

Bicuspid Aortic Valve Stenosis with Single Coronary Artery

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A 56-year-old female with congestive heart failure was transferred to our institution. Aortography demonstrated aortic valve stenosis (AS) with a congenitally bicuspid valve and dilatation of the ascending aorta. Preoperative coronary angiography showed a left single coronary artery. Replacement of the aortic valve and ascending aorta was performed. She had an uneventful postoperative course. We report the case of aortic bicuspid valve stenosis with single coronary artery as an extremely rare congenital cardiac anomaly combination. (Ann Thorac Cardiovasc Surg 2005; 11: 267–9)

Key words: bicuspid aortic valve, single coronary artery, aortic valve stenosis, aortic valve replacement

Introduction

Single coronary artery is a rare congenital anomaly. We describe a patient with bicuspid aortic valve stenosis (AS) with a single coronary artery who underwent replacement of the aortic valve and ascending aorta.

Case Report

A 56-year-old female was admitted to our hospital in 1987, for evaluation of a heart murmur. She had been followed up once a year with a diagnosis of mild AS and the left ventricular pressure gradient to ascending aorta was 30 mmHg. She had one episode of back pain. Echocardiography performed in June 2003 showed a severe AS with a pressure gradient of 100 mmHg. An electrocardiogram demonstrated sinus rhythm and left ventricular hypertrophy. A chest radiograph revealed mild cardiac enlargement with a cardiothoracic ratio of 0.53. Cardiac catheterization showed pulmonary arterial mean pressure of 21 mmHg, pulmonary capillary wedge pressure of 17

mmHg, left ventricular systolic pressure of 280 mmHg, left ventricular end diastolic pressure of 17 mmHg, with ascending aortic systolic pressure of 180 mmHg. The thermolilution cardiac index was 3.65 L/min/m². The left ventricular ejection fraction was 0.70. Aortic root angiography revealed grade 2 regurgitation, bicuspid aortic valve and dilatation of ascending aorta, with a diameter of 45 mm (Fig. 1). The coronary angiogram showed a left single coronary artery (Fig. 2). The operation was performed with cardiopulmonary bypass under the mild systemic hypothermia. After the cross-clamping of the distal ascending aorta, cardiac arrest was accomplished by initial antegrade cold blood cardioplegia via the aortic root and continuous retrograde cold blood cardioplegia. The surgery revealed a bicuspid aortic valve and a left single coronary artery. The two cusps with calcification were severely thickened and retracted. They were excised and replaced with a mechanical valve, and the dilated ascending aorta was replaced with a sealed graft. Histologically, the resected cusps showed fibrotic thickening with calcification. She had an uneventful postoperative course is well 14 months after operation.

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Discussion

The incidence of single coronary artery is reported to be 0.04–0.4% among patients having coronary angiography.^{1,2)} According to the site of origin and anatomical distribution



Fig. 1. Aortic root angiography shows grade 2 regurgitation, bicuspid aortic valve and dilatation of ascending aorta.



Fig. 2. Coronary angiography shows that a single coronary artery arising from the left coronary sinus gives off the anterior descending branch and the left circumflex branch in the usual fashion, and the left circumflex branch gives off the right coronary branch which passes posterior to the aorta and courses in right atrioventricular groove.

of the branches, Smith³⁾ and Lipton et al.²⁾ classified isolated single coronary arteries into three groups and nine subgroups. In our case, the single coronary artery arising from the left coronary sinus gives off the anterior descending branch and the left circumflex branch in the usual fashion, and then the left circumflex branch gives off the right coronary branch which passes posterior to the aorta

and courses in right atrioventricular groove. Therefore this case was classified into Group II, Type L II-P. Single coronary artery has often additional congenital cardiac anomalies. Ogden et al.⁴⁾ reported that 56 cases of 142 single coronary arteries had other congenital cardiac anomalies. Single coronary artery with bicuspid aortic valve was showed in 7 cases of the 56 cases of anoma-

lies. Sclerotic change and calcification of congenital bicuspid aortic valves progresses more rapidly than tricuspid aortic valves.⁵⁾ In this case, AS had progressed over several years.

Wariishi et al.⁶⁾ reported on a case with surgery for bicuspid AS developing ventricular fibrillation after cardiopulmonary bypass and required percutaneous cardiopulmonary support due to insufficient supply of cardioplegic solution at right coronary area because of the high-posterior take-off of the right coronary artery. Preoperative evaluation is important in cases with cardiac anomalies. Cardioplegia in aortic valve surgery with cardiac arrest is performed in antegrade fashion via the aortic root, and if there is aortic valve insufficiency, directly into the coronary ostia. In coronary artery anomalies, however, retrograde blood cardioplegia may be effective. In our case, cardiac arrest was accomplished by initial antegrade cold blood cardioplegia via the aortic root, and it was maintained by continuous retrograde cold blood cardioplegia.

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