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CASE REPORT

Accessory mitral tissue - an unusual cause of left ventricular outflow tract obstruction

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Abstract Accessory mitral valve tissue is one of the rare anomalies of embryonic development of the endocardial cushion. We describe here a case of a 9 year old male who presented with dyspnoea on exertion. Transthoracic and transesophageal echocardiography revealed aneurysm of membranous part of the interventricular septum producing left ventricular outflow obstruction. Left ventriculography showed a filling defect in the area of mitral aortic interventricular fibrosa probably a localized subaortic membrane. But intraopertive findings showed an accessory mitral valve tissue attached to the annulus of anterior leaflet with its chordal attachment to the papillary muscles of normal mitral valve and to the interventricular septum. The anomalous tissue was excised with its attachment through the aortotomy and left atriotomy. We emphasize, that fixed type of left ventricular outflow tract obstruction produced by an accessory mitral tissue can mimic an aneurysm of the interventricular septum on echocardiography and surgical excision through bicameral approach is recommended.

Keywords Mitral valve · Left ventricular · Echocardiography

Introduction

Left Ventricular Out Flow Tract Obstruction (LVOTO) can be caused by many pathophysiological mechanisms. Various conditions causing congenital left ventricular out flow tract obstruction have been reported in the literature and include aneurysm of the membranous interventricular septum, accessory mitral valve tissue, hypertrophy of anterolateral muscle of left ventricle and metastatic or primary cardiac tumours. We describe here a case of a 9 year old male with Accessory Mitral Valve Tissue (AMVT) producing fixed LVOTO with a review of literature.

Case report

A 9 year old male was admitted with a history of recurrent respiratory tract infection and shortness of breath on exertion New York Heart Association Class II. On physical examination heart rate was 90 per minute and regular, blood pressure was 90/60 mmHg, heart sounds were normal and there was a grade 3/6 ejection systolic murmur at the right second intercostal space. The chest X ray showed a cardiothoracic ratio of 60% with left ventricular type of apex. Electrocardiogram revealed sinus rhythm, left atrial enlargement and left ventricular hypertrophy. Echocardiogram revealed an intact ventricular septum, the perimembranous area being covered by a thin membrane and an aneurysmal out pouching from crest of the interventricular septum projecting into the Left Ventricular Outflow Tract (LVOT) causing a LVOT gradient of 157/96 mmHg. The tricuspid and aortic valves appeared to be normal. Cardiac catheterization showed normal right atrial and right ventricular pressures with no evidence of a shunt at any level. Left ventriculogram in left anterior oblique view showed, a filling defect in the LVOT in the area of Mitral Aortic Interventricular Fibrosa (MAIVF) producing narrowing of the outflow tract and probably a localized sub aortic membrane causing LVOT gradient of 149 mmHg (Fig. 1). The patient was taken up for surgery and the LVOT approached through an aortotomy after conventional median sternotomy and

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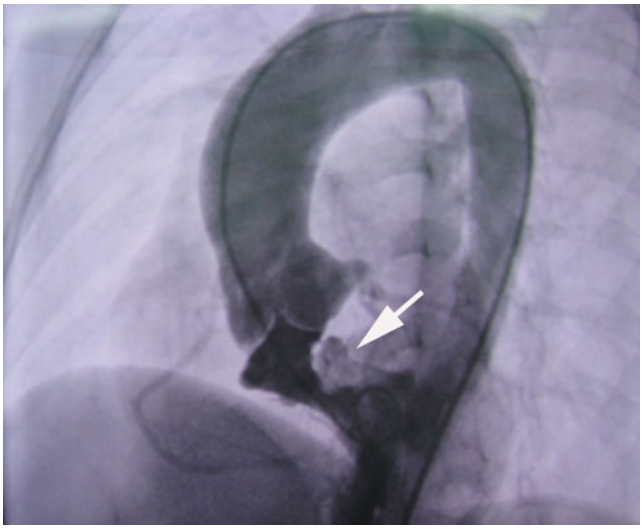

