Aortic valve replacement for the calcified ascending aorta in homozygous familial hypercholesterolemia

Tamotsu Yasuda*, Michio Kawasuji, Naoki Sakakibara, Yoh Watanabe

Department of Surgery (I), Kanazawa University School of Medicine, Takaramachi 13-1, Kanazawa 920-8641, Japan

Received 31 January 2000; received in revised form 7 April 2000; accepted 12 April 2000

Abstract

A 72-year-old woman who had been diagnosed as homozygous familial hypercholesterolemia was admitted for chest discomfort. Computed tomography and cardiac catheterization revealed severe calcification of the aortic root and a high grade stenosis of the proximal right coronary artery. Aortic valve replacement concomitant with coronary artery bypass was done using temporary hypothermic circulatory arrest. This is preferred method when dealing the calcified aorta.

Keywords: Calcified aorta; Aortic valve replacement; Coronary artery bypass grafting; Familial hypercholesterolemia

1. Introduction

Aortic valve replacement becomes a high-risk procedure in patients with calcified ascending aorta. We report a patient with homozygous familial hypercholesterolemia (FH) who underwent thromboendarterectomy of the porcelain aortic root, replacement of the calcified aortic valve, and gastroepiploic artery grafting for the right coronary artery stenosis, using temporary hypothermic circulatory arrest to reduce the risk of cerebral emboli.

2. Case report

A 72-year-old woman was admitted for frequent chest discomfort and congestive heart failure. She had been diagnosed as homozygous FH, FH- Tonami-2, based on identification of deletions in the gene for LDL receptor [1]. On admission, she had marked tendon xanthomas of the upper and lower extremities. Serum total cholesterol level was 394 mg/dl. There was no history of neurological event, and computed tomogram of the head revealed a mild brain atrophy. Computed tomogram of the thorax showed severe calcification of the ascending aorta, sinus of Valsalva, aortic valve (Fig. 1), and spotty calcification of the aortic arch. The exact diameters of the ascending aorta and sinus of Valsalva were 30 and 33 mm, respectively. Arteriosclerosis was observed on peripheral arteries, but there were no significant stenoses. Cardiac catheterization showed a high grade stenosis of the proximal right coronary artery, no stenosis of the left coronary artery, and aortic stenosis with a transvalvular pressure gradient of 60 mmHg.

At surgery, she was found to have a calcified aortic root diagnosed as grade IV plaque [2], but the calcification of the aortic arch was not severe as compared to the aortic root. After mobilization of the right gastroepiploic artery, cardiopulmonary bypass was established with femoral artery and bicaval cannulae. The patient was cooled to 16°C, and retrograde cardioplegia and left ventricular venting were used. On circulatory arrest, we found a so-called window 4 cm above the right coronary ostium detected by digital palpation, and the aorta could be entered through it. The ascending aorta was evaluated from inside under direct vision, revealing a totally calcified aortic root, but free floating plaques were not observed. Retrograde cerebral perfusion was performed through the superior vena cava to wash out emboli during circulatory arrest. Limited endarterectomy of the ascending aorta enabled us to find a soft area suitable for aortic cross-clamping, and we concluded that endarterectomy for the transverse aorta downstream from cross-clamp position was not needed. After aortic cross-clamping cardiopulmonary bypass was restarted. The aortic root including the entire Valsalva sinus was endarterectomized, and the calcified aortic valve was excised. A 19-mm SJM hemodynamic-plus valve (St Jude Medical, Inc., MN) was implanted in the supraannular position with interrupted mattress sutures. The aorta was closed with running sutures buttressed with the pericardium and sealed with fibrin glue.
During systemic rewarming, the right gastroepiploic artery was anastomosed to the right posterior descending artery. The patient was weaned from cardiopulmonary bypass without difficulty. The periods of circulatory arrest, aortic cross-clamp, cardiopulmonary bypass were 5, 120 and 240 min, respectively. The patient had an unremarkable recovery. Early follow-up with transthoracic echocardiography 6 months after surgery showed that exact diameters of the ascending aorta and sinus of Valsalva were 31 and 34 mm, respectively. The patient had no neurological event after surgery.

3. Discussion

Emboli from severely atheromatous or calcified aorta have been recognized as the most important factor for stroke after cardiac surgery. Surgical modification such as adjustment of aortic cannulation, no-clamp technique using hypothermic circulatory arrest, pedicle arterial revascularization, aortic graft replacement, or endarterectomy have reduced the incidence of adverse perioperative cerebral events [3]. On circulatory arrest we found so-called window by digital palpation with a lowered systemic pressure, and the aorta could be entered through it. We did not use deep hypothermia for the whole period of aortic valve replacement, because it was pressed for time to excise the severely calcified aortic valve during circulatory arrest. Aortic cross-clamp after limited thromboendarterectomy of ascending aorta was used in place of intra-luminal occlusion by balloon catheter. We think that balloon can lacerate the fragile aortic intima, cause aortic dissection or cerebral embolism. We chose the right gastroepiploic artery as the bypass conduit for ischemia of the inferior wall [4]. Moreover, we used retrograde cerebral perfusion through the superior vena cava to prevent cerebral emboli, and retrograde cardioplegia to avoid coronary emboli or injury of the coronary ostia.

Although the outer adventitial layer left after endarterectomy was fragile, bleeding was well controlled. An aggressive surgical approach to the porcelain aorta using radical thromboendarterectomy, reported by Vogt [5], appears to reduce the perioperative neurologic complications, and endoluminal diameter of the ascending aorta is reported to remain stable up to 44 months after endarterectomy. We consider that it is necessary to observe the long-term results of endarterectomy in patients with severely calcified aorta including this case.

References