

Isolated Fixed Discrete Subaortic Membrane- A Case Report

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Abstract:

Discrete subaortic membrane, which is an obstructing lesion of the left ventricular outflow tract, remains a surgical challenge. A 7 year old boy came with a discrete fixed sub-aortic membrane (SAM) causing severe Left ventricular outflow tract (LVOT) obstruction. The

membrane was excised totally and the patient had a satisfactory recovery. Discharge echocardiogram found no residual SAM, no gradient across LVOT, mild aortic regurgitation(AR) which was present preoperatively.

Key words: Subaortic Membrane, subaortic stenosis, SAM

(Bangladesh Heart Journal 2015; 30(1) : 43-45)

Introduction:

Fixed subaortic stenosis accounts for 8-20% of patients with congenital left ventricular outflow obstruction. Three anatomical types are described: (i) collar type, in which a fibromuscular ridge is present; (ii) diffuse long segment fibromuscular narrowing or tunnel type; and (iii) discrete membranous subaortic stenosis or subaortic membrane (SAM), which is characterised by a localised crescent-shaped or complete circular thickening of the endocardium on the ventricular septum in the form of a thin fibrous membrane, just caudal to the aortic valve cusps and on a plane usually corresponding to the level of the annulus fibrosus of the mitral valve. SAM is the most common of the obstructions.^{1,2} It is usually associated with other congenital anomalies in 60-70% of cases e.g. VSD, bicuspid AV, coarctation of aorta (COA), atrioventricular septal defects. The subaortic obstruction is almost always progressive but at a variable rate. The condition is rarely diagnosed antenatally or in infancy but often manifests in the 1st decade of life with features of progressive LVOT obstruction, LV hypertrophy and LV dysfunction, or aortic regurgitation. The jet from the narrowed subaortic tract damages the aortic cusps and causes regurgitation; this damage may also render the aortic valve prone to infective endocarditis.⁴ Treatment options have consisted of balloon dilatation or surgical excision.⁵

Case report:

A 7 years old boy got admitted with the history of repeated attacks of fever and respiratory tract infection since childhood and inadequate weight gain. Clinically he was having a Systolic murmur best heard in the aortic area. Chest X-ray was normal & there was sinus tachycardia on ECG. Echocardiography revealed a discrete fixed Sub-aortic membrane (SAM) about 2 mm apical to aortic valve annulus causing severe stenosis (PPG=81mm of Hg, MPG = 51 mm Hg) , mild AR, mild concentric LV wall hypertrophy & good biventricular function .

Surgery was planned. He was approached with median sternotomy with standard cardiopulmonary bypass. RA opened and IVS checked for VSD. LV vented through interatrial septum (IAS). Oblique aortotomy was done few centimeters above sinotubular junction. There was a circumferential thick SAM 2-3 mm below the aortic annulus, part of it was attached with anterior mitral leaflet (AML). SAM totally excised out. In process of taking out SAM, there was linear tear in AML and Right coronary cusp (RCC) of aortic valve. Tear of AML was repaired by 6-0 prolene & RCC was repaired by 0.1mm thick PTFE patch by using 7-0 prolene. Competency of the Aortic valve & Mitral valve were checked by saline test and found competent.

He had an uneventful postoperative recovery & per discharge echo revealed no residual SAM with mild AR and no pressure gradient across LVOT.

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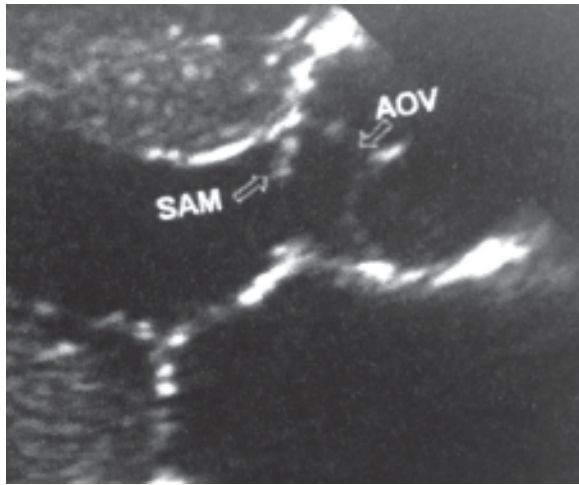