Fig. 1 Left ventriculogram in left anterior oblique view, showing a homogenous mass obstructing the left ventricular outflow tract from behind (*Arrow*)

aortic and bicaval cannulation. Exposure of the LVOT revealed an umbrella like tissue with a texture of anterior mitral leaflet measuring about 4 cm × 3 cm × 3 cm and arising from the mitral aortic interventricular fibrosa near the base of the anterior mitral leaflet. This tissue was partly attached to the papillary muscle of mitral valve and partly with the chordae like strands to the membranous part of the interventricular septum thus causing significant LVOT obstruction (Fig. 2). There was no ventricular septal defect. The left atrium was opened and the mitral valve was examined from the left atrial side. It was free from the LVOT mass. The abnormal tissue along with adjacent fibrous tissue was excised through the aorta (Fig. 3). After excision the aortic and mitral valves were tested for competence and were found to be intact. The post-operative course was uneventful an echocardiogram prior to discharge showed LVOT gradient of 13 mmHg with mild aortic and trivial mitral regurgitation. Histopathology of the excised tissue revealed normal valve tissue with myxoid degeneration. At 3 month follow up, left ventricular outflow gradient was 24 mmHg with mild aortic regurgitation and good ventricular function.

Discussion

AMVT is an extremely rare congenital anomaly of embryonic development of the endocardial cushion [1, 2]. Very few cases of AMVT have been reported in the literature [1–5]. It was first described by McLean in 1963 [1, 2]. Pathology studies suggest that there are two distinct types of AMVT—mobile and fixed. The mobile type is a dysplastic, sail or parachute-like, thickened leaflet that is projected into the LVOT during ventricular systole. In contrast, the fixed type is firmly anchored to the interventricular septum by short chordae, lacks mobility, and demonstrates normal leaflet morphology [3]. The membrane insertion of the accessory mitral valve is usually to the area of aortomitral continuity; however, many variations in the attachment of the chordal component have been reported, such as to the anterior mitral valve chordae, anterior papillary muscle, anterior mitral leaflet valve, left ventricular wall, accessory papillary muscle, or interventricular septum [6]. AMVT is usually diagnosed in the first or second decade of life with clinical manifestations of exercise intolerance, chest pain, syncope, tachycardia resulting from left ventricular outflow tract obstruction. Rarely it can be associated with thromboembolic events or they can present in adult life with an

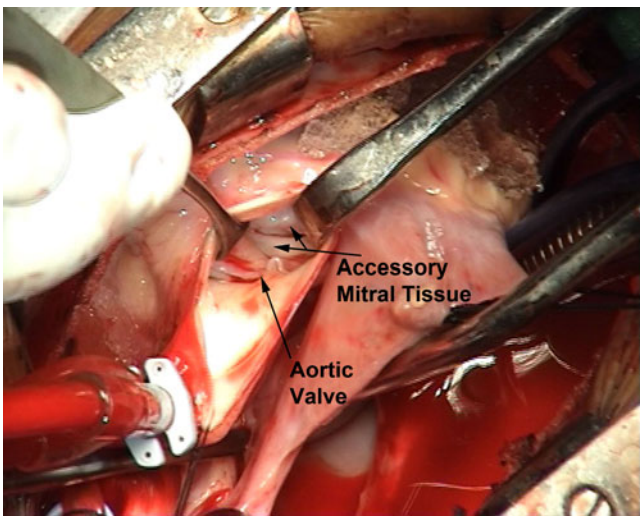

Fig. 2 Aortotomy showing an umbrella like Accessory mitral valve tissue obstructing the left ventricular outflow tract

