Case Report

A Case of Acute Type A Aortic Dissection with Double Aortic Arch

Katsuaki Tsukioka, MD, PhD,[1] Tetsuya Kono, MD, PhD,[2] and Kohei Takahashi, MD, PhD[1]

Double aortic arch (DAA) is extremely rare in adults. A 71-year-old woman suffered from syncope, and an acute-type aortic dissection with a DAA accompanied by a massive pericardial effusion was shown in a non-enhanced computed tomography (CT). Enhanced CT was not performed because of her hemodynamic instability. She was rushed to the operating theater after immediate pericardiocentesis without more precise morphological evaluation. Ascending aortic replacement was performed by clamping both aortic arches without systemic circulatory arrest. She survived the operation, but her respiratory function was affected by tracheomalacia and remaining DAA with residual dissection.

Keywords: double aortic arch, aortic dissection, tracheomalacia

Introduction

While the true occurrence of a double aortic arch (DAA) in adults is unknown, a right-sided aortic arch occurs at 0.1% in adults accompanied with a high possibility of a complete vascular ring, in which DAA is thought to be the most common form.[1,2] In adult cases of DAA, surgical intervention has been reported due to respiratory complaints, dysphagia, or other symptoms. Clinical presentation of an acute type A aortic dissection (ATA-AD) in DAA is extremely rare, and to the best of our

knowledge, only two cases of surgical intervention have been described so far.[3,4] Precise morphological study by enhanced computed tomography (CT) was performed in the previous two cases, and the location and characteristics of the false lumen were identified. Replacement of the ascending aorta was also performed. Development of a postoperative respiratory disorder is also often described in adult cases of vascular ring,[1,2,5] derived from tracheal remodeling tracheomalacia, which develops secondary to long-standing compression by DAA.

In this report, we describe the surgical strategy for an ATA-AD with an extraordinal anatomy in a patient with critical status and uncleared morphological study, and present how we managed her postoperative respiratory dysfunction.

Case Report

A 71-year-old woman complained of syncope and was transferred to a neighboring hospital. She had a medical history of appendectomy and hypertension, and no persisting respiratory symptoms or dysphagia was documented. Endotracheal intubation was performed because of her hemodynamic instability and altered consciousness. Thereafter, a non-enhanced CT revealed an ATA-AD with a DAA accompanied by a massive pericardial effusion (**Fig. 1a**). The false lumen was not extending to the descending aorta (**Fig. 1b**). She was then referred to our hospital. On her arrival, her systolic blood pressure was 50 mmHg and pulse rate was 88 beats per min with a high dose of inotropic support. Her Glasgow Coma Scale score was E3VTM6. Neither paralysis nor difference in limb blood pressure was observed. Laboratory test showed severe thrombocytopenia with a platelet count of 23×10^9/L. Enhanced CT was not performed because of her hemodynamic instability, and she was rushed to the operating theater after an immediate pericardiocentesis at the emergency bay to prevent sudden cardiac arrest. After induction of general anesthesia, median sternotomy was performed. Transesophageal echocardiography revealed no cardiac anomalies, a good left ventricular contraction and a competent aortic valve. Cardiopulmonary bypass was established by cannulation to the left femoral artery and bicaval venous drainage,

[1] *Department of Cardiovascular Surgery, Iida Municipal Hospital, Iida, Nagano, Japan*
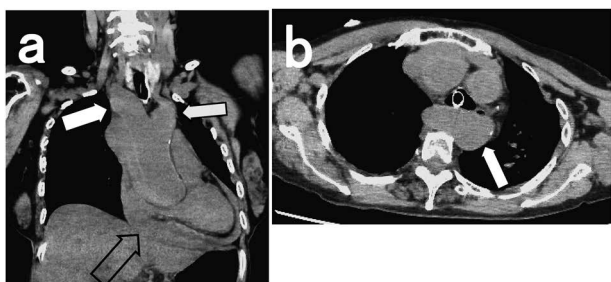[2] *Department of Cardiovascular Surgery, Suwa Red Cross Hospital, Suwa, Nagano, Japan*

