SINGLE STAGE SURGICAL TREATMENT OF THE CONGENITAL SUPRAAORTIC STENOSIS ASSOCIATED WITH ACQUIRED MULTIPLE CORONARY ARTERY DISEASE

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Supravalvular aortic stenosis is an uncommon congenital disease. Tortuosity or aneurysmatic enlargement of coronary arteries is more common than in normal population and atherosclerosis in the coronary arteries can be accelerated because of hypertension in the aortic root. A 29-year-old male patient was operated on for the supraaortic stenosis and coronary artery disease in the same operation. Double arterial and one venous graft were used for the coronary revascularization and the stenotic segment of the aorta was enlarged with single sinus aortoplasty technique. This one-stage procedure is safe and can be protective against perioperative or late mortality in these patients.

Key words: Supravalvular aortic stenosis, coronary artery disease, single sinus aortoplasty

Supravalvular aortic stenosis can be alone or associated with other cardiac anomalies. The most common associated pathology is multiple stenosis in the peripheral pulmonary arteries, but other cardiac pathologies like coronary artery aneurysm, intracardiac defects or other types of left ventricular outflow tract obstructions can be observed (1,2). This uncommon congenital disease is associated with normal systolic

pressure in the aorta and its branches with the singular exception of the coronary arteries which are hypertensive (2). Supravalvular aortic stenosis causing high blood pressure can cause dilatation, tortuosity, aneurysm formation or early atherosclerotic changes at the coronary arteries (2-4). These changes at the coronary arteries can be the reason of sudden death. We report an adult patient with a congenital supravalvular aortic stenosis associated with three-vessel coronary artery disease. Both pathologies were treated surgically in the same operation and we have not found any report like our case in the literature.

CASE REPORT

A 29-year-old man came to our clinic with sternal pain since two years and his supravalvular aortic stenosis and coronary artery disease were diagnosed echocardiographically and angiographically. At echocardiography, the interventricular septum was measured as 1.3 cm and systolic gradient of the stenosis was measured as 60 mm Hg. Cardiac catheterization revealed supravalvular aortic stenosis with a 58 mm Hg pressure gradient and severe atherosclerotic lesions in three coronary arteries. He had a normal pulmonary arteriography. The levels of his cholesterol and lipoprotein subgroups were normal. ECG showed left ventricular hypertrophy and anterior wall ischemia. He underwent the operation and arterial cannulation was carried out through the distal ascending aorta and single venous cannulation was applied through the right atrium. Retrograde continuous blood cardioplegia was used for myocardial protection. When aortotomy was performed toward the noncoronary sinus, the stenotic segment in the aorta was observed to be 1 cm above the coronary ostia. The intimal surface of the aorta was normal and did not have any calcification or atheromatous plaque. Firstly, distal coronary anastomoses (LIMA-LAD, RIMA-RCA, and venous graft-Cx system) were performed. Before proximal anastomosis was performed on the pericardial patch, the intimal flap was resected and the stenotic segment of the aorta was enlarged with single sinus aortoplasty technique using the glutaraldehyde-treated autologous pericardium. In the first postoperative year, he was examined echocardiographically and there was no residual gradient at the stenotic level. Left ventricular hypertrophy has regressed significantly.

DISCUSSION

Supravalvular aortic stenosis is inherited as an autosomal dominant trait, although sporadic cases can be observed (5). Congenital supravalvular aortic stenosis is an uncommon form among the other left ventricular outflow tract obstructions. Surgically, two types are present: a localized form and a diffuse form. This anomaly appears as a localized form in 85% of patients and it develops just above the attachment of the aortic cusps at the commissure (6). On the other hand, the intimal flap at this level increases the stenosis. The opposite interaction between them decreases the inflow of sinus of Valsalva, and that may interrupt coronary blood flow. Especially, obstruction of the left coronary ostium can cause sudden death. Coronary and left ventricular insufficiency is the most common cause of death in patients who have not been operated on. This uncommon lesion is unique, because only the coronary arteries among the other systemic vessels are exposed to high systolic pressure, which leads to aneurysmatic dilatation, tortuosity, and premature atherosclerosis of these arteries. Aneurysmatic enlargement of the coronary artery system can be observed in these patients (2,4).

Coronary artery lesions are categorized as congenital or acquired (7). Important congenital lesions are intimal hyperplasia, intimal fibrosis, and medial hypertrophy. Generally, luminal narrowing is seen in the proximal segment of the coronary arteries and the most common type is ostial narrowing (4,6,8,9). Although atherosclerotic coronary artery disease associated with supravalvular aortic stenosis is common in patients with familial hypercholesterolemia (10), atherosclerotic coronary artery disease is very rare in patients with sporadic congenital supravalvular aortic stenosis (2,7). In the

report of the Mayo Clinic, it was stated that secondary structural changes in the coronary arteries caused by high systolic pressure in the aortic root included intimal hyperplasia and atherosclerosis (7). The high smooth muscle content of the media of the coronary arteries may predispose to these early occurring and progressive processes (7). If this associated acquired disease is not treated, it can cause early or late death after correction of supravalvular stenosis in the adult patients.

Although atherosclerotic disease is rare in the adult patients with congenital supravalvular aortic stenosis, supraaortic stenosis should be corrected with CABG in the same operation. Although we have not observed any complication with the pericardial patch until now, to avoid late complications of the patch, we recommend that full arterial revascularization must be the first choice of revascularization in these patients. We have not used radial artery as arterial conduit, when we performed this one-stage procedure. If radial artery must be used, its proximal end can be anastomosed to internal mammarian artery (end-to-side). This one-stage procedure is safe and can be protective against perioperative or late mortality in these patients.

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