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Discrete subaortic stenosis in an adult patient

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ABSTRACT

Discrete subaortic stenosis is an unusual cause of the left ventricular outflow tract obstruction in the adults and characterized by a discrete subaortic membrane. A 52-year-old female patient presented with chief complaints of progressive dyspnoea, chest pain and fatigue. Echocardiographic study showed a discrete fibromembranous ridge located in the subaortic region, which resulted in severe subaortic stenosis, with a mild aortic regurgitation and a mean gradient of 65 mmHg. She underwent surgical resection of the subaortic membrane without any complications. The postoperative course was uneventful, and she was discharged from hospital on the 7th postoperative day. At one-year postoperative follow-up, the patient was doing well without recurrence on echocardiogram. A close follow-up is mandatory for a possible recurrence despite sufficient surgical resection.

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Keywords: Aortic valve; discrete subaortic stenosis; echocardiography; surgery

Introduction

Discrete subaortic stenosis (DSS) is a rare cause of left ventricular outflow tract (LVOT) obstruction in the adults. It is characterized by a membranous or fibromembranous tissue that partly or fully surrounds the subaortic region. This membranous shelf causing flow turbulence in the LVOT can result in progressive subaortic stenosis, concentric left ventricular hypertrophy, and secondary aortic regurgitation [1-3].

DSS is referred to as a congenital disease, but it is mostly considered to be due to an acquired disorder. However, the incidence, rate of progression, the treatment options including the timing and technique, and postoperative outcomes in adults have not been fully elucidated [3-6]. Surgical membranous resection, with or without septal myectomy in patients with DSS, is mostly a successful modality and may provide sufficient relief of LVOTO with low morbidity [1, 7, 8].

Herein, we describe a case of a discrete subaortic membranous ridge in an adult female undergoing surgical intervention.

Case Presentation

A 52-year-old female patient had been suffering from complaints of exertional dyspnoea, chest pain and weakness for 2 years. One year earlier, she had been diagnosed with subaortic membrane using echocardiography, which was performed for the evaluation of cardiac murmur. The patient was strongly urged to consider surgical resection, which

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she refused. The patient was re-admitted to our hospital due to increasing complaints in the last 6 months and surgery was planned.

Physical examination revealed a grade III-IV/VI harsh ejection systolic murmur at the left sternal border. She had New York Heart Association (NYHA) functional classification II. She had hypertension and type 2 diabetes mellitus for 10 years and these disorders are under control with medication. There were no abnormalities in routine blood tests. An electrocardiogram showed sinus rhythm of 75 beats/min and left ventricular hypertrophy with a strain pattern. Chest radiography revealed mild cardiomegaly. There was no dilatation of the ascending aorta. Two-dimensional echocardiographic study including both transthoracic and transoesophageal echocardiography showed a discrete fibromembranous ridge located in the subaortic region, which resulted in severe subaortic stenosis, with mild aortic regurgitation and a mean transmembranous gradient of 65 mmHg. Coronary artery disease was ruled out by the normal coronary angiogram. The patient had no additional cardiac pathologies.

Surgery was performed with standard conventional cardiopulmonary bypass under moderate hypothermia. An oblique aortotomy was carried out and extended into the non-coronary sinus for subaortic resection. Aortic valve appeared tricuspid and aortic annulus was normal. Afterwards, the aortic valve leaflets were carefully retracted to explore the subaortic membrane. The discrete semilunar fibrous membranous structure of 5 x 20 mm was located about 10 mm below the aortic valve (Figure 1). Crescent-shaped fibrous membrane was carefully resected to avoid injury to the conduction tissue and the anterior leaflet of the mitral valve (Figure 2). Residue fibromuscular tissues were also excised (Figure 3). Upon further inspection, interventricular septum was shown to be of normal anatomic structure.

