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Case Report / Olgu Sunumu

Repair of congenital supravalvular aortic stenosis in an adult patient

Erişkin bir hastada doğuştan supravalvüler aort stenozu tamiri

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Supravalvular aortic stenosis (SVAS) is an unusual form of obstructive lesions of left ventricular outflow tract (LVOT) located above the aortic valve. A 45-year-old male case who was admitted with a complaint of dyspnea was diagnosed of SVAS, leading to significant gradient and luminal narrowing. The case was treated successfully through two-sinus reconstruction and ascending aorta replacement. We conclude that the graft replacement of aorta in such cases would be a safe and an effective procedure.

Key words: Ascending aorta; supravalvular aortic stenosis; surgical procedure.

Supravalvüler aort stenozu (SVAS), aort kapağının üzerinde bulunan sol ventrikül çıkım yolu (SVÇY) obstrüktif lezyonlarının nadir görülen bir şeklidir. Nefes darlığı yakınması ile başvuran 45 yaşında bir erkek hastada önemli bir gradyana ve luminal daralmaya neden olan bir SVAS tespit edildi. Olgu iki sinüs rekonstrüksiyonu ve çıkan aort replasmanı ile başarılı bir şekilde tedavi edildi. Bu tür olgularda aort greft uygulamasının güvenli ve etkili bir işlem olabileceği kanısındayız.

Anahtar sözcükler: Çıkan aort; supravalvüler aort stenozu; cerrahi işlem.

Supravalvular aortic stenosis (SVAS) is an unusual form of obstructive lesions of the left ventricular outflow tract (LVOT) just above the aortic valve.[1] We present an adult patient diagnosed with SVAS who was treated with two-sinus reconstruction along with ascending aorta replacement.

CASE REPORT

A 45-year-old male patient with dyspnea was admitted to our hospital, and a physical examination revealed a grade 3-4/6 systolic murmur at the second right intercostal area. Transthoracic echocardiography (TTE) revealed supravalvular aortic stenosis with a gradient of 62/30 mmHg, and computed tomography (CT) detected an hourglass appearance at the sinotubular junction (Figure 1a). The diameter of the ascending aorta was 37 mm at the level of the sinus of Valsalva, and there was a narrowing at the supravalvular level, with luminal diameter of 13 mm. An operation was performed on cardiopulmonary bypass (CPB) using an aortic and a single, two-stage venous cannula. An aortic cross-clamp was applied after initiating CPB, and cardioplegic solution was administered first in an antegrade manner and then continued from the retrograde cannula. An aortotomy was performed superior to the thickened segment of the aorta. The sinotubular junction was severely narrowed. The left and non-coronary sinus of Valsalva were hypoplastic, and the aortic leaflets were thickened. In addition, there were small nodular calcifications on the leaflets (Figure 2). However, aortic leaflet coaptation and mobility were normal, and there was an adequate orifice for the aortic valve. The thickened and narrowed segment of the aorta was then excised. The aortic wall of the left and non-coronary sinuses also seemed to be thickened and was thought to be diseased, so a longitudinal incision was made in the center of the non-coronary sinus of Valsalva to widen the sinus. Next, the left

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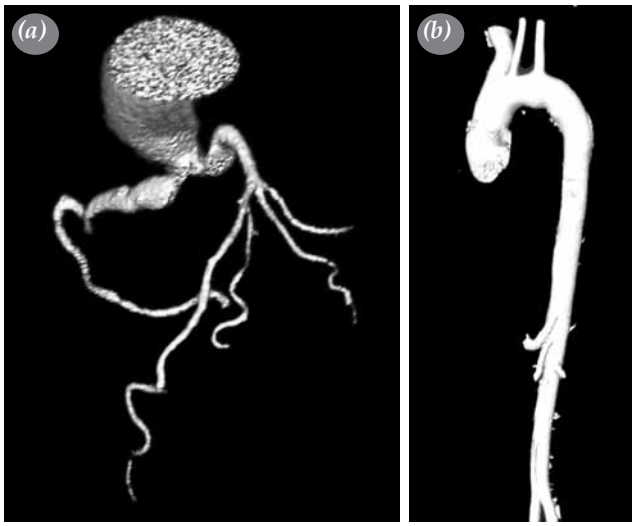

