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Successful Surgical Repair of Semilunar Valves in an Adult Female with Congenital Ventricular Septal Defect

Doğumsal Ventriküler Septal Defektli Erişkin Kadın Hastada Semilunar Kapakların Başarılı Cerrahi Onarımı

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Öz

Ventriküler septal defect (VSD), sağ kalbi ve pulmoner vasküler sistemi, pulmoner vasküler rezistansı arttırarak etkileyen en sık asiyanotik hastalıktır. Tanı almamış subaortik VSD, zaman içinde, ciddi pulmoner arter hipertansiyon yanı sıra aort kapak yetmezliğine de neden olabilir. Ek olarak, VSD yoluyla oluşan soldan sağa şant ile, sağ kalp dilatasyonu gelişebilir. İlerleyen dönemde geri dönüşsüz pulmoner arter hipertansiyonu (PAH) (Eisenmenger Sendromu) gelişebilir. Olgu sunumumuzda, efor dispnesinden yakınan 45 yaşında kadın hastamızı sunmaktayız. Hastada geç tanı almış VSD nedeniyle gelişmiş aort ve pulmoner kapak yetmezliği mevcuttu. Başarılı semilunar kapak onarımı ve eş zamanlı doğumsal kalp defekti onarımı yapıldı. Her 2 kapak otolog perikard dokusu kullanılarak onarıldı. Hastamızda çift mekanik kapak replasmanı yapmak yerine, ömür boyu antikoagülasyona bağlı komplikasyonlardan kaçınmak için kapak onarımı tercih ettik. Olgu sunumumuz erişkin hastadaki kapak patolojisine doğumsal kalp cerrahı yorumunu sunmayı amaçlamaktadır. Olgumuzun VSD'nin hem aort hem pulmoner kapağı komplike etmesi bakımından ve semilunar kapakların yetmezliği yanısıra bu kapakların prolapsusu ile pulmoner overflowun engellenmesi bakımından tek örnek olduğuna inanıyoruz.

Anahtar Kelimeler: Aort Kapak Yetmezliği, Erişkin, Pulmoner Kapak Yetmezliği, Ventriküler Septal Defekt

Abstract

Ventricular septal defect (VSD) is the most common acyanotic congenital heart disease that affects the right heart and pulmonary vascular system by increasing pulmonary vascular resistance. Undiagnosed subaortic VSD may over time cause aortic valve insufficiency as well as severe pulmonary arterial hypertension. Moreover, right heart dilatation can be seen due to the left-to-right shunt through the VSD. The final stages may include irreversible pulmonary artery hypertension (PAH) (Eisenmenger's syndrome) and right-to-left shunting. In our case report, we present a 45-year-old female patient suffering from exertional dyspnea. She was diagnosed with severe aortic and pulmonary valve insufficiency caused by late-diagnosed VSD. Successful surgical repair of degenerated semilunar valves with concomitant heart defect was performed. Both valves were repaired using autologous pericardium tissue. Instead of performing double mechanical valve replacement in our patient, we preferred valve repair to avoid complications related to lifelong anti-coagulation. This case report aims to present the congenital heart surgeon's interpretation of valvular pathology in the adult patients. We believe that our case is unique in that VSD complicated both the semilunar valves insufficiency, limited the pulmonary overflow due to the prolapsus of these valves until adult ages.

Keywords: Aortic Valve Insufficiency, Adult, Pulmonary Valve Insufficiency, Ventricular Septal Defect

Introduction

Ventricular septal defect (VSD) accounts for 20% in congenital heart diseases in childhood and 10% in adult congenital heart malformations (1). It is usually diagnosed in pediatric ages due to incidentally detected cardiac murmur in routine physical examination, in routine examination of fetal echocardiography or further evalution of severe symptoms of congestive heart insufficiency. It is well known that in case of subaortic VSD, the aortic valve leaflet tends to prolapse into the VSD due to the Venturi pulling effect, and aortic insufficiency occurs. In some of cases, tricuspit septal leaflet may produce a pouch to limit the shunt flow through the defect. Therefore, pulmonary overflow can be reduced by this effect. For this reason, VSD can be occult until adult ages. In literature, there are a few cases about adult VSD patients and their outcomes (2,3). In the present study, we introduce an adult female patient diagnosed with VSD, aortic and pulmonary valve insufficiency and her preoperative and postoperative management.

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Case

A 45-year-old female patient was admitted to our clinic due to acute onset severe dyspnea. She had not been diagnosed with any cardiac disease previously. Physical examination showed systolic murmur in pulmonary and aortic valves. Transthoracic echocardiography revealed severe pulmonary and aortic valve insufficiency and a subarterial VSD 20 mm in diameter. After cardiac catheterization, no coronary artery disease was observed and mean pulmonary artery pressure was 60 mmHg, pulmonary vascular resistance (PVR) value was 5 Wood Unit (WU); Qp/Qs was >1.5. Reversibility test was performed at the same time and results were in favour of reversible state. Due to severe dyspnea, diuretic medical treatment was given following the diagnosis. Written informed consent was provided by the patient. Through median sternotomy and under cardiopulmonary bypass (CPB), right atriotomy,

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transverse pulmonary arteriotomy, and transverse aortotomy were performed to obtain an adequate exposure. A pouch made by the tricuspid septal leaflet had covered the perimembranous VSD. Additionally, the right coronary cusp of the aortic valve and adjacent cusp of pulmonary valve were prolapsed due to the Venturi effect of chronic jet flow through the VSD (Figure 1A,1B). Moreover both cusps were adherent to this pouch. The pouch, prolapsed aortic right coronary cusp and adjacent prolapsed pulmonary valve cusp were explored and they were excised carefully (Figure 1A, 1B). The VSD was repaired using a polytetrafluoroethylene patch with a continuous 5/0 propylene suture (Figure 2).