Fig. 1 Preoperative computed tomography.
(**a**) The false lumen was seen in the ascending aorta and both aortic arches (open arrow). A massive pericardial effusion resulted in cardiac tamponade (closed arrows). (**b**) The false lumen was not extending to the descending aorta. The preoperative diameter of the right aortic arch was 32 mm.
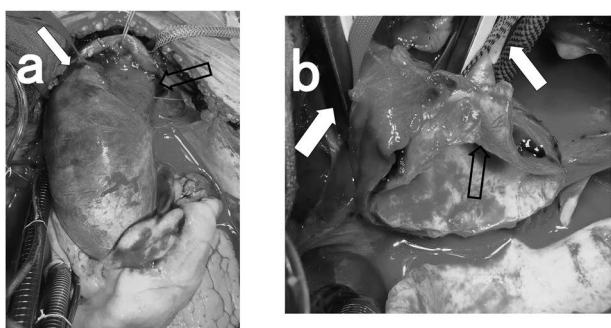

Fig. 2 Intraoperative findings.
(**a**) The left aortic arch was free from dissection (open arrow), but the right aortic arch was affected by dissection (closed arrow). (**b**) After clamping both aortic arches (closed arrows), we found an intimal tear in the distal end of the ascending aorta (open arrow).

Fig. 3 Postoperative computed tomography.
(**a**) A new communicating dissection appeared from the right aortic arch (RAA) (open arrow) to the descending aorta (closed arrow). (**b**) An intimal tear was seen near the distal anastomosis (open arrow). The postoperative diameter the RAA increased to 35 mm.

and systemic cooling was initiated subsequently. Intraoperatively, we found that the left aortic arch was free from dissection, but the right aortic arch (RAA) was affected (**Fig. 2a**). We feared that malperfusion to the right common carotid artery branching off from the RAA might occur if we clamped the RAA; thus, we added another arterial perfusion to the right axillary artery branching off separately from the RAA. After clamping both aortic arches at a bladder temperature of 28°C, significant decline in neither direct measurement of blood pressure of the left radial and the right dorsalis pedis arteries nor cerebral oxygenation monitored by near-infrared spectroscopic oximetry (NIRO-200, Hamamatsu, Herrsching, Germany) was observed. Confirming that malperfusion was avoided through these findings, we decided to avoid deep hypothermic circulatory arrest (DHCA) with retrograde cerebral perfusion. Systemic cooling was also discontinued. During the resection of the ascending aorta without opening both aortic arches, systemic perfusion was not interrupted. We found an intimal tear in the distal end of the ascending aorta (**Fig. 2b**) and the distal anastomosis with a 26-mm
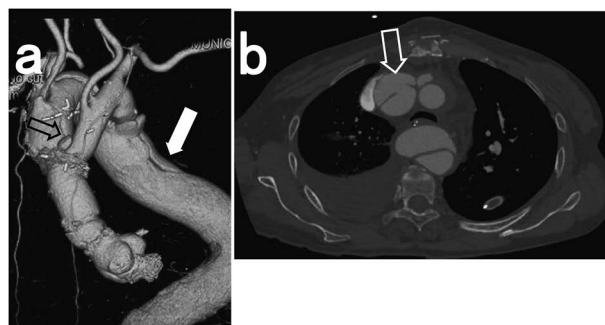
J-graft (Japan Lifeline, Tokyo, Japan) was performed excluding the intimal tear. Then, we removed the femoral arterial cannulation and resumed antegrade reperfusion through an epiaortic branch of the graft followed by ascending aorta replacement. After extubation on postoperative day (POD) 4, no neurological deficiencies were noted, but she developed respiratory dysfunction accompanied by a severe strider, which demanded transit support of non-invasive positive pressure ventilation (NPPV) for 5 days. She was discharged on POD 60. A new communicating dissection from the RAA to the descending aorta was shown on postoperative CT on POD 11. An intimal tear was found near the distal anastomosis (**Fig. 3a**) and the preoperative diameter of 32 mm (**Fig. 1b**) of the RAA increased to 35 mm (**Fig. 3b**).

