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CASE REPORT

SURGICAL EXCISION OF DISCRETE MEMBRANEOUS SUB AORTIC STENOSIS

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ABSTRACT

A 7 years old boy presented Cardiac Center of Ethiopia (CCE) with dyspnea on exertion. His chest X ray showed evidence of cardiomegaly and his electrocardiography (ECG) showed left ventricular hypertrophy (LVH). His Doppler 2D echocardiography revealed that there is a discrete sub aortic membrane and gradient in the left ventricular out flow tract (LVOT) was 80mmHG.

Using trans-aortic approach, excision of the sub aortic membrane done. Post-operative the heart picked up in sinus rhythm and weaning from cardiopulmonary bypass was routine. Epicardial Doppler echocardiography showed mean gradient across LVOT of 19mmHG. LVOT was also shown free from remnant membrane. The patient got discharged from hospital on the fifth day.

Keywords: *Discrete membranous sub aortic stenosis, surgical excision,congenital heart disease*

INTRODUCTION

Trans aortic excision of the sub aortic membrane appears to be the most effective mode of management in relieving the hemodynamic obstruction and the patient's subjective symptoms. However several complications can result from the surgical treatment. Overzealous resection in the ventricular septum can lead to heart block from damage to the conduction system, iatrogenic ventricular septal defect, or damage to the anterior leaflet of the mitral valve.

Sometimes worsening of aortic insufficiency may occur, which may end up with requirement of valve replacement. Postoperative endocarditis may complicate the post-operative condition especially in situation of significant gradient across the LVOT.

Since the commencement of open-heart surgery by an all Ethiopia team in our hospital, various open-heart surgeries have been performed with excellent outcomes. We would like to share this specific case (excision of sub aortic membrane) to the medical community, which was performed by the team with an outstanding result.

CASE SUMMARY

A seven years old boy from Addis Ababa came to the center in Addis Ababa with easy fatigability and repeated respiratory infection since childhood. He has no history of sore throat or treatment for tonsillitis. Doppler 2D echocardiography revealed there was a discrete sub aortic membrane just below the aortic valve about 1cm caudal to the valve. The gradient across the LVOT was found to be peak 84mmHG and mean gradient of 45mmHG. Mild to moderate aortic regurgitation with a thickened aortic valve leaflets was noted. There was no mitral or tricuspid regurgitation.

Surgical intervention was planned and open-heart surgery performed through midline sternotomy. Routine aortic and right atrial cannulation performed. Aorta cross-clamped and cold blood regular cardioplegia administered. Aorta opened and a discrete sub aortic membrane about 1cm below the valve was excised. Aorta closed and cross clamp released and heart picked up in sinus rhythm. Epicardial echocardiography after bypass done shows that the dynamic obstruction resulting from the hypertrophy was trivial, thus strengthening the decision not to perform myectomy.

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Weaning was successful and patient was shifted to ICU uneventfully. In the fifth post-operative day, patient was discharged home with no issues. After three weeks, the patient was re-evaluated with TTE and showed mean gradient of 19mmHG and peak gradient of 30mmHG (Fig 1).

