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Case Study

Subvalvular Aortic Stenosis in Adults: How Complex It Can Be

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Abstract

Subaortic stenosis is yet a misunderstood lesion. We report two surgical cases which illustrate the diversity and the complexity of this left ventricular outflow tract disease: one isolated discrete form treated with enucleation, and one with associated left ventricular hypertrophy and double-orifice mitral valve treated with extensive ridge excision, myomectomy and mitral valve repair.

Keywords: *Subvalvular Aortic Stenosis; LVOT Obstruction; Double-Orifice Mitral Valve*

Introduction

Subvalvular aortic stenosis (SAS) is one of the common adult congenital heart diseases that can occur either alone or in combination [1]. It encompasses a variety of anatomic lesions from a simple membrane that obstructs the left ventricular outflow tract (LVOT) to an irregular ridge of fibroelastic tissue involving the subaortic region, the aortic and mitral valve, the septum and the fibrous trigones [2]. We report two recent surgical cases of SAS which illustrate the diversity and the complexity of this polymorphic condition.

Case Report

Case 1 Presentation

A 57-year old lady with regular follow-up for a well-documented SAS was referred to surgery because of the worsening of her dyspnea on exertion and the occurrence of palpitations. Echocardiography confirmed the diagnosis of a discrete subaortic membrane (Figure 1) with a mean gradient of 54 mmHg and a pick LVOT velocity of 4.8 m/s. The aortic valve was normal with a mild central regurgitation. There was no other heart abnormality. Surgery was done through median sternotomy using crystalloid arrest and moderate hypothermic cardiopulmonary bypass. After the opening of the ascending aorta with a hockey-stick excision, the subvalvular region was exposed by retracting the aortic leaflets. The enucleation of the fibromuscular ridge was done with a deep excision into the fibrous trigones; the anterior mitral valve leaflet was covered by a layer of abnormal fibrous tissue and a peeling off this layer was associated. There was no associated marked LV hypertrophy and a concomitant additional myomectomy was not necessary. It was confirmed by the immediate postoperative TEE, showing a large open LVOT without residual anatomic obstruction (Figure 2). The postoperative outcome was event-free. Cardiac echocardiography confirmed a good hemodynamic result with a residual gradient less than 10 mmHg and a LVOT velocity of 1.2 m/s. The patient has been asymptomatic till the last 3-year follow-up, with a persistent mild aortic valve regurgitation.

