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Case Report

Aortic Valve–Sparing Surgical Treatment of Supravalvar Aortic Stenosis in a 65-Year-Old Adult

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Supravalvar aortic stenosis (SVAS) is a rare congenital cardiac disease that usually co-occurs with Williams syndrome. In the adult population, a few SVAS cases have been reported in patients affected by homozygous familial hypercholesterolemia. However, because of the rarity of this disease entity, there is no standard surgical treatment for SVAS. Here, we present a case of successful surgical treatment using an autologous excised aortic patch in a 65-year-old patient with SVAS.

Keywords: Supravalvar aortic stenosis, Aortic valve

Case report

Supravalvar aortic stenosis (SVAS) is a rare cardiac anomaly. It usually occurs in combination with Williams syndrome—with a typical facial appearance and mental retardation [1]—but can also present in adult patients affected by homozygous familial hypercholesterolemia (HFH) [2,3]. Because of the rarity of this disease, the surgical technique for SVAS is not standardized and has evolved from a plain patch technique to simple sliding aortoplasty [4,5]. In adult patients with SVAS, conventional surgical treatment is difficult to apply due to reduced flexibility and atherosclerotic changes of the vasculature. We performed autologous excised aortic patch aortoplasty and ascending aorta replacement sparing the aortic valve in a 65-year-old adult patient with SVAS who did not have either Williams syndrome or HFH.

A 65-year-old female patient with a history of transient ischemic attack. hypertension, dyslipidemia, and paroxysmal atrial fibrillation had SVAS. She took medication for dyslipidemia, hypertension, and atrial fibrillation. Her blood cholesterol level was 230 mg/dL and no other familial member had dyslipidemia. Preoperative echocardiography showed SVAS with a peak velocity of 4.5 m/sec and mild aortic regurgitation with an ejection fraction of 65%. Computed tomography showed severe focal stenosis at the aortic root with diffuse soft tissue thickening and calcification with a diameter of 14×10 mm (Fig. 1).

The operative approach was through a median sternotomy. Cardiopulmonary bypass was instituted with a cannula for arterial return in the ascending aorta and a venous single cannula in the right atrium. The aortic cross-clamping point was decided after manual palpation of the area of calcification. Cardiac arrest was achieved using cold antegrade cardioplegic solution. The aorta was transected several millimeters distal to the point of stenosis. The calcified ascending aorta was removed to a few millimeters below the ascending aorta cross-clamping site. After a careful inspection of the stenotic segment of the sinotubular junction, as well as the conditions of the coronary opening and the aortic valve, we meticulously excised the stenotic calcified tissue, taking care not to damage other tissues. Even though the intimal defect of the sinus portion appeared serious (Fig. 2B), it was not especially remarkable because the aortic wall was thickened. Since the stenotic tissue was close to the coronary opening and aortic valve commissure, the procedure was time-consuming. After the removal of stenotic tissue, an incision was made in the non-coronary sinus of the proximal aorta. Autologous healthy aortic tissue from the previously excised ascending aorta was used for patch aortoplasty. After a saline test to detect possible aortic regurgitation, ascending aorta replacement was

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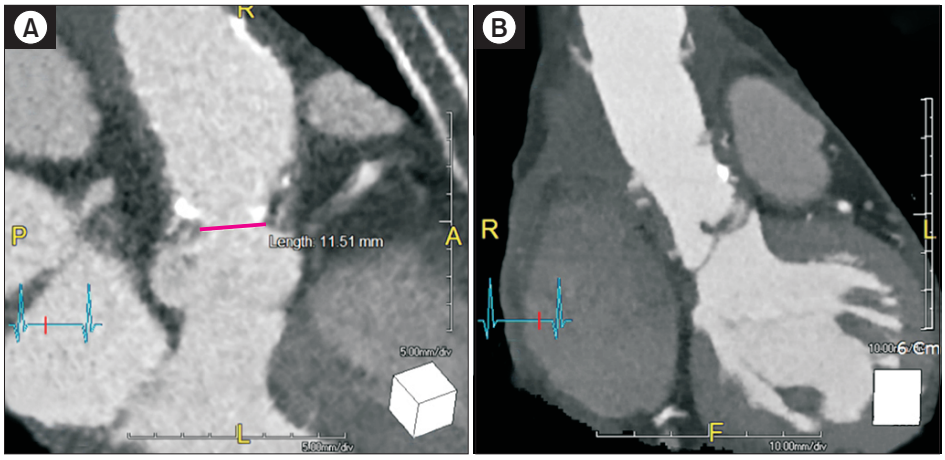

Fig. 1. Preoperative (A) and postoperative computed tomography (B) showed the relief of supravalvar aortic stenosis.

