» **Case Report** «

Successful Surgical Exclusion of Rapidly Expanding Kommerell Diverticulum Following a Total Arch Replacement for an Acute Type A Aortic Dissection

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A 50-year-old man presented with an acute type A aortic dissection with an aberrant right subclavian artery. Emergent total arch replacement with an elephant trunk was performed. Intraoperatively, the origin of the aberrant right subclavian artery could not be resected because it was located too far from the distal arch. After two weeks, the patient became aware of dysphagia. Postoperative computed tomography showed the esophagus was compressed anteriorly by the aneurismal origin of this aberrant vessel (Kommerell diverticulum) with a patent false lumen. Additional replacement of the descending aorta via left thoracotomy was performed immediately to exclude a Kommerell diverticulum.

Keywords: aorta/aortic, aortic arch, aortic dissection

Introduction

A left aortic arch with an aberrant right subclavian artery (ARSA) is the most common aortic arch anomaly even though it occurs in approximately 0.5% of the general population.^{1,2)} It has been recommended that resection of the origin of an aberrant subclavian artery as well as reconstruction of the vessel should be performed during the initial surgical intervention for the aneurysm or dissection of the adjacent descending thoracic aorta.^{3–5)} In an acute type A aortic dissection (TAAD), the above-mentioned procedures are difficult and challenging because of the potentially catastrophic condition and the distal location of this anomaly. In view of survival, replacement of

the ascending aorta and total arch for TAAD is an acceptable procedure, even if the ARSA cannot be resected. However, the dissected origin of the ARSA is prone to dilate to an aneurismal formation (Kommerell diverticulum) that will cause a rupture or an aorto-esophageal fistula.⁶⁾ In this paper, we report a patient with dysphagia after a palliative total arch replacement for a TAAD with an ARSA. The patient successfully underwent a second-stage repair for a Kommerell diverticulum before suffering from critical complications.

Case Report

A 50-year-old man had sudden chest pain and was brought to our hospital. The patient did not present with dysphagia or respiratory discomfort and the aortic anomaly was not detected before this event. Enhanced computed tomography (CT) revealed a TAAD from the ascending aorta down to the left iliac arteries with a false lumen remaining patent. CT also showed an ARSA originating from the proximal descending aorta as the fourth aortic arch branch that passed behind the esophagus and trachea (Fig. 1A).

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Fig. 1 (A) Preoperative enhanced CT revealed an acute aortic dissection type A with an ARSA originating from the proximal descending aorta as the fourth aortic arch branch. (B) Postoperative CT showed a patent false lumen and an esophagus compressed anteriorly by a Kommerell diverticulum (arrows). CT: computed tomography; ARSA: aberrant right subclavian artery.

Emergent total arch replacement was performed via a median full sternotomy. Cardiopulmonary bypass was established with the arterial inflow through the artificial conduit anastomosed to right axillary artery in end to side fashion and the cannula inserted into the right femoral artery, and drainage from the right atrium. Under circulatory arrest, the ARSA was ligated on the right side of the trachea, and the left subclavian artery was also ligated at its origin from aortic arch. Then antegrade selective cerebral perfusion was started through artificial conduits anastomosed to the right and left axillary arteries, and balloon-tipped catheters into the right and left common carotid arteries. The intimal tear could not be found in the ascending aorta and aortic arch. The open distal arch anastomosis was performed using a modified elephant trunk procedure. The elephant trunk anastomosis was placed between the left subclavian artery and ARSA. The artificial conduit to the right axillary artery was anastomosed to a side branch of a quadrifurcated graft. The patient recovered without any neurological complications but he became aware of dysphagia in the short period of 2 weeks after the operation. Postoperative CT revealed wrinkling of the elephant trunk graft, a patent false lumen, and the esophagus compressed anteriorly by a dilated blind-sac of the ARSA (Kommerell diverticulum) that measured 25 mm

in diameter (**Fig. 1B**). Because of the potential for a rupture of the Kommerell diverticulum, the second-stage repair was performed immediately via a left thoracotomy. A partial cardiopulmonary bypass was established with return into the right femoral artery and drainage from the right atrium using a long cannula inserted into the right femoral vein. The entry site was identified at the orifice of the ARSA. The distal circumferential graft anastomosis was placed between Th7 and Th8 and was carried out to perfuse into both the true and false lumen. After completion of the distal anastomosis, the graft was sutured end-to-end to the elephant trunk graft. The postoperative course was uneventful. The patient became asymptomatic after the second-stage procedure. Postoperative CT revealed complete exclusion of the Kommerell diverticulum (**Fig. 2**).