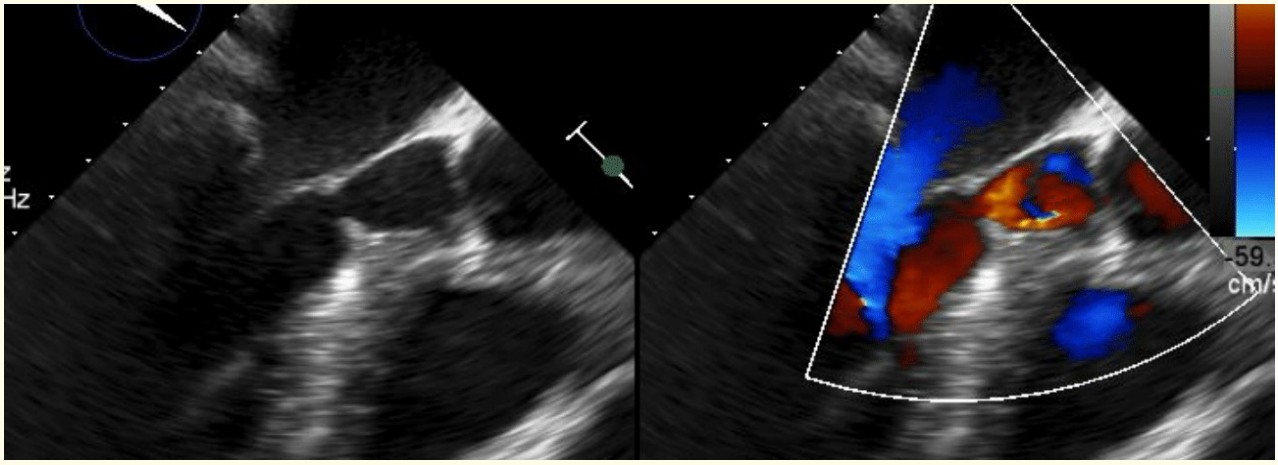

Figure 1: *Case 1: Preoperative TEE showing a typical discrete subvalvular aortic stenosis as a ring into the LVOT.*

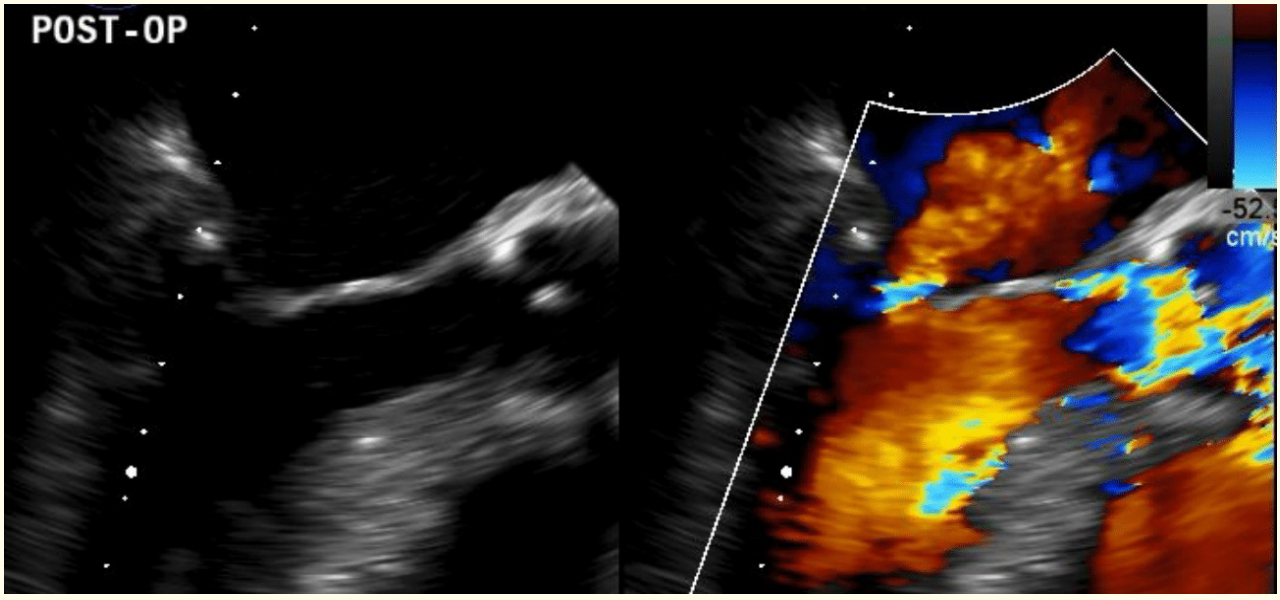

Figure 2: *Case 1: Postoperative TEE assessment showing a free LVOT without residual gradient.*

Case 2 Presentation

A 22-year old female was referred to surgery because of the worsening of her symptoms related to a severe SAS associated with a moderate mitral regurgitation and a mild aortic regurgitation. Echocardiography confirmed the diagnosis of severe SAS due to a thick fibromuscular ridge, associated with a significant LV hypertrophy (Figure 3). Mean gradient was 65 mmHg and pick LVOT velocity was 5.8 m/s. The aortic valve was normal with a mild central regurgitation. Mitral valve analysis confirmed a moderate mitral regurgitation

