Co-Existence of Severe Coarctation of the Aorta and Aortic Valve Stenosis in a 65-Year-Old Woman: A Case Report

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Coarctation of the aorta is usually diagnosed and corrected early in life. Survival to more than 60 years of age of a patient with unrepaired coarctation of the aorta is extremely unusual, and the optimal management strategies for such patients are controversial. We describe the case of a woman who was first diagnosed as having coarctation of the aorta and aortic valve stenosis at the age of 65 years and underwent successful aortic valve replacement.

Keywords: coarctation of the aorta, bicuspid aortic valve, aortic valve stenosis

Introduction

Coarctation of the aorta is a congenital vascular lesion typically diagnosed and corrected in childhood or early adulthood. Most untreated patients with coarctation of the aorta die before the age of 50 years, and very few survive to old age. There are a few case reports of patients first diagnosed as having coarctation of the aorta after 60 years of age, but their management remains controversial. We report the case of a woman who was first diagnosed as having coarctation of the aorta and aortic valve stenosis at the age of 65 years and underwent successful aortic valve replacement.

Case Report

We report the case of a 65-year-old woman who was diagnosed as having hypertension in her mid-thirties, in whom the blood pressure was, however, well-controlled with a combination of antihypertensive agents (angiotensin II receptor antagonist, calcium channel blocker). She was referred to our hospital because of nocturnal dyspnea. Clinical examination revealed a blood pressure of 115/60 mmHg in the right arm and 123/63 mmHg in the left arm. The lower extremity blood pressure was 78/51 mmHg, with a marked radio-femoral delay. A systolic ejection murmur was heard at the right upper sternal border. Electrocardiography revealed sinus rhythm and changes suggestive of left ventricular hypertrophy. An enlarged cardiac silhouette and rib notching were observed on the plain chest X-ray (Fig. 1). Echocardiography revealed severe systolic dysfunction, with an ejection fraction of only 34%. The aortic valve was bicuspid with a peak/mean transvalvular pressure gradient of 61/32 mmHg, and the valve area was reduced to 0.4 cm².

Therefore, aortic valve replacement was planned and cardiac catheterization was performed preoperatively to evaluate the heart function. It was impossible to engage a catheter into the coronary artery ostium via the right radial artery because of a severely tortuous brachiocephalic artery, and total occlusion of the descending aorta was detected when approach via the right femoral artery was attempted. Computed tomography (CT) angiography clearly showed complete occlusion of the descending aorta with extensive collateral circulation; there was no significant coronary artery disease (Fig. 2).

Successful aortic valve replacement was performed. After a median sternotomy, cardiopulmonary bypass was established with double-arterial cannulation in the
Co-Existence of Severe Coarctation of the Aorta and Aortic Valve Stenosis

ascending aorta and right femoral artery, to ensure adequate perfusion proximal and distal to the coarctation. The distal ascending aorta was cross-clamped, and antegrade cold blood cardioplegia was used to induce cardiac arrest. After incision of the ascending aorta, the calcified bicuspid aortic valve was excised, and a stented bovine pericardial prosthesis (21 mm, Carpentier-Edwards PERIMOUNT Magna Ease; Edwards Life-sciences, Irvine, California, USA) was implanted. The cardiopulmonary bypass and aortic cross-clamp times were 95 min and 69 min, respectively. We propose to correct the coarctation in a second operation, if needed.

The postoperative course was uneventful. Six months after the surgery, significant reduction of the plasma levels of BNP from 438 pg/mL to 62 pg/mL was observed. Currently, the patient is asymptomatic, and her blood pressure remains well-controlled under a combination of antihypertensive agents (angiotensin II receptor antagonist, calcium channel blocker, and very low dose of beta-blocker).

Discussion

Coarctation of the aorta is a congenital vascular lesion typically diagnosed in early life, accounting for 5% to 10% of all congenital cardiovascular malformations. It is frequently associated with other congenital defects, and as many as 50%–85% of patients have a bicuspid aortic valve. The natural history of unrepaired coarctation of the aorta includes the development of systemic hypertension and subsequent morbidity and death from cardiovascular disease. Most untreated patients with coarctation of the aorta usually die before 50 years of age.

Death in these patients is usually secondary to heart failure, coronary artery disease, aortic rupture/dissection, concomitant aortic valve disease, infective endarteritis/endocarditis, or cerebral hemorrhage.