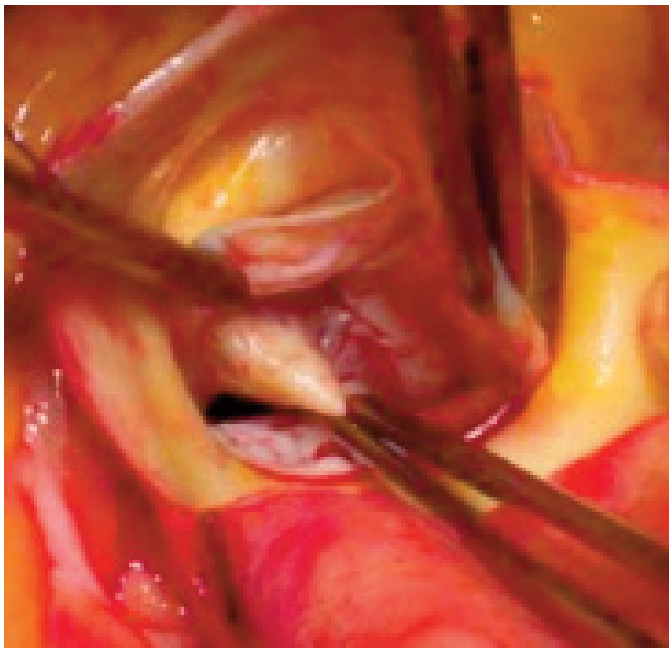

Figure 1. Operative view of discrete membrane below the aortic valve, which is attached to the septum

Histopathological study revealed fibrous membranous tissue, collagen, fibrin tissue, and spindle-shape fibroblasts in haematoxylin-eosin staining. Early after surgery, postoperative echocardiography showed trivial aortic regurgitation. The postoperative course was uneventful, and she was discharged from hospital on postoperative day 7. She was doing well and there was no recurrence in her one-year postoperative follow-up echocardiogram.

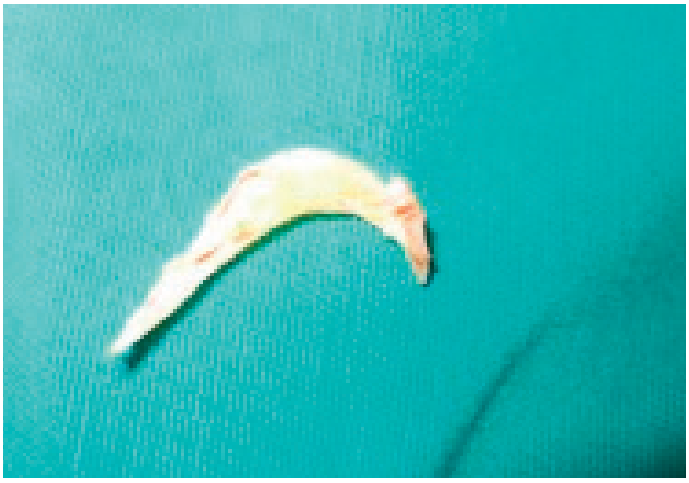

Figure 2. The resected specimen showing a crescent-shaped fibrous membrane.

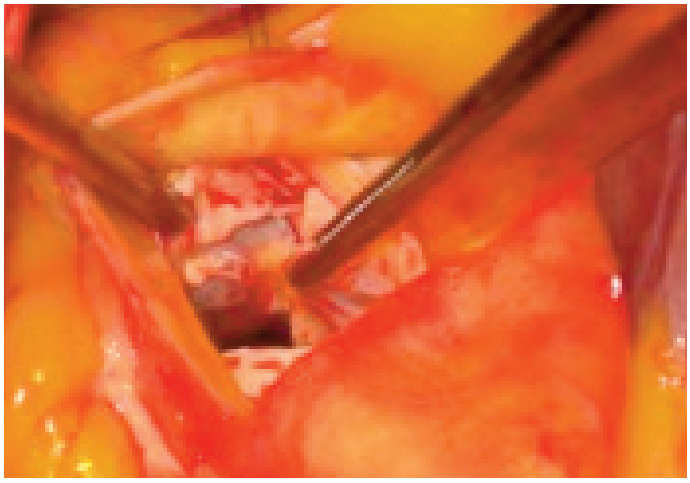

Figure 3. Excision of the residue fibromuscular membrane

Discussion

The prevalence, etiologic characteristics, therapeutic options, and postoperative outcomes for DSS in adults have not been well established [3-6]. DSS is more common in children and accounts for 8% to 20% of all cases of LVOT obstruction requiring surgery [6]. The prevalence of DSS has been increasing in adults with the development of diagnostic methods. In the largest series of 134 adult patients with DSS, Oliver *et al.* [4] reported that the prevalence was relatively frequent of 6.5% for all adults with congenital heart disease.