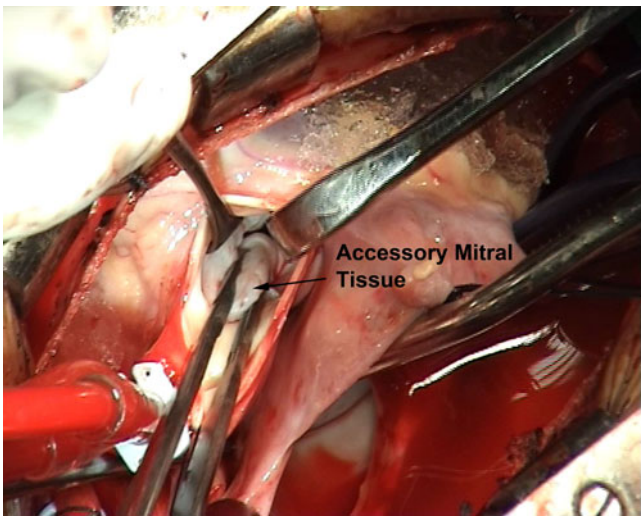

Fig. 3 Accessory mitral valve tissue attached to the anterior mitral annulus and part of its chordae attached to the membranous part of interventricular septum

asymptomatic heart murmur [1, 2, 4]. Despite interest in other non-invasive imaging modalities (i.e. computed tomography and magnetic resonance imaging), echocardiography is currently favoured for both the diagnosis and for determining the clinical significance of AMVT [1–3]. Because other types of left ventricular masses such as tumours, vegetations or aneurysms of membranous septum can produce similar echocardiographic appearances, an AMVT should be considered in the differential diagnosis of any LVOT mass [2, 3]. In our case the mass in the LVOT looked like an aneurysm of the membranous part of interventricular septum. An angiography can visualize a mass in the subaortic area and a high quality cineangiocardiographic techniques should demonstrate the type and severity of subaortic stenosis [2, 3]. In our case a left ventriculogram in left anterior oblique view showed a subaortic stenosis caused by a homogenous contrast medium recess in the vicinity of the mitral aortic interventricular fibrosa of the LVOT. Origin from this area can differentiate it from the aneurysm of the membranous part of the interventricular ventricular septum but cannot differentiate it from the localized subaortic membrane. Surgical excision is the method of choice and surgical removal is indicated in patients presenting with significant LVOTO, even if they are asymptomatic. Asymptomatic patients without substantial LVOT obstruction, aortic insufficiency, or other associated congenital disease do not require removal of AMVT. Antibiotic prophylaxis for endocarditis is indicated and a regular follow up is recommended to identify any progression in LVOTO [2, 7]. AMVT without serious LVOT obstruction carries a risk of thromboembolic complication. Antiplatelet drugs should be administered even in the absence of predisposing factors for cerebrovascular or coronary thromboembolism. [8]. Large AMVT may produce degenerative changes in the leaflets of the aortic valve and severe aortic valvular insufficiency which may require aortic valve replacement [9]. In case of hypertrophic obstructive cardiomyopathy associated with AMVT, an alcohol septal ablation therapy is a treatment of choice provided, the possibility of the presumed AMVT to actually be a flail subvalvular membrane is definitively excluded. Flail subvalvular membranes have been reported as an etiology of subvalvular aortic stenosis [10]. Various surgical approaches have been described in the literature and they are robotic endoscopic [11] or an open access through the aorta, left atrium or the left ventricle. The bicameral approach is adequate and safe [3, 5]. A left ventriculotomy adds to the problem of postoperative ventricular dysfunction [3, 5]. The aortic approach helps in excision of the AMVT and the left atrial view helps in assessing its relation with mitral valve and mitral valve competence after excision. Although an endoscopic technique is the least invasive surgical method to approach intracardiac lesions, such as the AMVT, it can be technically challenging [11].

Conclusion

Fixed type of left ventricular outflow tract obstruction produced by an AMVT can mimic an aneurysm of the interventricular septum. Surgery is the only treatment for significantly obstructive lesion and bicameral approach is safe and recommended.

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