» **Case Report** «

Two Cases of Thoracic Aortic Aneurysm with Right Aortic Arch: Comparison of Two Operative Strategies for Hybrid Thoracic Endovascular Repair

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Hybrid TEVAR was performed in 2 patients with right aortic arch accompanied by Kommerell's diverticulum and aortic aneurysm. In patient 1, total debranch + TEVAR was performed with 1-stage median sternotomy. In patient 2, total arch replacement and insertion of a peripheral elephant trunk were performed first, followed by TEVAR. No endoleaks or aortic events were observed in either case during the observation period, and both patients had good postoperative clinical courses. We report our experience with two such cases that were treated with two different methods of hybrid TEVAR, and discuss the merits and demerits of each treatment method.

Keywords: aortic aneurysm, thoracic, endovascular surgery

Background

Right-sided aortic arch and Kommerell's diverticulum with aneurysmal dilatation are relatively rare conditions, and there is no consensus on the appropriate surgical strategy for the conditions. Here, we report our experience with two cases of right aortic arch respectively accompanied by Kommerell's diverticulum and aortic aneurysm that were treated with different methods of hybrid thoracic endovascular aortic repair (TEVAR). We also examine the advantages and disadvantages of the treatment methods.

Case Report

Case 1

A 76-year-old man was pointed out right aortic arch and abnormality at the origin of the left subclavian

artery (Kommerell's diverticulum) by computed tomography (CT) when the patient underwent colon cancer surgery at another hospital in January 2009. One year after surgery, no recurrence of colon cancer was noted; however, Kommerell's diverticulum had increased to 56 mm, so the patient was referred to our department for further examination and treatment. He was admitted to our department for surgery in August 2010. On his chest CT, the aortic arch was observed to the right of the trachea and esophagus. A diverticulum with a maximum diameter of 56 mm that protruded to the left was observed on the descending aorta. The cervical branches bifurcated in the order of left common carotid artery, right common carotid artery, and right subclavian artery; the left subclavian artery bifurcated from the diverticulum (Fig. 1A).

Based on the aforementioned findings, a diagnosis of right aortic arch Edwards IIIB type accompanied by Kommerell's diverticulum was made. When informed of the possibility of an aneurysm or rupture, the patient expressed a strong desire for treatment, so surgery was decided. As this was a tumor-bearing patient, TEVAR with total debranching of cerebral vessels through median sternotomy was selected to maintain extracorporeal circulation at a minimum

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Fig. 1 Pre- and post-operative computed tomography angiography (CTA) scan of Case 1. **(A)** Preoperative CTA scan. Please note the right-sided aortic arch and the diverticulum with a maximum diameter of 56 mm that protruded to the left on the descending aorta. **(B)** Scheme of operative procedure. **(C)** Postoperative CTA scan. No endoleaks inside the diverticulum.

level, with coil embolization of the left subclavian artery bifurcating from the diverticulum to be performed at a later date. If possible, simple TEVAR without debranching was considered to be safe. However, it was difficult to obtain the enough landing zone in the descending aorta.

Operation: With the patient in the supine position, an incision was made below the left clavicle to expose the left subclavian artery that was anastomosed end-to-side to an 8-mm artificial blood vessel. The mediastinum was accessed via median sternotomy. The ascending aorta was partially blocked and connected to the central side of a 4-branched artificial vessel (16 mm × 8 mm × 8 mm) with Cardio pulmonary bypass. Under brain separation, the legs of the 4-branched artificial vessel were connected to the right common carotid artery, the left common carotid artery, and the right subclavian artery. After connecting the artificial vessel, that was anastomosed to the left subclavian artery to the remaining artificial vessel leg. Stent graft manipulations were then started. Two GORE® TAG® thoracic endoprostheses (37 mm × 20 cm and 37 mm × 10 cm) (WL Gore & Associates, Flagstaff, Arizona, USA) was inserted and the stent graft was retained from the tip of the artificial vessel anastomosed to the ascending aorta (**Fig. 1B**). The total time under extracorporeal circulation was 80 min, with the operation lasting 394 min.

Postoperative course: The patient went well after the surgery without significant complications on postoperative day (POD) 22, coil embolization of the left subclavian artery was performed. On POD 27, contrast

CT confirmed no endoleaks inside the diverticulum (**Fig. 1C**). On POD 31, the patient was discharged from the hospital. At three postoperative years, the diameter of the diverticulum was unchanged and no endoleaks were observed. The patient currently undergoes follow-up examinations at our hospital as an outpatient.