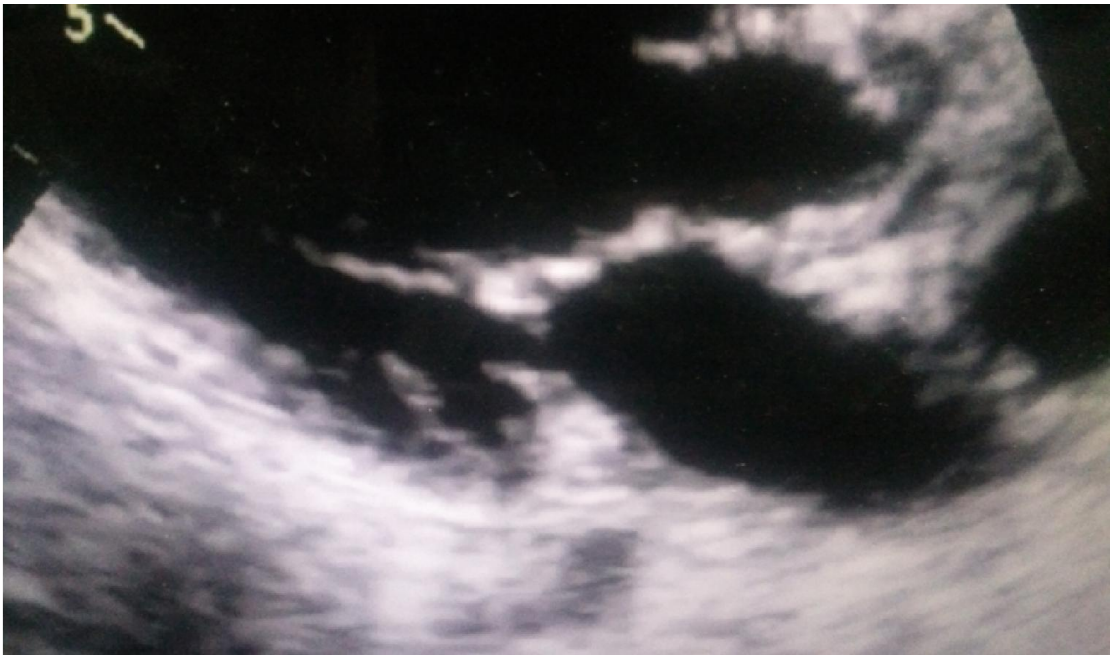

Fig 1: Long axis parasternal view showing clear LVOT after surgical excision of the obstructing membrane.

DISCUSSION

Congenital aortic stenosis is classified into three major anatomical subtypes: valvular, subvalvular, and supravalvular (1). Subvalvular aortic stenosis is further subdivided into the discrete membranous (DMSS) type and the diffuse muscular hypertrophic type, referred to as idiopathic hypertrophic subaortic stenosis (IHSS).

Discrete membranous sub-aortic stenosis is characterized by a localized crescent-shaped thickening of the endocardium on the ventricular septum, 1cm caudal to the aortic valve cusps and on a plane usually corresponding to the level of the annulus fibrosis of the mitral valve (2). The description classically fits the intra operative and echocardiographic finding in our patient.

Sub aortic stenosis is a relatively uncommon cause of left ventricular outflow tract obstruction, encompassing from 8 to 20% of left-sided obstructive lesions (3). The pathogenesis of sub aortic stenosis is elusive and multiple theories have been proposed. According to Somerville, discrete sub-aortic stenosis is thought to be an acquired lesion that results from abnormal motion, growth, or hypertrophy of left ventricular muscle (4).

The natural history of discrete sub aortic stenosis is that in most patients it follows a progressive obstructive course over several months or years (5). With long-standing stenosis of moderate or severe degree, there is associated left ventricular hypertrophy, which may result in secondary dynamic outflow obstruction. This in turn may account for the persistence of gradients in the out flow tract after membrane removal.

The presence of a high-velocity systolic jet impinging on the aortic valve has been suggested as the cause of valve damage and regurgitation. There is also a high reported incidence of bacterial endocarditis, either on the aortic valve or on the membrane itself (6).

The optimal timing of therapeutic intervention for patients with DMSS is uncertain. Some have advocated that operation for DMSS to be undertaken earlier than would be recommended for valvar aortic stenosis with a similar gradient (7). The rationale for such aggressive treatment is that earlier intervention for DMSS prevents the development of abnormal muscular hypertrophy, reduces the incidence of aortic insufficiency, and protects the patient against bacterial endocarditis. It has even been proposed that operation be performed for the diagnosis alone, independent of the patient's symptoms or the gradient (8).

Although routine myectomy, (9) may eliminate the substrate for recurrent subvalvar stenosis, it may create concerns about the possibility of postoperative heart block because of the proximity of the sub aortic obstruction and its attached septal muscle to the bundle of His.

Significant immediate postoperative LVOT gradients are risk factors for recurrence, re-operation, and mortality (10).

CONFLICT OF INTEREST

There is no affiliation to any company or any organization in reporting and writing the above case.

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