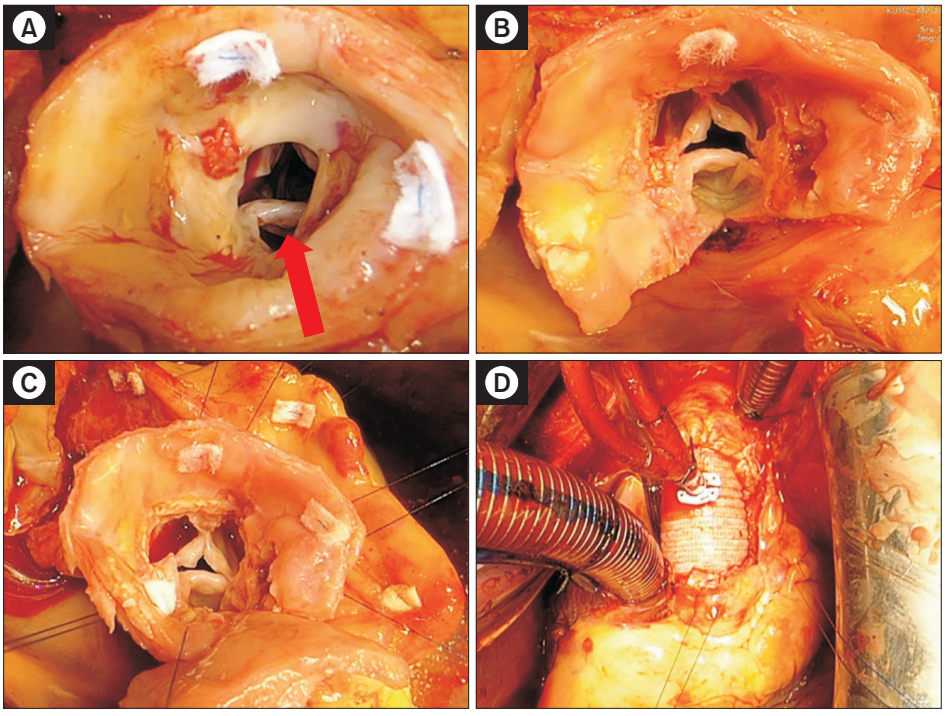

Fig. 2. Supravalvar aortic stenosis repair procedure. After transection of the aorta distal to the narrow point, the calcified tissue was carefully inspected (A). After removal of the stenotic tissue, an incision was made into the noncoronary sinus of the proximal aorta (B). Patch aortoplasty using autologous healthy aortic tissue from the previously excised ascending aorta was performed (C). Finally, ascending aorta graft interposition was performed (D).

performed using a 24-mm Gelweave graft (Vascutek, Renfrewshire, Scotland) (Fig. 2). After surgery, the patient had an uneventful postoperative course with arrhythmic medication and electrocardiogram monitoring. Follow-up echocardiography and computed tomography showed decreased SVAS, with a peak velocity of 2.3 m/sec and an increased sinotubular junction diameter of 21×21 mm (Fig. 1B). The pathologic report of the aortic tissue was simply atherosclerosis with calcification. The patient did not have either HFH or Williams syndrome.

The patient provided written informed consent for the publication of clinical details and images.

Discussion

SVAS is a rare cardiac disease that is often progressive in childhood, and scant data are available on its outcomes in the adult population [6]. In particular, only anecdotal reports exist of SVAS in patients older than 60 years [2]. Irrespective of whether its origin is congenital or acquired, the surgical goal is to enlarge the aortic root and to maintain aortic valve function. Because this patient showed extensive calcification inside the aortic root and ascending aorta, as well as aortic regurgitation associated with old age, we could not rule out the possibility of performing a Ben-

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tall operation.

However, after careful observation and meticulous removal of the calcified lesions, we were able to preserve the coronary opening and aortic valve. Since it appeared that removal of the calcified tissue itself was not sufficient to decrease the pressure gradient, we decided to perform an additional patch aortoplasty using the autologous excised ascending aorta. From our experience, we knew that sliding aortoplasty is a good surgical option for handling SVAS [4,5]. However, as this procedure is not suitable for adult patients with stiff aortic tissue, we used a synthetic graft for ascending aorta replacement and performed a modified procedure using patch aortoplasty with autologous aortic tissue. With regard to the choice of patch material, a Dacron patch is stronger than other tissue types (e.g., pericardium) for preventing aneurysm formation. However, because using a Dacron patch would have caused difficulties in handling needle-hole bleeding and resulted in an uneven reconstruction of the aortic wall in terms of its ability to endure aortic pressure, we decided to use autologous healthy aortic tissue that had been removed for the ascending aorta replacement. Using autologous aortic tissue not only avoided the need for foreign material, but also had the advantage of enabling easy handling during the suturing procedure; furthermore, it may prevent future aneurysm formation, which is a possible complication of using pericardial tissue. In conclusion, we were able to treat SVAS in an older patient safely using modified patch aortoplasty without aortic valve replacement.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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