Figure 1. (a) Preoperative computed tomography showing the hourglass appearance. **(b)** Postoperative computed tomography at the third month of follow-up.

coronary sinus of Valsalva was also excised during the preparation of the left coronary ostium. A 30 mm Dacron graft containing to extensions to the left and non-coronary sinuses was then inserted into the excised segment, and the left coronary ostium was reimplanted to the graft. After removing the cross-clamp, the patient was weaned from CPB, and the operation was completed. The postoperative course was uneventful, and postoperative TTE showed a normally functioning aortic valve. Furthermore, control CT at the postoperative third month of follow-up was normal (Figure 1b).

DISCUSSION

Supravalvular aortic stenosis is an uncommon anomaly that is defined by the presence of a narrowing at the level of the sinotubular junction. [1] It is mainly associated with Williams-Beuren syndrome in children.[2] The etiology in our patient was speculated to be congenital in nature because no evidence of inflammatory arteritis or systemic disease was found and no other segments of the aorta or its branches were involved. Surgery is usually performed when the peak gradient across the stenotic segment is 50 mmHg or higher, and since SVAS is progressive, an operation should be carried out when the surgical criteria are met. Simple, localized lesions can be treated by patch aortoplasty, extended aortoplasty, three-patch repair, or sliding aortoplasty.[3,4] The disease is mainly diagnosed in childhood, and at that time, reconstruction of the aortic root rather than the use of prosthetic material

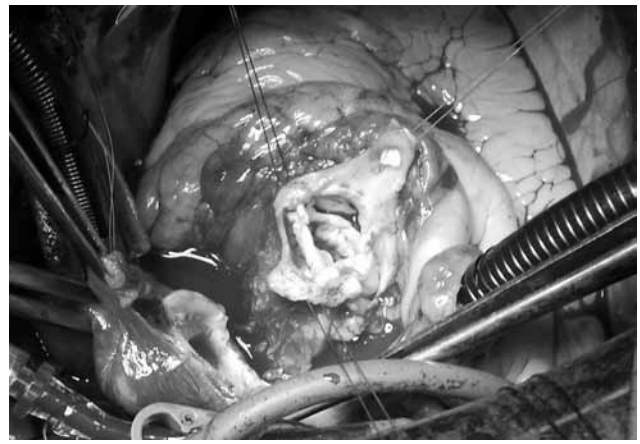

Figure 2. Sinuses of Valsalva and aortic leaflets shown intraoperatively.

is thought to be more reliable because it leads to improved growth of the aortic root.

In this case, the aortic leaflets were thickened, and they had small nodular calcifications. Aortic valve replacement or repair was thought to be unnecessary as the aortic leaflet coaptation and the aortic orifice were normal. Additionally, there was no pathology on the preoperative echocardiographic examination of the aortic valve. However, a study by Imamura et al.[5] documented that the presence of aortic valve stenosis and a bicuspid aortic valve were risk factors for reoperation. Because of this, the pathology of the aortic leaflets may progress; therefore, echocardiographic follow-up is essential for monitoring this situation. Furthermore, patients with a bicuspid aortic valve and aortic valve stenosis should be considered for aortic valve intervention.

There was a very narrow luminal orifice of the aorta at the level of the sinotubular junction in our patient. Moreover, the non-coronary and left sinus of Valsalva were hypoplastic, and the sinus walls appeared to be thickened, although it was reported that the diameter of the ascending aorta diameter was 37 mm at the level of sinus Valsalva on the preoperative CT. For this reason, the diseased segments in this patient were fully excised to avoid reoperation, and multiple sinus reconstruction with graft material extending to the ascending aorta to enlarge the sinotubular junction was the preferred surgical option.

In an adult patient, reconstruction of the aortic sinuses and replacement of the ascending aorta at the level of the sinotubular junction using graft material can be an effective, safe, and non-time-consuming procedure.

Declaration of conflicting interests

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