Case 2

A 75-year-old man visited his physician for annual check-up and was pointed out abnormal shadow on the chest X-ray. His CT scan revealed right aortic arch and a 55-mm distal aortic arch aneurysm. The internal membrane of the aneurysm was irregular and accompanied by mural thrombus, and there appeared to be arteriosclerotic changes. The branches of the arch area bifurcated in the order of left brachiocephalic artery, right common carotid artery, and right subclavian artery (**Fig. 2A**).

Based on the aforementioned findings, a diagnosis of right aortic arch with distal aortic arch aneurysm was made, and surgery was decided. The surgical method selected was total arch replacement with elephant trunk (ET), performed first through median sternotomy followed by the second stage TEVAR.

The first surgery: With the patient in the supine position, a horizontal incision was made below the right clavicle to expose the right subclavian artery, which was connected to an 8-mm artificial blood vessel via end-to-side anastomosis. The mediastinum was accessed via median sternotomy. Cardio pulmonary bypass was started. After inducing hypothermic circulatory arrest (minimum body temperature,

Fig. 2 Pre- and post-operative computed tomography angiography (CTA) scan of Case 2. (A) preoperative CTA scan. Please note the aneurysm in the aortic arch with right-sided aortic arch. (B) Scheme of operative procedure. (C) Postoperative CTA scan. No endoleaks was shown. The aneurysm had shrunk to 40 mm after 2 years.

27.0°C), antegrade cerebral perfusion was established. A 24-mm Hemashield graft (Maquet, Wayne, New Jersey, USA) was inserted 5 cm into the descending aorta as an elephant trunk (ET) and then reconstruct aortic arch with a 24-mm J-graft 4-branch graft (Japan Lifeline Co., Ltd., Tokyo, Japan). The right subclavian artery was connected to a lateral branch of the 4-branch graft with end-to-end anastomosis, replacing the entire aortic arch with artificial vessels. The total time under extracorporeal circulation was 152 min, the circulatory arrest duration was 21 min, and the surgery lasted for 390 min.

The second surgery: TEVAR was performed under general anesthesia on POD 14 of the 1st surgery. Two GORE® TAG® endografts (31 mm × 15 cm) were placed from inside the ET (Fig. 2B). On POD 19 of the 1st surgery (day 5 after endograft insertion), contrast CT confirmed no clear endoleaks (Fig. 2C). On POD 21 of the first surgery (day 7 after stent graft insertion), he was discharged from the hospital, independently ambulatory. At two postoperative years, outpatient examination results showed that the aortic aneurysm had shrunk to 40 mm with no sign of endoleaks.

Discussion

Right-sided aortic arch is a relatively rare congenital disorder occurring in approximately 0.1% of the population.¹⁻³ Many cases are accompanied by other congenital heart anomalies or visceral heterotaxy. Kommerell's diverticulum, first reported by

Burckhard Kommerell in 1936, is a remnant of the dorsal aorta.⁴ Since cases of the right aortic arch accompanied by Kommerell's diverticulum or aortic aneurysm are rare, consensus on the surgical indications and methods has not been reached.⁵

According to Austin, et al.,⁶ in 19% of patients with Kommerell's diverticulum, the diverticulum ruptures causing death. Moreover, Cina, et al.⁷ reported that of 32 cases of Kommerell's diverticulum, 53% presented with rupture or dissociation, and surgery was indicated in patients who could withstand surgery and had a diverticulum with a diameter >3 cm. Therefore, we indicated surgery in the present cases.

Approximately 14% of right aortic arch cases are Edwards IIIB, which is accompanied by an abnormality at the origin of the left subclavian artery.¹ Case 1 in this report was also Edwards IIIB. Surgery in such cases can be problematic owing to anomalies in the path of the brachiocephalic vein, difficulty in determining the appropriate method to expose the descending aorta, and the complexity of reconstruction. These cases may require methods such as right thoracotomy, bilateral thoracotomy, or median sternotomy + left thoracotomy, as well as thoracotomy on the side of the aneurysm or dissociation, if one exists. However, the manipulations involved in thoracotomy are associated with risks such as respiratory complications, phrenic nerve paralysis and other nerve disorders, hemorrhage, and postoperative pain compare with left aortic arch. Thus, high operative mortality rates of 12%–16.6% have been reported

for open surgeries.⁷⁾ A few recent studies report the use of median sternotomy in hybrid therapies with a combination of cervical branch reconstruction and stent graft treatments.⁸⁻¹⁰⁾ Patients who require only median sternotomy have a major advantage. Neither of the patients required additional thoracotomy, and both surgeries were completed using only a mediastinal approach via median sternotomy.

