Case report

Surgery for calcific aortic root stenosis in homozygous familial hypercholesterolemia

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Abstract

A 35-year-old female with homozygous familial hyperlipidemia (IIa) was referred to our hospital for an operation against supravalvular and valvular aortic stenosis. She had been treated with low-density lipoprotein apheresis for 20 years, and total cholesterol ranged between 200 and 400 mg/dl under this treatment. She had undergone percutaneous coronary intervention for ostial stenosis of the right coronary artery three times since the age of 19. Unenhanced three-dimensional computed tomography showed supravalvular stenosis, funnelling and heavily calcified aorta. An operation was performed under deep hypothermic circulatory arrest without aortic cross clamping. After the ascending aorta had been replaced with a one-branched vascular graft, arterial perfusion was resumed. The stenosed ascending aorta was resected at the sinotubular junction. Because the aortic root was still extremely small, the noncoronary sinus and the commissure between left and right coronary cusp were incised, and the aortic root was enlarged with linguiform vascular-graft patches. A 21-mm mechanical valve was implanted. The postoperative course was uneventful.

Keywords: Homozygous familial hypercholesterolemia; Aortic root; Stenosis; Calcification; Circulatory arrest

1. Introduction

Cardiovascular involvement in patients with homozygous familial hypercholesterolemia (HoFH) is well known. Stenosis of the aortic root, including the aortic valve, is considered to be cardinal [1]. Only a few cases of surgical repair have been reported for this condition, probably due to the short life of the patient and the difficulty of the operation [2—4]. For patients with a severely calcified ascending aorta, a particular surgical strategy is required. We report a case with aortic root stenosis who underwent aortic valve replacement with patch-augmentation of the root, following replacement of the heavily calcified ascending aorta under circulatory arrest.

2. Case report

This case was a 35-year-old female. A xanthoma was first detected at the age of 3, when she was diagnosed as type IIa homozygous familial hypercholesterolemia. Activity of low-density lipoprotein (LDL) receptor was 14%. LDL apheresis was performed from age 16, and serum total cholesterol ranged between 200 and 400 mg/dl. Chest pain on effort became apparent at age 19, and ostial stenosis of the right coronary artery was found by coronary arteriogram. Since then, the patient has undergone three percutaneous coronary interventions for the same lesion. Aortic stenosis (AS) of pressure gradient 80 mmHg was first detected at the age of 31 by echocardiogram. Angina on effort began at age 34, but no stenotic lesion was detected on the coronary system. Echocardiography revealed progression of AS, with a pressure gradient of 129 mmHg. Unenhanced computed tomography (CT) revealed supravalvular aortic stenosis, funnelling of the ascending aorta, and severe calcification of the entire aorta (Fig. 1). On cross-sectional CT, the outer and inner diameters at the sinotubular junction were 20 and 10 mm, respectively.

The operation was performed under deep hypothermic circulatory arrest, without aortic cross clamping. A small space for cannulation was found by epiaortic echography on the heavily calcified ascending aorta. Retrograde femoral arterial perfusion was avoided in fear of embolic events. Extracorporeal circulation was established by ascending aortic perfusion and bicaval venous drainage, and core cooling was applied. Under circulatory arrest at a pharyngeal temperature of 21 °C, the ascending aorta was incised at the cannulation site. Because calcification of the proximal arch...
was heavy, careful endoarterectomy was carried out. A 24-mm one-branched vascular prosthesis was then anastomosed at the site with 3-0 braided polyester sutures using a Dacron-felt strip for reinforcement. Following deairing and flash-out of debris with retrograde cerebral perfusion, the proximal side of the prosthesis was clamped and extracorporeal circulation was restarted via the side branch. The aortic root showed remarkable calcification and extreme narrowing on the sinotubular junction and the annulus. Though the coronary ostia were confirmed to be patent, the adjacent aortic wall showed marked calcification. There was no mobility of the cusps, though adhesion was not seen. The ascending aorta was resected just above the coronary ostia. For adequate exposure of the aortic root, the noncoronary sinus was incised extending to the annulus. However, the aortic annular size was still small, so that an additional incision was made at the commissure of the left and right coronary cusp. Linguiform patches made from the vascular prosthesis were inserted at the incised sites, and the annulus was enlarged (Fig. 2). It was then possible to implant a 21-mm mechanical valve (CarboMedics Inc., Austin, TX, USA). Proximal anastomosis was next carried out. Though a 60-

Fig. 1. (a) Unenhanced three-dimensional computed tomography (CT) showed supravalvular stenosis (arrow), funnelling of ascending aorta and calcification of the entire aorta. (b and c) Preoperative two-dimensional CT. (d) Postoperative enhanced CT demonstrates no stenosis in the ostium of the right coronary artery.

Fig. 2. The annulus was enlarged with linguiform vascular-graft patches. LCC: left coronary cusp; RCC: right coronary cusp; NCC: noncoronary cusp.
19.9 respectively, and the lowest pharyngeal temperature was 19.9 °C.

3. Discussion

Coronary arteries and the aortic root, including the valve, are well known to be the affected site of atherosclerosis in HoFH. In particular, supravalvular AS and funnelling of the ascending aorta derived to poststenotic dilatation are considered to be peculiar to this disease. Exposure to high concentrations of blood cholesterol in early life is likely to cause lipid infiltration to the aortic root, leading to supravalvular and valvular stenosis [1,5,6]. Our patient had undergone three percutaneous coronary interventions since the age of 19. Access to the coronary artery became more difficult with time. This fact implies that the progression of supravalvular stenosis should be attributed to hypercholesterolemia rather than having a congenital origin.

Severe calcification of the ascending aorta, also frequently seen in HoFH [7], makes the operation more complicated. Yasuda et al. [4] performed endoarterectomy of the ascending aorta under temporary circulatory arrest, followed by aortic valve replacement under aortic cross clamping, in a patient with AS and HoFH. We carried out replacement of the ascending aorta under circulatory arrest and retrieved systemic perfusion via the branched prosthesis. In cases with aortic root narrowing, it is mandatory to resect and replace part of the ascending aorta to release supravalvular stenosis. It is therefore safer to replace the entire ascending aorta under circulatory arrest, rather than risk clamping the endoarterected and fragile aorta.

It is often difficult to expose the aortic annulus in patients with heavily calcified and small annulus. In such cases, incision into the Valsalva sinus of the noncoronary cusp provides easier resection of the cusps and placement of sutures at the annulus. The incised part is augmented with a linguiform vascular-graft patch. Because the annular enlargement was insufficient in our case, patch augmentation was used at the commissure between the left and right coronary cusps.

Although a full root replacement might be a possible choice, severe calcification around the coronary ostia discouraged us from adopting this procedure because coronary reimplantation was considered difficult. Proactive patch plasty of the right coronary ostium was not done because of this heavy calcification.

In recent years, improved prognosis is expected in patients with HoFH as a result of LDL apheresis, more potent statins and a newly introduced cholesterol absorption inhibitor [8—10]. However, these modalities do not yet completely avert cardiovascular complications, and aggressive surgery for aortic root stenosis is still necessary to prolong survival. We believe our method is useful in this setting.

References