Cohen, et al. reported the long-term outcomes after repair of coarctation of the aorta. They identified age at the time of the repair as a significant factor determining the long-term outcomes of these patients: the outcomes improved as the patient’s age at the initial operation decreased (until the age of 9 years). This could be explained by the fact that operation at an early age is associated with a reduced incidence of postoperative relapse of hypertension, thereby leading to a lower incidence of complications such as coronary artery disease, which remain the primary causes of death in patients with coarctation of the aorta. While the patients ranged widely in age from 1 week after birth to 72 years old in their study, the results covered only patients who underwent repair for coarctation of the aorta, while excluding patients who underwent simultaneous surgery or surgery for concomitant heart disease alone. As such, while there are reports on the long-term outcomes after surgery for coarctation of the aorta, there are only scarce reports of the outcomes in elderly patients aged 65 years or older and the long-term outcomes of elderly patients with aortic coarctation is unknown. Therefore, there is still no consensus about the best surgical approach for the management of elderly patients with coarctation of the aorta and concomitant cardiac lesions; whether a single-stage or 2-stage surgery should be performed, and whether the coarctation or the intracardiac lesion should be repaired first.

Our patient reported herein had coarctation of the aorta with a poorly functioning heart associated with severe aortic valve stenosis, for which surgery was indicated. Preoperatively, the patient showed well-controlled blood pressure and no claudication symptoms in the lower limbs. However, the outcome of the patient was considered to be influenced by the evidence of cardiac failure associated with the low left ventricular ejection fraction. As severe aortic valve stenosis was suspected to be the cause rather than the coarctation of the aorta, it was decided that aortic valve replacement would be carried out first to improve the outcome of the patient.

The next problem that we faced was whether it was necessary to simultaneously undertake repair of coarctation of the aorta. While the blood pressure was favorably controlled in the patient at the time of the surgery, it
could become potentially difficult to control, at least in part, due to resolution of the aortic valve stenosis in the future. There is the possibility that with the improvement of the cardiac failure after surgery, the consequent improvement of the ADL may precipitate the onset of claudication symptoms in the lower limbs. Therefore, we would consider simultaneous surgery if a low-invasive technique were available.

In order to perform anatomic bypass surgery, it is necessary to add a lateral thoracotomy. Therefore, there are such issues as excessive surgical invasiveness and complex surgical procedures requiring postural change during surgery. In addition, there is the risk of the operation becoming difficult due to adhesions when a second surgery is required. With the median sternotomy approach alone, it is difficult to perform anatomic bypass surgery for patients with coarctation, and only extra-anatomic bypass surgeries such as ascending-to-descending aortic bypass and axillo-bifemoral bypass may be possible. As the latter has a problem of long-term graft patency, it may have been appropriate to perform ascending-to-descending aortic bypass in this patient. As the latter has a problem of long-term graft patency, it may have been appropriate to perform ascending-to-descending aortic bypass in this patient. There are several reports of the outcomes of this operative procedure indicating favorable operative results. However, Pethig, et al. reported heart failure and life-threatening ventricular arrhythmias in patients with one-stage aortic valve repair and extra-anatomic bypass grafting for coarctation of the aorta. These complications may be attributable to global myocardial ischemia and impaired coronary blood supply in the hypertrophied hearts with a low perfusion pressure.

As the patient had poor heart function, there was concern about fatal arrhythmias being induced by the decreased coronary blood flow after resolution of the aortic coarctation. Considering that the blood pressure was controlled without difficulty at the time of the surgery, we decided to carry out aortic valve replacement alone. Fortunately there was no notable complication in the perioperative period, and the postoperative blood pressure control was also favorable. In regard to the heart function, the ejection fraction improved from 34% before the operation to 62% at 1 year after the operation. It is necessary to continue to carefully observe the patient and to evaluate whether the patient would become a candidate for surgery for coarctation of the aorta.

**Conclusion**

We report the case of a woman who was first diagnosed as having coarctation of the aorta and aortic valve stenosis at the age of 65 years and underwent a successful aortic valve replacement. It will be necessary to examine whether one-stage operation or two-stage operation might be suitable in such patients, depending on the condition of the individual patients.
Disclosure Statement

We have no conflicts of interest to disclose.

References