Discussion

Burckhard F. Kommerell described an aortic diverticulum in 1936 in a patient with a left aortic arch and ARSA,¹⁾ which is a rare congenital abnormality of the aortic arch and its branches (prevalence of 0.5% in the normal population²⁾). It is considered an anomaly of the 4th aortic arch and results from abnormal regression of the right aortic arch between the carotid

Fig. 2 Postoperative transverse and coronal enhanced computed tomography after a two-stage repair revealed complete exclusion of the origin of the aberrant right subclavian artery (ARSA) (arrows).

and subclavian arteries. In the most common type, the right subclavian artery arises as the last branch of the aortic arch and travels by the proximal descending aorta to the right arm, passing behind the esophagus.³⁾ Although a patient with an aberrant subclavian artery may be asymptomatic, this anomaly may cause catastrophic complications such as dissection or rupture of an aneurysm. Cina, et al. reported the rate of dissection or rupture was 53% among 32 patients in a case review.³⁾ Kieffer, et al. reported an acute aortic dissection occurred in 2 of 33 adult patients with this anomaly.⁴⁾

The surgical strategy for a TAAD with an ARSA has not been standardized because of the rarity of this anomaly. The key component in a TAAD is replacement of the dissected ascending aorta or ascending to aortic arch including the entry site. In a patient with an ARSA, however, resection of the entry site is extremely difficult during initial repair because the entry usually exists around an orifice of the ARSA and is located far from the distal arch. To facilitate the second-stage repair, we performed a total arch replacement with an elephant trunk of which an anastomosis was performed between the left subclavian artery and ARSA. Unexpectedly, he felt dysphagia in a brief period of postoperative day 14 because of esophageal compression by a Kommerell diverticulum that measured 25 mm in diameter. The size above which intervention should be recommended in mildly symptomatic patients has not been clearly established. Cina, et al. recommended intervention for aneurysms 30 mm or greater in diameter³⁾ and Ota, et al. recommended surgical treatment for aneurysms 50 mm or greater.⁵⁾ However, Agematsu, et al. reported a patient

that had undergone the palliative procedure for a TAAD died postoperatively from a rupture of the dissected Kommerell diverticulum and esophageal bleeding through an aorto-esophageal fistula despite the diverticulum size being less than 30 mm in the maximum diameter.⁶⁾ Therefore, we decided that the diverticulum should be treated immediately with additional intervention before potential critical complications.

Although the early and mid-term results of the endovascular repair for distal aortic dissection appear favorable, controversy exists regarding open surgery versus endovascular repair in patients that require intervention, especially those with chronic distal aortic dissection disease. In the largest studies on endovascular repair, 37% of patients died or required endovascular or surgical reintervention within a 3-year follow-up period.⁷⁾ Pujara, et al. reported good early and late outcomes after open repair for chronic distal aortic dissection.⁸⁾ They recommended that endovascular repair should be limited to only patients with limited dissection or those considered high risk for open surgery. Therefore, we chose an open surgery in a second-stage repair for the dissected diverticulum and adjacent descending aorta.

One staged repair such as the total arch replacement with open stent grafting may be preferable for our case because cumulative mortality and morbidity associated with the staged repair for TAAD is not insignificant. Tsagakis, et al. demonstrated good operative results with an in-hospital mortality of 12% and 97% immediate false lumen thrombosis in the peri-stent-graft level for TAAD with the use of a hybrid stent graft with integrated vascular prosthesis

for arch replacement.⁹⁾ However, introducing the stent graft into an acutely dissected aorta may be cumbersome and may carry the risk of new intimal tear formation, malperfusion syndrome, aortic disruption and paraplegia. For these reasons, we performed the conventional repair for TAAD regardless the exclusion of the origin of ARSA was not obtained at a first-stage operation.

In conclusion, we experienced a rare case of acute type A aortic dissection of the left aortic arch with an aberrant right subclavian artery. The conventional two-staged repair was feasible and provided a good clinical outcome.

Disclosure Statement

Keizo Tanaka and other co-authors have no conflict of interest.

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