LVOT Obstruction in Young Adult

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Abstract: sub aortic stenosis is an important cause of left ventricular outflow tract (LVOT) obstruction in young adult. They usually present with angina, syncope or dyspnoea during the second decade of life. Surgical resection is the intervention of choice for treatment of SAS. Surgical mortality is low, and complications are generally minimal.

Keywords: subaortic stenosis, LVOT, Discrete subaortic membrane , Tunnel.

I. Introduction

The causes of left ventricular outflow tract (LVOT) obstruction in young adult includes sub aortic stenosis, bicuspid aortic valve, supravalvular aortic stenosis, hypertrophic cardiomyopathy and coarctation of aorta. 8% to 20% of cases of congenital left ventricular outflow obstruction are due to subaortic stenosis(SAS) [1]. The causes of Subaortic stenosis (SAS) are discrete fibrous membrane or a diffuse muscular tunnel causing LVOT obstruction. 90% SAS is due to discrete sub aortic membrane, but the tunnel-type lesions are associated with a greater degree of stenosis.

The discrete SAS constitute 6.5% of congenital heart disease of adult,[2] with a male to female ratio of 2:1 . The SAS usually results in symptoms of LVOT obstruction in the second decade of life. The classical symptoms includes angina, syncope and dyspnoea. Here we report a case of discrete subaortic membrane presented with classical symptoms of LVOT obstruction.

II. Case Report

19 year old female patient presented with 2 years history of progressive dyspnoea and exertional chest pain. She also give history of 3 episodes of exertional syncope during the past 6 months. On examination, pulse - 84/mt, low volume, BP – 112/70 mm of Hg. Examination of cardiovascular system revealed heaving type of apex and an ejection systolic murmur at the base radiating to the carotids. Routine blood investigations were within normal limits. ECG showed left ventricular hypertrophy (LVH) with strain pattern. Chest x-ray showed LV type of apex without apparent cardiomegaly. Echo cardiogram in parasternal long axis(Fig: 1) showed concentric LVH with subaortic stenosis and mild aortic regurgitatin (AR). Gradient across the LVOT was 90/53 mm Hg(Fig:2) . M mode across the aortic valve showed systolic fluttering of aortic valve typical of subaortic membrane (Fig:3). To delineate the subaortic membrane further patient was taken for TEE. TEE in aortic long axis showed discrete subaortic membrane with mild AR (Fig :4) . Patient underwent successful Surgical resection.
Figure 1- ECHO in parasternal long axis showing concentric LVH with subaortic stenosis and mild aortic regurgitation.

Figure 2- CW Doppler across LVOT showing Gradient of 90/53 mm Hg.

Figure 3- M mode across aortic valve showing Aortic valve systolic fluttering typical of subaortic membrane.
SAS is an important cause for LVOT obstruction in young patients. SAS can be discrete or the tunnel type [3]. The presence of this discrete ridge or membrane usually results in symptoms of obstruction during the second decade of life.

In 1884 Cheevers first described the Discrete subaortic membrane [4]. The tunnel type of subaortic obstruction has a congenital basis [5], where as discrete stenosis is said to develop because of hemodynamic and rheologic abnormalities localized to the left ventricular outflow tract (LVOT). The lesion may be isolated or associated with other congenital lesions. 37% of patients with SAS have associated perimembraneous ventricular septal defects (VSDs) [6]. A bicuspid aortic valve (BAV) is present in 23% of patients of SAS. SAS may also present as part of a complex of obstructive lesions, as in Shone's complex.

Patients with SAS present with angina, heart failure, or syncope. Severe forms of SAS result in an increased gradient across LVOT and are usually evidenced on physical examination by a harsh late-peaking systolic murmur, delayed and diminished peripheral pulses, and a displaced and sustained left ventricular systolic impulse.

The clinical course of SAS is generally progressive, with increasing obstruction and progression of aortic regurgitation. Aortic regurgitation may result from damage to the valve by the turbulent systolic jet caused by SAS. Some degree of aortic regurgitation occurs in 50% of patients with SAS, where as moderate or severe aortic regurgitation occurs in 12% of patients.

In the absence of left ventricular hypertrophy, dilatation, or failure, intervention can be deferred, but there should be careful lifelong follow-up for symptoms and progression. A continuous wave Doppler derived peak instantaneous gradient of 50 mm Hg is considered severe and portends a poor prognosis if left untreated [7]. Patients with SAS are at a high risk for developing infective endocarditis, which frequently involves the aortic valve [8].

Surgical resection is the intervention of choice for treatment of SAS and is usually done via a transaortic approach. Surgical mortality is low, and complications are generally minimal. [9] Patients with a resting catheter-determined or Doppler-derived estimated peak instantaneous pressure gradient of 50 mm Hg have severe SAS and should undergo operative resection of SAS. Surgical intervention should be considered in patients with lower gradients, if there is left ventricular systolic dysfunction, moderate/severe aortic regurgitation, VSD or symptoms of LVOT obstruction. Tunnel-type SAS is more surgically challenging and often necessitates concomitant myomectomy or application of the Konno-Rastan procedure to reconstruct the LVOT [10]. Concomitant repair of the aortic valve is performed if aortic regurgitation severity is more than mild. SAS recurs in up to 37% of cases after surgical resection.

IV. Conclusion

The causes of LVOT obstruction in young adult includes subaortic stenosis, bicuspid aortic valve, supravalvular aortic stenosis, hypertrophic cardiomyopathy and coarctation of aorta. Subaortic stenosis is an important cause of LVOT obstruction in young females. Surgical resection is the intervention of choice for treatment of SAS. Surgical mortality is low, and complications are generally minimal.
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References


Figure Title:
Figure 1- ECHO in parasternal long axis showing concentric LVH with subaortic stenosis and mild aortic regurgitation
Figure 2- CW Doppler across LVOT showing Gradient of 90/53 mm Hg
Figure 3– M-mode across aortic valve showing Aortic valve systolic fluttering typical of subaortic membrane.
Figure 4- TEE in aortic long axis showing discrete subaortic membrane with mild AR.