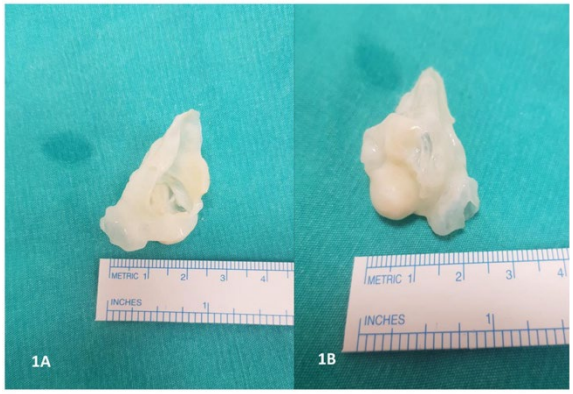

Figure 1. Resected parts of degenerated pulmonary valve and right coronary cusp of aortic valve.

Afterward, the defect of the pulmonary valve cusp was repaired with glutaraldehyde-treated autologous pericardium with a 6/0 propylene suture. A similar repair was performed for the right coronary cusp of the aortic valve in addition to aortic resuspension of three commissures. CPB was terminated with a low dose of inotropic agent. The postoperative course was standard except she required sildenafil 25 mg three times a day. Postoperative transthoracic echocardiogram showed minimal aortic and pulmonary valve insufficiency and no residual VSD, with a pulmonary artery systolic pressure of 25 mmHg. On the 10th postoperative day, she was discharged without any complication. Sildenafil treatment was continued until the 3rd month postoperatively. Postoperative mid-term outcome was reasonable.

Discussion

A VSD is generally diagnosed in childhood due to cardiac murmur during routine physical examination, except for a large VSD which results congestive heart failure in the neonatal period. Depending on its size, location in the heart, PAH value, and PVR value symptoms can occur at different ages. The most common VSD in adults is a small VSD and it remains asymptomatic (4). Rarely, VSD causes valvular infective endocarditis over time (5). Other complications of unrepaired VSD in adult are aortic regurgitation, aortic right coronary cusp prolapsus, pulmonary stenosis, pulmonary regurgitation, mitral valve prolapsus and (1,2). VSD repair in adults is rare in literature (1,4).

On behalf of European Society of Cardiology (ESC) guide in 2020, in patients with evidence of left ventricle volume overload and no PAH (PVR <3 WU in case of such signs), VSD closure is recommended regardless of symptoms (Class I). In patients with no significant left to right (LR) shunt, but a history of repeated episodes of infective endocarditis, VSD closure should be considered (Class IIa), in patients with VSD-associated prolapse of an aortic valve cusp causing progressive aortic regugitation, surgery should be considered (Classs IIa). In patients who have developed PAH with PVR 35 WU, VSD closure should be considered when there is still significant LR shunt (Qp:Qs >1.5) (Class IIa). In patients who have developed PAH with PVR >5 WU, VSD closure may be considered when there is still significant LR shunt (Qp:Qs >1.5), but careful individual decision in expert centres is required (Class IIb). It should be considered that in adults, coronary artery disease might be concomitant with VSD (3).

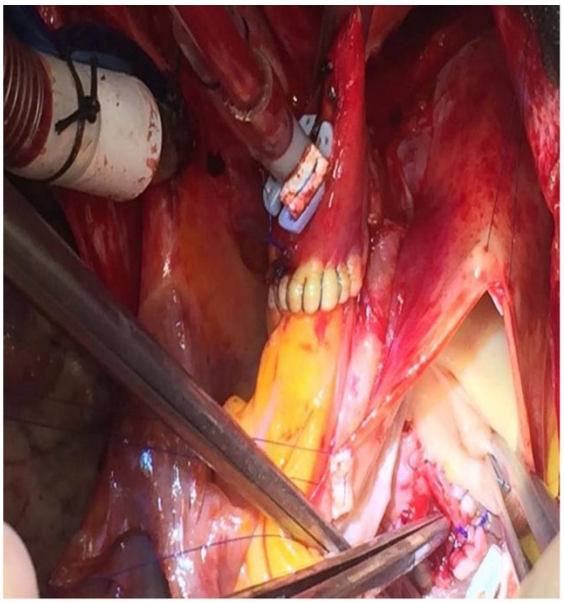

Figure 2. Intraoperative view of patch closure of VSD.

In this case, a pouch limited the PAH by covering the VSD. Additionally, the pulling effect of the pouch and the jet flow, degenerated and enlarged the right coronary aortic valve and adjacent cusp of pulmonary valve. Therefore, this effect probably diminished the flow through the VSD. Thus, the patient tolerated the left to right shunt until the 4th decade. Despite this limitation, our patient had significant pulmonary hypertension at the time of diagnosis and she required sildenafil treatment postoperatively. Prolapsus of cusps and severe valvular insufficiency can be repaired using mechanical valves in general.

However, in our case, removing the excess tissue of prolapsed aortic and pulmonary cusps, and repair the missing part using glutaraldehyde treated autologous pericardium provided adequate coaptation for both damaged semilunar valves.

Conclusions

It is important to conduct a further examination in the case of pulmonary hypertension, to eliminate undiagnosed VSD in adult people. The damaged cusps of aortic and pulmonary valves can be repaired using autologous pericardium in adult patients safely instead of mechanical valve replacement to protect the patient from complications of lifelong anticoagulation in appropriate cases.

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