The advantage of total debranch + TEVAR is mainly in that it does not require hypothermia, selective cerebral perfusion, and cardiac arrest. So, this method could be less invasive than arch replacement + ET followed by the second stage TEVAR. It is true that the total debranching of the cerebral vessels can be performed without cardio pulmonary bypass. However, in our department, we routinely use normothermic selective cerebral perfusion to avoid cerebral ischemia during the procedure. The disadvantage of this method is that it does require proximal landing zone in the ascending aorta. In cases of right aortic arch, the aortic arch is often sharp, which can make it difficult to deploy an endograft up to the ascending aorta.¹¹⁾ It might be feasible by using hard wires such as a Lunderquist guide wire or applying the pull-through wire technique, but there is still a risk of possible endoleak due to the bird-beak configuration resulting from the sharp angulation of the aortic arch.

On the other hand, the arch replacement + ET followed by the second stage TEVAR could be more invasive than total debranch + TEVAR, but the deployment of the endograft would be much easier because the proximal landing would be the elephant trunk, namely the prosthetic graft elephant trunk sitting in the descending aorta. Also, this method is considered to be more effective in preventing the risk of endoleak, than the total debranch + TEVAR.

We think that the arch replacement + ET followed by the second stage TEVAR is more preferable to total debranching + TEVAR. However, in the case which seems to be difficult to endure the arch replacement, we may choose the treatment that is a low invasion such as total debranching + TEVAR.

Conclusion

We experienced two cases of right aortic arch accompanied by Kommerell's diverticulum and aortic

aneurysm that were treated with surgery using two different methods of TEVAR. While both patients experienced positive outcomes, arch replacement + ET + 2nd TEVAR was considered the better choice for patients who can withstand this procedure.

Disclosure Statement

None of the authors declare any conflict of interest.

References

- 1) Knight L, Edwards JE. Right aortic arch. Types and associated cardiac anomalies. *Circulation* 1974; **50**: 1047-51.
- 2) Hastreiter AR, D'Cruz IA, Cantez T, et al. Right-sided aorta. I. Occurrence of right aortic arch in various types of congenital heart disease. II. Right aortic arch, right descending aorta, and associated anomalies. *Br Heart J* 1966; **28**: 722-39.
- 3) Shuford WH, Sybers RG, Gordon IJ, et al. Circumflex retroesophageal right aortic arch simulating mediastinal tumor or dissecting aneurysm. *AJR Am J Roentgenol* 1986; **146**: 491-6.
- 4) van Son JA, Konstantinov IE, Burckhard F, Kommerell and Kommerell's diverticulum. *Tex Heart Inst J* 2002; **29**: 109-12.
- 5) Ota T, Okada K, Takanashi S, et al. Surgical treatment for Kommerell's diverticulum. *J Thorac Cardiovasc Surg* 2006; **131**: 574-8.
- 6) Austin EH, Wolfe WG. Aneurysm of aberrant subclavian artery with a review of the literature. *J Vasc Surg* 1985; **2**: 571-7.
- 7) Cinà CS, Althani H, Pasenau J, et al. Kommerell's diverticulum and right-sided aortic arch: a cohort study and review of the literature. *J Vasc Surg* 2004; **39**: 131-9.
- 8) Idrees J, Keshavamurthy S, Subramanian S, et al. Hybrid repair of Kommerell diverticulum. *J Thorac Cardiovasc Surg* 2014; **147**: 973-6.
- 9) Kawajiri H, Shimizu H, Yoshitake A, et al. Hybrid repair of a Kommerell diverticulum associated with a right aortic arch and a left descending aorta. *J Vasc Surg* 2012; **56**: 1727-30.
- 10) Frigatti P, Grego F, Deriu GP, et al. Hybrid endovascular treatment of aneurysm degeneration in a rare right-aortic arch anomaly with Kommerell diverticulum. *J Vasc Surg* 2009; **50**: 903-6.
- 11) Sato S, Matsuda H, Fukuda T, et al. Hybrid repair combined with open surgery and endografting for lesions in right aortic arch: report of three cases. *Ann Vasc Dis* 2012; **5**: 61-4.