Case report - Vascular thoracic

Normothermic total arch replacement without hypothermic circulatory arrest to treat aortic distal arch aneurysm in a patient with cold agglutinin disease

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Abstract

Cold agglutinin disease although rare, can lead to serious complications for patients undergoing cardio-thoracic surgery, especially when cardiopulmonary bypass is applied under hypothermic circulatory arrest. We describe normothermic total arch replacement without hypothermic circulatory arrest in a patient with cold agglutinin disease. The patient tolerated all procedures well and did not develop cerebral ischemia due to surgical maneuvers or thrombotic or haemolytic complications due to cold agglutinin disease. Although endovascular aortic repair is the first choice under such complex conditions, this method could also serve as an alternative strategy when endovascular aortic repair is precluded.

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1. Introduction

Total arch replacement with cardiopulmonary bypass under hypothermic circulatory arrest is usually safe. The results of these procedures have recently been improved with fewer complications when combined with selective antegrade or retrograde cerebral perfusion. However, hypothermia causes serious complications for patients with cold agglutinin disease as hemagglutination occurs at low temperature followed by hemolysis during rewarming [1]. We describe normothermic total arch replacement without hypothermic circulatory arrest to treat an aortic distal arch aneurysm concomitant with cold agglutinin disease.

2. Case report

A 71-year-old male was referred to our hospital with an aneurysm of the aortic distal arch (Fig. 1a, b). Coronary-pulmonary artery fistulae, partial anomalous pulmonary venous return and paroxysmal atrial fibrillation with a history of congestive heart failure were also preoperatively diagnosed.

The preoperative routine work-up showed an abnormal clotting reaction and further work-up revealed a cold autoantibody and cold agglutination that started at 25 °C, although this patient was asymptomatic regarding hemagglutination or hemolysis. Although the cold agglutinin titer was not measured, we diagnosed this patient as having cold agglutinin disease and therefore, reconstructed the cervical branches under a beating heart and performed total arch replacement under normothermic partial extracorporeal bypass without hypothermic circulatory arrest.

With the patient in the supine position, grafts were anastomosed on each bilateral axillary artery via small subclavian skin incisions and pushed into the thorax through the intercostal spaces. After changing the patient's position to the left hemilateral decubitus, the clamshell approach proceeded via a thoracotomy in the left fourth and right third intercostal spaces with a transverse sternotomy. A trifurcated customized graft fashioned from a bifurcated prosthetic graft for abdominal aortic repair was anastomosed on the partially clamped ascending aorta (Fig. 2a), and each cervical branch was sequentially reconstructed under the beating heart without cardiopulmonary bypass. The middle branch was connected to the graft from the right axillary artery to maintain temporary cerebral circulation from the right carotid and right vertebral arteries during permanent anastomosis of the brachiocephalic artery and the right branch (Fig. 2b). The right branch was anastomosed to the brachiocephalic artery, and the left branch was connected to the graft from the left axillary artery while maintaining cerebral circulation from the left vertebral artery.
The branch connected to the graft from the right axillary artery was finally, disconnected and re-anastomosed to the left carotid artery (Fig. 2d). After dislocating the aortic arch, a partial extracorporeal bypass was established with cannulation on the descending aorta and bivalve drainage of the peripheral circulation. A 24-mm straight graft was anastomosed to the descending aorta and pulled through inside the aneurysm, followed by proximal anastomosis (Fig. 2e). Because the vein with partial anomalous pulmonary venous return adhered to and was located across the wall of the aortic aneurysm, it was not opened. Thus, the aortic arch was replaced under cerebral perfusion with autologous cardiac output. After completing the total arch replacement, the coronary-pulmonary artery fistulae were resected and the right atrium was cryoablated.

The patient tolerated all procedures well and was discharged without complications, such as cerebral ischemia due to surgical maneuvers or thrombosis or haemolysis due to cold agglutinin disease (Fig. 1c).

3. Discussion

Total arch replacement under cardiopulmonary bypass with hypothermic circulatory arrest has become a standard type of aortic arch surgery. In addition, combining these procedures with selective antegrade or retrograde cerebral perfusion has reduced operative risk for neuropsychological deficits. However, surgery under hypothermic circulatory arrest is occasionally contraindicated under circumstances, such as cold agglutinin disease.

Cold agglutination is a primary or acquired autoimmune disease that involves autoantibodies that lead to hemagglutination at low temperature followed by hemolysis during rewarming. The critical temperature at which this condition is elicited should be determined during preoperative evaluation. Operative strategies, such as preoperative steroid administration and plasmapheresis [2], surgery under systemic normothermia or mild hypothermia, and cardiac arrest with warm cardioplegia above the critical temperature [3] should be considered. Valvulopathy and coronary artery disease can be surgically treated under normothermic or mildly hypothermic conditions, the thermal amplitude of which is higher than that required for antibody activation [4]. However, patients with cold agglutinin disease are considered intolerant of hypothermic surgery.

Total arch replacement under cardiopulmonary bypass with hypothermic circulatory arrest is a standard procedure at our institution, and patients with conditions, such as cold agglutinin disease who cannot tolerate hypothermic procedures are rare. Repair of the simple distal aneurysm was considered in the present patient, but the form of this aneurysm and the adherent vein seemed to threaten the allowance of the cross clamp and the seam for anastomosis. We therefore, proceeded with normothermic complete arch repair without circulatory arrest. Several reports have described normothermic total arch replacement under extracorporeal bypass without circulatory arrest [5, 6]. We reconstructed the cervical branches while maintaining the cerebral circulation with autologous cardiac output, which is a novel technique.

Alternatively, endovascular repair with debbranching or revascularization of the aortic branches should be considered for high-risk patients, but endovascular repair of the thoracic aorta was still rare in Japan when we operated...
on the present patient. Endovascular repair is becoming accepted and early results from selected patients have been very encouraging [7, 8]. We normally perform endovascular repair for such patients because this method has now become routine at our institution. Endovascular repair of aortic arch lesions should be further refined to improve long-term results and become a standard alternative strategy for unusual situations. We believe that our method represents an alternative strategy with which to treat atypical patients for whom an endovascular approach is precluded by the presence of a severely atherosclerotic, calcified, tortuous and occluded aorta.

References


Fig. 2. Total arch replacement after reconstruction of cervical branches. Pre, Preoperative status of thoracic aorta. (a) Grafts anastomosed to bilateral axillary arteries and proximal anastomosis of trifurcated graft on ascending aorta using partial clamp. (b) Connection of middle branch to graft anastomosed to right axillary artery. (c) Connection of left branch to graft anastomosed on left axillary artery and anastomosis of right branch with brachiocephalic artery. (d) Disconnection and anastomosis to middle branch of left carotid artery. (e) Total arch replacement with partial extracorporeal circulation.