Discussion

The true occurrence of DAA in adults is unknown, but it is reported to be extremely rare because it tends to cause severe respiratory symptoms and results in early diagnosis and correction in infancy or childhood.[2] Only two cases on the repair of the aortic dissection in DAA have been also reported thus far.[3,4] Although we resected the primary intimal tear in the ascending aorta, a new communicating dissection with a new intimal tear in the RAA near the anastomosis appeared postoperatively. This indicated that a clamp injury might have occurred. While we kept clamping both aortic arches, in a previous case reported by Zeigler et al.,[3] retrograde cerebral perfusion under DHCA was used during open distal anastomosis. In principle, cross-clamping on the dissecting aorta should be avoided, especially in cases of retrograde femoral perfusion because it might generate new intimal tears other than the clamp site causing new false lumens and malperfusion.[6] This patent false lumen can cause dilatation of

the residual aorta and affect late prognosis,[7] demanding close observations in the long term. Further, Geirsson et al. reported that the open distal anastomosis technique in ATA-AD resulted in better mid-term survival compared to the clamp-on technique.[8] However, our patient had a severe thrombocytopenia resulting from presumable consumption coagulopathy on admission, which also indicated increased mortality in ATA-AD patients.[9] We found that DHCA is not preferred for our patient because it might further induce perioperative coagulation problems. Further, we thought that selective antegrade cerebral perfusion (SACP) was also unfeasible because we could not obtain a precise cannulation design due to insufficient evaluation of the morphology of cerebral branches as preoperative-enhanced CT was not performed. Moreover, postoperative-enhanced CT revealed that all cerebral arteries branched off separately from both aortic arches. Thus, SACP cannulation to each ostium would have been less feasible and would prolong the duration of distal anastomosis. This could be disadvantageous for such a critical patient with preoperative hemodynamic instability. Fortunately, we could avert malperfusion after clamping both aortic arches and preserve systemic perfusion that could mitigate adverse ischemic effects to the organs. We could also perform distal anastomosis without distortion or crowding at the aortic wall because clamps to both aortic arches could be located distantly from the distal end of the ascending aorta (**Fig. 2b**), which can be hardly expected in ordinary ATA-AD cases. However, the open distal technique with cerebral perfusion would have been inevitable if malperfusion had occurred when we clamped the dissected vascular ring.

With regard to her postoperative respiratory function disturbance, her long-term exposure to the direct compression from the vascular ring could presumably result in the weakness of the trachea, also called tracheomalacia,[1,2,5] which induced preexisting susceptibility to airway collapse caused by unfavorable factors such as transtracheal pressure differences, postoperative tissue edema, and convective acceleration derived by compressed segments. As shown in **Fig. 3**, the postoperative dilatation of RAA could result in an additional tracheal compression and contribute to the stridor, which became apparent after extubation. She overcame this situation by NPPV support for 3 days, which might reduce her respiratory effort and, consequently, turbulence, worsening airway resistance. The NPPV support was removed with decrease in tracheal edema caused by systemic inflammatory fluid accumulation. However, the remaining communicating dissection in RAA would also cause respiratory disorder or dysphagia resulting from the compressive effects of the dilated aortic arches in the late postoperative course. In these circumstances, surgical interventions such as resection or division

of the RAA through right lateral thoracotomy might be performed for her long-term follow-up.

Conclusion

Although we salvaged a patient severely affected by an extremely rare ATA-AD of DAA without using open distal anastomosis by clamping both aortic arches, the residual aortic dissection with an intimal tear and a patent false lumen would cause not only aortic events but also respiratory or esophageal symptoms related to the vascular ring.

Disclosure Statement

The authors have no conflicts of interest.

Author Contributions

Study conception: KT
Data collection: KT
Analysis: KT
Investigation: KT
Funding acquisition: KT
Critical review and revision: all authors
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Accountability for all aspects of the work: all authors

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