related to a complete cleft of the anterior leaflet, associated with a double-orifice mitral valve (Figure 4); there was no prolapse or restriction of the both leaflet. According to the complexity of the lesions and the young age of the patient it was decided to perform a complete repair to stabilize the LVOT and to avoid the recurrence of the SAS. Surgery was done through median sternotomy using crystalloid arrest and moderate hypothermic cardiopulmonary bypass. After the opening of the ascending aorta with a hockey-stick excision, the subvalvular region was exposed by retracting the aortic leaflets. A circular sub-valvular membrane was easily identified and resected. A thick secondary chordae of the anterior mitral valve leaflet was resected and a large hemi-circular concomitant additional myomectomy was done resulting in a good enlargement of the LVOT. Then, The mitral valve was exposed through the left atrium and a mitral valve repair surgery was performed: the diagnosis of true double-orifice mitral valve was confirmed with a central thick band between the free edge of both leaflet at their middle and a complete cleft of the anterior leaflet; the central band was respected, the cleft was closed with 4/0 running suture, and an annuloplasty was associated with insertion of a 28-mm Physio-ring (Edwards Lifesciences, Irvine, CA). The immediate postoperative TEE showed a large open LVOT without residual anatomic and hemodynamic obstruction, and without residual mitral regurgitation (Figure 5). The postoperative period was uneventful. At 2-year follow-up, the patient is symptom free but last echocardiography has detected a worsening of the preoperative aortic regurgitation from mild to moderate, associated the a stable good result of LVOT and mitral valve repair.

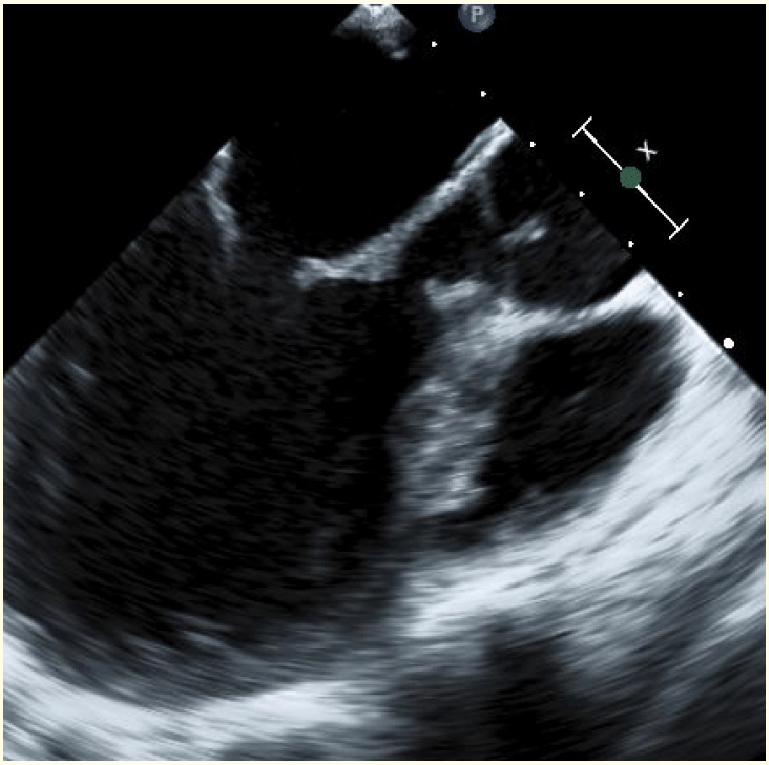

Figure 3: *Case 2: Preoperative TEE showing an extensive subvalvular stenosis with an irregular ridge associated with a LV hypertrophy.*