In adults, DSS is a rare pathology with an unknown aetiology. However, it is a well-described cause of isolated LVOT obstruction in children with its rapid haemodynamic progression and secondary aortic regurgitation. DSS is a progressive and probably acquired cardiac anatomical abnormality, in which the LVOT is characterized by the presence of the obstructing membrane immediately below the aortic valve [1, 8, 9]. This pathology can occur as a primary isolated lesion or in combination with additional subaortic anomalies such as abnormal septal attachments of mitral valve, accessory mitral valve tissue, abnormal left ventricular papillary muscle, anomalous muscular band, and muscularization of the anterior mitral valve leaflet [1]. The lesion is recognized as a result of an ongoing dynamic process and has obvious haemodynamic significance and consequences that reach far into adulthood [3-6].

DSS can also be associated with the presence of other congenital structural anomalies including ventricular septal defect, atrioventricular canal defects, bicuspid aortic valve, coarctation of the aorta, interrupted aortic arch, patent ductus arteriosus, double-outlet right ventricle, and persistent superior left vena cava [1, 8, 10, 11]. In a study, DSS has been determined in 44% of the cases associated with other congenital cardiac anomalies. Two most frequent lesions were ventricular septal defect and aortic coarctation [4]. The lesion may surprisingly appear as a secondary pathology year after the surgical repair of the associated congenital anomaly or mitral valve surgery for rheumatic heart disease [4, 6].

DSS is a manifestation of geometric abnormalities in the LVOT. These abnormal morphological arrangements including small LVOT, increased mitral-aortic fibrous distance, malaligned ventricular septal defect and steepened aorto-septal angle result in altered flow patterns such as increased turbulence [4, 12]. These abnormalities increase septal shear stress producing local fibroproliferative reaction of the endocardium, eventually stimulating development of the subaortic membrane [3, 8, 9]. The high-velocity subvalvular systolic jet in the LVOT can result in progressive significant LVOT obstruction, concentric left ventricular hypertrophy, and aortic valve destruction, which may cause an aortic regurgitation. Patients with DSS are at increased risk to develop acquired aortic valve endocarditis [8, 10].

In a large cohort study of 149 adults at 4 centres, van der Linde *et al.* [3] evaluated the natural history of DSS and identified risk factors for DSS progression, aortic regurgitation progression, and the need for surgery. Interestingly, in contrast to children, longitudinal follow-up (median; 6.3 years) data showed that DSS progressed very slowly in adulthood. Their study demonstrated that the baseline LVOT gradient was 32.3±17 mmHg, with <1 mmHg gradient increase per year. They also documented that, particularly the patients with associated congenital heart disease were at risk for faster progression (p=0.005), while progression did not influenced by the baseline LVOT gradient or age. In their study, mild aortic regurgitation was common (58%), but non-progressive over time (p=0.701). LVOT gradient ≥50 mmHg, LVOT gradient progression, and moderate to severe AR were independent predictors for surgery [3]. Another study by Oliver *et al.* [4] showed a similar slow progression rate (2.3 mmHg increase per year) during a mean follow-up of 4.8 years in only 25 patients with sequential echocardiographic studies. However, they suggested that DSS progression was influenced by the patient age and a significant relationship between age and LVOT gradient (r=0.61, p<0.0001) was found. Their data showed that aortic regurgitation detected by colour Doppler imaging in adults (81%) with DSS, but was hemodynamically significant (moderate to severe) in <20% of the patients [4].

DSS remains a clinically challenging diagnosis in the adults [8]. It may range from asymptomatic to varying degrees of symptoms such as syncope, chest pain, palpitations, weakness or exertional dyspnoea. The diagnosis is made by echocardiography in patients with LVOT obstruction with associated aortic regurgitation. Multimodality imaging is needed to distinguish DSS from hypertrophic cardiomyopathy with obstruction. Echocardiography assesses the

anatomy of the subaortic lesion, dimensions and function of the left ventricle as well as integrity of the aortic and mitral valves [8, 10]. Echocardiographic parameters that should be considered in patients with DSS include mitral-aortic separation, aorto-septal angle, LVOT width, aortic valve dextroposition, left ventricle wall and septal thickness, and indexed left ventricular end-diastolic and end-systolic diameters [10].

