no regurgitation on either aortic or mitral valve.

Symptomatology of the patient depends on the presence of AMVT associated with LVOT obstruction, moderate affection of aortic and mitral valves and on concomitant cardiovascular malformations. Echocardiography plays a principal role in the diagnosis, follow-up and indication of surgery of patients with this congenital lesion. Clinically silent AMVT requires follow-up, but it is not an indication for surgery itself. On the other hand AMVT associated with LVOT obstruction or mitral stenosis is an indication for surgery, especially combined with other cardiac malformations.

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Introduction

Accessory mitral valve tissue (AMVT) is a rare congenital malformation of the heart [1]. The first case of AMVT was described in 1842. The first surgical treatment of this lesion was published by MacLean et al. in 1963 [1,2]. The first echocardiogram (ECHO) diagnosis of AMVT was made in 1985. The prevalence of AMVT is not fully known because this finding is often asymptomatic [1]. Golias et al. presented that fewer than 100 cases of AMVT have been reported in the literature [2]. The biggest review article of AMVT was published in 2003 and is based on 63 reports of 90 patients published from 1963 to 2002. The age of AMVT diagnosis ranged from newborns to 77 years. More authors describe incidence of AMVT in adults 1/26,000 ECHOs [1,3,4]. Literature data show a larger prevalence in young men, with a male-to-female ratio of 1.5:1 [1].

AMVT as a congenital heart defect is associated with other congenital anomalies of the heart or great vessels. The most frequent anomalies encountered in the literature are ventricular septal defect (19%), subaortic stenosis (10%), left ventricular hypertrophy (9%), transposition of the great arteries (8%), atrial septal defect (8%), aortic coarctation (5%), dextrocardia (3%), situs viscerus inversum (3%), coronary artery anomalies (2%), double outlet right ventricle (2%) and Ebstein's anomaly (1%) [1,5]. Association of AMVT and congenital left ventricle apical aneurysm was also presented [6]. AMVT can be asymptomatic with the presence of a murmur, but more frequently it is presented with mild to severe left ventricular outflow tract (LVOT) obstruction [4,6]. Rarely AMVT can be presented by congenital mitral stenosis [1,7]. Congenital mitral stenosis may be caused by many conditions such as supramitral ring, commissural fusion, shortened chordae, anomalous mitral arcade, anomalous position of the papillary muscle, parachute mitral valve and AMVT [7–11]. Other possible causes of LVOT obstruction include septal shift due to left-to-right ventricle pressure gradient, pulmonary valve abnormalities, subvalvular fibromuscular tunnel, accessory tricuspid valve tissue, anomalous insertion of mitral valve chordate or papillary muscle to outlet septum, straddling tricuspid valve and accessory endocardial cushion tissue relating to a ventricular septal defect [4,12].

From anatomical point of view, two basic types of AMVT exist. Type I is fixed and type II is mobile. Type I includes nodular (type IA) and membranous (type IB) type and type II includes pedunculated (type IIA) and leaflet-like (type IIB) type [2–4]. Accessory tissue of atrioventricular valve can affect one or both atrioventricular valves; however, mitral valve is the most often affected valve [1]. Panduranga et al. presented six locations of AMVT insertion: the left ventricle wall, interventricular septum, accessory papillary muscle, anterolateral papillary muscle, anterior mitral valve leaflet and the anterior mitral valve chordae [3]. AMVT can vary in appearance from amorphous gelatinous tissue to a mature duplicate of the mitral valve. AMVT has been described in several ways, including sac, balloon, parachute, sail, leaflet, sheet, membranous or pedunculated [7]. Embryologic mechanism of AMVT formation is not clear. More authors presented formation of AMVT as an incomplete separation of mitral (or tricuspid) valve from endocardial cushions [1]. From the pathophysiological point of view, two mechanisms of LVOT obstruction are recognized. If the first type is presented, mass of the accessory tissue causes obstruction and clinical presentation is immediately significant. The second type presents progressive deposition of fibrous tissue due to turbulent flow created by AMVT and progressive increase of outflow gradient [2,4].

Case report

A 50-year-old male with medical history of surgery due to aortic coarctation 39 years ago was admitted to the department of cardiology because of dyspnoea and constant pain in the left thorax. The patient was treated for hypertension and thrombocytopenia. He was long-term monitored for some degree of dilation of the ascending aorta. After admission ECHO revealed bicuspid aortic valve and dilated aortic root including the ascending aorta (aortic ring diameter of 37 mm, Valsalva sinuses diameter of 51 mm, sinotubular junction diameter of 52 mm and ascending aorta diameter of 61 mm) with mild aortic regurgitation up to grade 2. Concurrently mitral annulus dilatation was also found with diameter up to 43 mm. Moderate mitral regurgitation with accessory mitral valve tissue with trivial obstruction of LVOT was also present. Maximal pressure gradient across the LVOT was 14 mm of Mercury column and maximal speed was 1.9 m/s (Figs. 1 and 2). Computer tomography confirmed dilation of the ascending aorta (67 mm) and root respectively (60 mm) and 55% stenosis in the site of resected aortic coarctation (Fig. 3). Coronary angiography did not show any coronary artery stenosis. Surgical treatment of the ascending aorta, root and aortic-mitral valve was indicated.

Standard mid-sternotomy was performed (Fig. 4). Afterwards, a cardiopulmonary bypass was established in standard fashion and the heart was arrested using an anterograde intermittent cold blood cardioplegia. Left atrium of the heart and ascending aorta was opened. Accessory mitral valve tissue in the localization of the anterolateral commissure along with

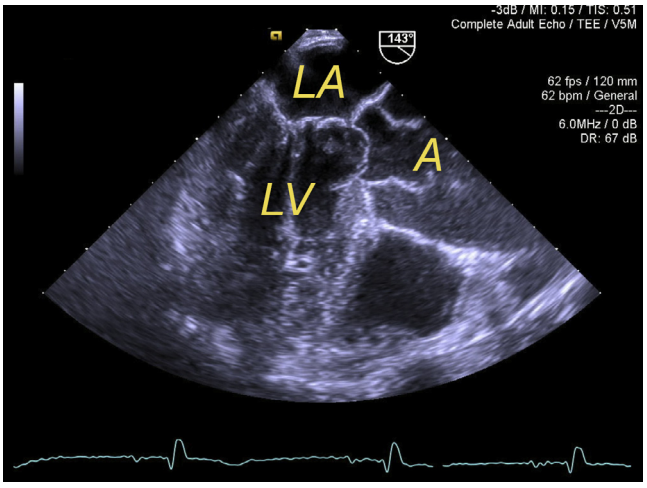

Fig. 1 – Echocardiographic depiction of accessory mitral valve tissue (A, ascending aorta; LA, left atrium; LV, left ventricle).

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