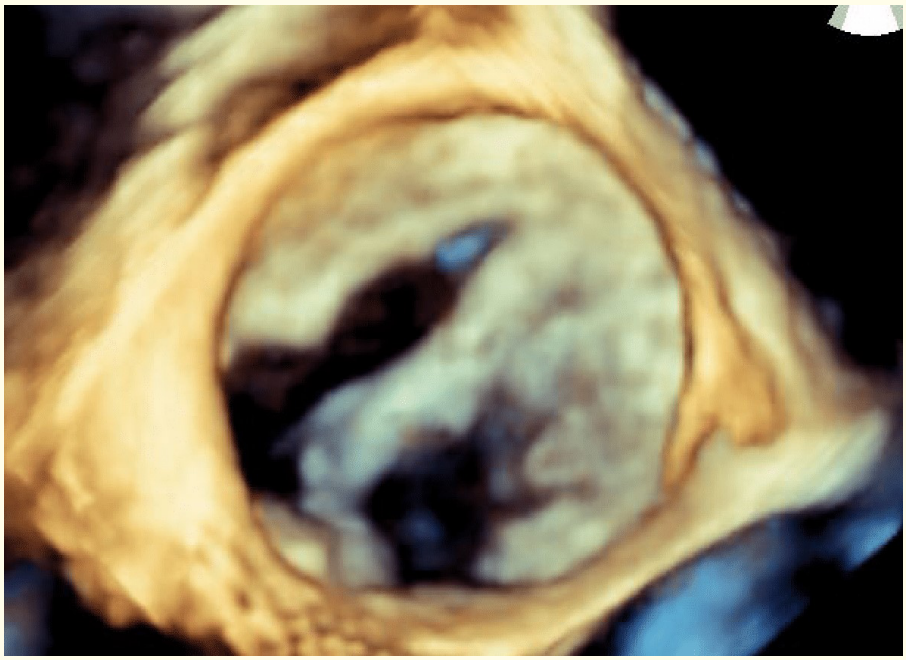

Figure 4: *Case 2: Preoperative TEE with 3D image showing a double-orifice mitral valve associated with a complete cleft of the anterior leaflet along the central band.*

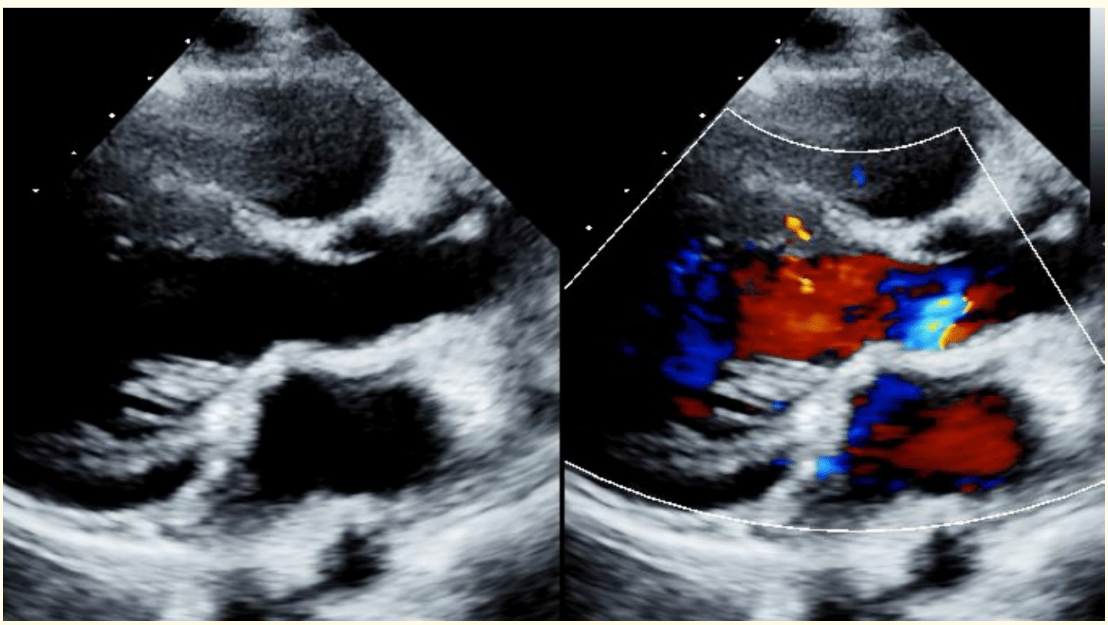

Figure 5: *Case 2: Postoperative TEE showing an open LVOT without residual gradient.*