Surgery is the intervention of choice for the treatment in severe and symptomatic patients with DSS [8, 10, 13]. The optimal timing of surgical repair and proper surgical technique remains controversial [8, 10]. Surgical decision in adults should be based on the anatomic finding of the lesion, clinical evaluation, left ventricular hypertrophy, systolic function and aortic regurgitation [4]. To prevent damage to the aortic valve and rapid progression of LVOT obstruction, early surgical resection of the subaortic membrane may be recommended in patients with DSS. Indications for surgery include the mean LVOT pressure gradient greater than 50 mmHg and/or left ventricular systolic dysfunction, echocardiographic or angiographic evidence of progressive aortic regurgitation, and coexisting cardiac lesions that require surgery [8]. Definitive treatment for DSS consists of surgical correction of the subaortic obstruction, which may range from simple membrane excision to extensive ring resection, with or without myectomy [11]. Surgery allows sufficient relief of LVOT obstruction with low mortality and morbidity. In our case, she was diagnosed as DSS by patient's clinical status and echocardiography findings, and recommended for surgical intervention. She underwent surgical resection of the subaortic membrane with preservation of aortic valve without complications.

There are no data related to the benefits of early surgery in adults. Data from a study by Oliver *et al*. [4] showed that the benefits of early surgical repair in adults should be questioned. Recently, according to multicentre study by van der Linde *et al.* [3], early surgery in asymptomatic adults with DSS is not indicated solely to prevent rapid progression of the LVOT obstruction or progressive aortic regurgitation.

The surgery for DSS can lead to some complications such as incomplete relief of the obstruction or persistent gradient across the LOVT, an increase in the degree of aortic regurgitation, iatrogenic ventricular septal defect, mitral valve damage, bundle branch or complete heart block, and risk of endocarditis. These early complications are usually associated with aggressive resection during circumferential myectomy. A postoperative transoesophageal echocardiographic examination is essential to identify any iatrogenic complication [5, 6, 8 ,10, 11, 14].

Mortality following surgery for DSS is very low and survival is excellent, with 97% at 20 years [14]. Recurrent DSS requiring reoperation even after a successful repair still is an important problem, especially in the presence of a predisposing associated congenital heart abnormality and depending on the type of the preoperative lesion. Inadequate relief of the obstruction is a major factor in DSS recurrence. Therefore, concomitant selective myectomy is recommend to achieve complete relief of the LVOT obstruction [8, 10, 11, 14].

Surgery is associated with high DSS recurrence (20-30%) requiring reoperation. Recently, in a retrospective multicentre study, van der Linde *et al.* [14] identified risk factors for DSS recurrence, aortic regurgitation worsening and reoperation in a large cohort of 313 adult patients who previously underwent surgical intervention for DSS during the postoperative follow-up period of 12.9 years. They reported that 80 patients (25.6%) underwent at least one reoperation for recurrent DSS (1.76% per patient year). In this cohort, nearly all patients had adequate surgical relief. Mean LVOT gradient decreased from 76 mmHg preoperatively to 15 mmHg postoperatively (p<0.001), and there was an overall increase of 1.3 mmHg per year in the gradient (p=0.001). There was also mild aortic regurgitation in 68% of the patients, but generally did not progress over time (p=0.76).

Predictors for reoperation included female sex (hazard ratio [HR]: 1.53) and progression of LVOT obstruction (HR: 1.45) [14]. In the same study, additional myectomy did not reduce the risk of reoperation (p=0.92), but significantly increased the risk of complete heart block requiring pacemaker implantation (8.1% versus 1.7% of patients who underwent isolated enucleation; p=0.005) [14].

Risk factors for DSS recurrence include younger age at initial surgery, increased age at the time of diagnosis (>30 years old), female sex, closer lesion proximity to the aortic valve (<7 mm), preoperative peak instantaneous LVOT gradient ≥80 mmHg, and intraoperative peeling of the membrane from the aortic or mitral valves [13-15]. These risk factors are associated with higher incidence of reoperation.

Conclusion

It would be important to keep DSS in mind as a cause of aortic stenosis in adults. It can be treated successfully with low mortality and morbidity, with or without concomitant septal myectomy. The anatomic findings of the lesion play an important role on the extent of surgical resection for DSS. The long-term outcome is not predictable in the adults. Therefore, meticulous follow-up is mandatory for DSS recurrence.

Informed Consent

Written informed consent was obtained from the patient for the publication of this case report.

Conflict of interest

The authors declared that there are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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