Single-Stage Repair of Thoracic Aortic Aneurysm through a Median Sternotomy in a Patient with Pseudocoarctation of the Aorta and Severe Aortic Valve Stenosis

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Introduction

Pseudocoarctation of the aorta is an uncommon anomaly characterized by congenital elongation and kinking of the aorta. Pseudocoarctation of the aorta is associated with other congenital cardiac anomalies and aneurysm formation of the thoracic aorta. Here, we present a case of pseudocoarctation of the aorta associated with thoracic aortic aneurysm (TAA) and severe aortic valve stenosis (AS).

Case

A 72 year-old woman was admitted to our hospital because of congestive heart failure (CHF) with severe AS. On admission, her blood pressure was 80/42 mmHg, and her pulse was regular with a rate of 82 bpm. Blood test showed elevation of the brain natriuretic peptide (1577 pg/ml). A chest X-ray showed pulmonary congestion, cardiac enlargement and widening of the superior mediastinum. The results of transthoracic echocardiography showed an ejection fraction of 30%, a peak gradient across the aortic valve of 93 mmHg and an effective orifice area of 0.55 cm². Enhanced computed tomography (CT) performed subsequently revealed kinking in the distal aortic arch with stenosis (Fig. 1A), poststenotic dilatation of the descending aorta (45 mm), a left subclavian artery aneurysm (30 mm) and no significant collateral vessels (Fig. 1B). There was no systolic pressure gradient between the upper and lower extremities. As descending aorta tended to expand, we thought that the aneurysm was needed for resection. After treatment of CHF, we performed single-stage surgery through a median sternotomy (Fig. 1C). Total cardiopulmonary bypass was initiated by cannulating the ascending aorta. The common femoral artery was isolated and cannulated with an 18 Fr cannula. After the ascending aorta was clamped and cardiac arrest was obtained, the ascending aorta was opened. We found a bicuspid aortic valve and performed aortic valve replacement (AVR) with a 21-mm bovine pericardial bioprosthetic aortic valve. A wide incision of the left pleura was made from the aortic arch to the diaphragm. The left lung was mobilized to the right side and the narrowed segment and descending aortic aneurysm were clearly visible when the heart with the pleura and pericardium were retracted. After the patient was cooled to 28°C and the descending aorta was clamped, antegrade cerebral perfusion and femoral artery perfusion were initiated (Fig. 2A). The aneurysm wall was thinned and diameter of the narrowed segment was 9 mm (Fig. 2B). The distal aortic anastomosis was completed with a four-branched woven Dacron graft. After antegrade flow to the lower body was established through a side branch of the graft, the ascending aorta, brachiocephalic and carotid arteries were anastomosed. The third graft limb was anastomosed to the left axillal artery through the pleural cavity and the left subclavian artery aneurysm was resected and...
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The pathological examination of the resected aortic aneurysm under H&E staining revealed a dilated aortic wall with arteriosclerosis. Her postoperative course was uneventful, and she had no complications at the time of discharge.

Discussion

Pseudocoarctation of the aorta is an uncommon anomaly characterized by congenital elongation and kinking of the aorta. The four criteria for the diagnosis of pseudocoarctation: an abnormal posterolateral chest radiograph, less than 25 mmHg pressure gradient between the upper and lower extremities, no evidence of increased collateral circulation or rib notching, and a diagnostic aortogram.

Pseudocoarctation of the aorta has been described as a benign entity warranting no specific therapy. However, some authors suggested that pseudocoarctation of the aorta should not be regarded as a benign condition because it is sometimes associated with aneurysm formation of the thoracic aorta, which may cause sudden rupture or dissection. Therefore, surgery is recommended for all symptomatic patients and those with aneurysm formation. The etiology of aneurysm formation is considered to be anomalous development of the aorta in association with pseudocoarctation, with secondary arteriosclerotic changes in the intima. Furthermore, turbulent flow through the narrowed segment results in damage to the arterial wall with subsequent aneurysm formation.

In this case, AVR and aneurysm resection were needed because the patient was diagnosed with pseudocoarctation of the aorta associated with growing aneurysm of the descending aorta and severe AS. Endovascular aneurysm repair is not appropriate for our patient because of the complex kinking and tortuosity of the aorta. We felt that performing a median sternotomy along with a left thoracotomy or anterolateral partial sternotomy was too invasive; therefore, we performed just a median sternotomy and used our “pleural-window approach.”

Fig. 1  Preoperative enhanced computed tomography. Computed tomography revealed kinking in the distal aortic arch with stenosis (white arrow), poststenotic dilatation of the descending aorta, left subclavian artery aneurysm and no significant collateral vessels (B). We performed the descending aortic aneurysm resection, the left subclavian artery aneurysm resection and aortic valve replacement.

Fig. 2  A: Surgical schema. After the patient was cooled to 28°C and the descending aorta was clamped, antegrade cerebral perfusion and femoral artery perfusion were initiated (black arrow). B: Surgical view through the wide incision of the left pleura. The aneurysm wall was thinned and diameter of the narrowed segment was 9 mm (arrow head).
In previous reports of pseudocoarctation of the aorta with TAA, left thoracotomy was performed to approach the TAA. However, there are no reports of pseudocoarctation with descending aortic aneurysm in which a concomitant cardiac procedure is required. We conjecture that this is the first case of pseudocoarctation in which aneurysm resection and AVR were performed as a single-stage procedure through only a median sternotomy using our “pleural-window approach.”

We think that this approach could be a good option for descending aortic aneurysm resection and concomitant cardiac surgery in similar cases.

Disclosure Statement
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