Discussion

The mechanisms that specifically mediate the late presentation of first-time subaortic stenosis (SAS) in adults are only partially understood. SAS was generally considered a congenital heart disease, commonly associated with congenital abnormalities such as ventricular septal defect, coarctation of the aorta, and patent ductus arteriosus. Nowadays, SAS is more considered a progressive disease that can be isolated or associated with other heart defects. It means that SAS is not a true congenital lesion as it is not usually present at birth, but an acquired, progressive lesion, which grows during postnatal life and may also recur after surgical correction [1,2]. SAS occupies a short channel between the aortic valve and the left ventricular inflow tract, and it is surrounded by important dynamic structures. Persistence of the endocardial cushion tissue in this region and turbulences produced by any associated anatomic factors may drive fibrous proliferation, which can progress and worsen the disease [1]. There appear to be a number of causes, which can be broadly classified into those secondary to congenital heart disease and those not related to congenital heart disease. However, the exact relationship between LVOT anatomy, acquired cardiovascular disease, and late presentation of the de novo SAS in adults remains unclear.

The two reported cases illustrate well the diversity and the complexity of the disease. According to the conceptual classification, our first case was an isolated discrete SAS which needed a simple resection of the sub-aortic membrane, and in our second case it was a fibromuscular ridge with associated LV hypertrophy and mitral valve abnormality requiring a deep enucleation with concomitant myomectomy and associated mitral valve repair. Additional myomectomy did not reduce SAS recurrence or reoperation risk and significantly increased the risk of a complete heart block [3]. Therefore, myomectomy should not be encouraged in most patients and should be performed only in case of marked LV hypertrophy [4,5]. The tubular type is mainly seen earlier in children and requires extensive ablation as Konno procedure.

Associated aortic valve regurgitation (AR) is common but generally mild and nonprogressive over time in most patients; however around 10% of patients progressed from mild to moderate AR, but progression to severe AR is very rare [1,5]. Associated mitral valve disease is not exceptional [1,2], but double-orifice mitral valve (DOMV) as in our second case, is an unusual cardiac anomaly that consists of two anatomically distinct orifices separated by an accessory fibrous tissue. In most cases, the anomaly consists of an eccentric hole; by contrast, the central bridge type with two orifices almost equal in size remains rare [6,7]. DOMV is frequently associated with abnormality of the subvalvular apparatus which may contribute to mitral regurgitation and probably to SAS process [8,9]. In the case reported, a very thick secondary chordae was identified and was resected; the associated moderate MR was related to anterior cleft without prolapse or flailed leaflet. Consequently, in this case, surgery of associated mitral valve repair was mainly indicated to prevent secondary recurrence of SAS and/or worsening of the MR.

In both case, patients were referred to surgery at the step of symptomatic severe LVOT obstruction with high gradient. An adapted surgical strategy allowed to restore a good LVOT anatomy without residual obstruction or gradient. In case 1, SAS was probably related to an isolated fibrous proliferation in the LVOT, and a simple enucleation was enough. In case 2, SAS was possibly the consequence of the congenital abnormality of the mitral valve associated with the hypertrophy of the septum, and needed an extensive surgical repair. Clinical improvement is the rule after surgery; however, the risk of progression of AR and recurrence of SAS justify regular follow-up with echocardiography assessment [1,5].

Conclusion

SAS remains a misunderstood disease in adults. The reported cases confirm that surgery must be adapted to the conceptual lesion: enucleation alone provides good results in discrete SAS, concomitant additional myomectomy is recommended in associated cardiac anomalies.

Disclosure

The authors declare no conflict of interest.

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