Fig.-1: Preoperative echocardiography showing a discrete SAM

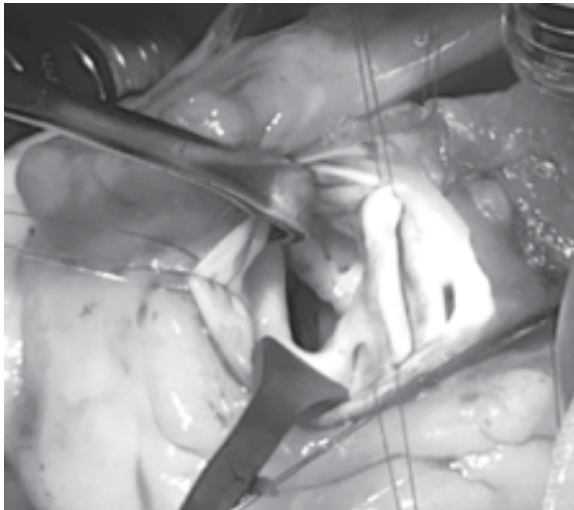


Fig.-2: SAM visible through aortotomy

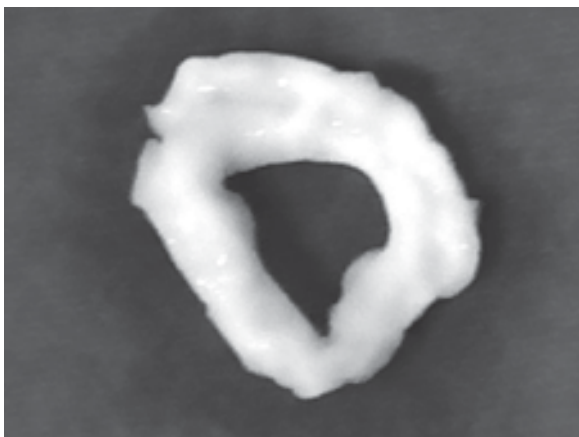


Fig.-3: SAM after excision

Discussion:

Discrete subaortic stenosis is a manifestation of a geometric anatomic alteration in the LVOT. This endocardial abnormality involves not only the subaortic ridge but also the leaflets of the adjacent valves.⁷ Although substantial pressure gradient and aortic regurgitation are the main indications for surgery, controversy persists about the timing of surgical repair and the surgical technique.⁸

Many authors have suggested surgery for patients who have left ventricle–aorta gradients that exceed 30 mmHg or a coexisting cardiac defect that requires surgical correction, while others advocate surgical resection for SAM of any degree because of concerns about the developmental role that subaortic stenosis may play in aortic insufficiency, LV hypertrophy and infective endocarditis. Likelihood of gradual progression as well as development of these complications favour early operation. Moreover surgical resection of fixed subaortic stenosis before the development of a significant outflow tract gradient (>40 mm Hg) may prevent recurrence & reoperation.¹¹

The optimal surgical method for patients with SAM is debatable. Although some surgeons prefer enucleation of the discrete membrane and in selected patients its fibromuscular ridge, many others believe that surgery is not sufficient without routine myomectomy along with resection of the hypertrophied muscle. Radical excision of all diseased tissue, which attains a minimal early postoperative gradient, may reduce the occurrence of late aortic regurgitation.⁹ However; this more aggressive approach increases the risk of iatrogenic damage to the conduction tissue (injury to the conduction tissue between the right and noncoronary cusps), ventricular septum (VSD), and mitral valve.

Recurrence rate is significant but the cause of recurrence of LVOT obstruction is unknown, obstruction occurring even after adequate excision. It is unlikely that a small residual gradient after resection increases the likelihood of recurrence. Regrowth from the region of the septum that was the initial site of fibromuscular obstruction may be an important cause,¹⁰ and routine removal of the underlying septal muscle may prevent this. The other cause of recurrence may be that scar formation from the original excision fixes the diameter of the LVOT and limits its growth. However, all should consider the dynamic nature of the LVOTO may not be evident in the relaxed, cardioplegic heart; therefore, measures should be taken to ensure that the obstruction is completely alleviated once tone returns.

Conclusion:

As surgical resection can now be accomplished with very low mortality and minimal complications, it is attractive to consider surgery at the time of diagnosis to provide better outcome by resisting the natural consequences due to LV outflow obstruction, valvular involvement or infective endocarditis.

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