

➤ **Case Report** ◀

Endovascular Treatment for Kommerell's Diverticulum with a Right-Sided Aortic Arch

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We report a rare case of type A dissection involving a right-sided aortic arch with an aberrant left subclavian artery originating from Kommerell's diverticulum in a 76-year-old woman. Endovascular treatment for Kommerell's diverticulum including intimal tear of the dissection was performed. At the 5-year follow-up, the patient was doing well, with no endoleak or dilatation of the Kommerell's diverticulum.

Keywords: right-sided aortic arch, Kommerell's diverticulum, aortic dissection

Introduction

The occurrence of a right-sided aortic arch with an aberrant left subclavian artery (ALSA) is an uncommon anomaly.¹⁾ An aneurysmal change at the origin of ALSA, known as Kommerell's diverticulum (KD), poses a high risk of rupture and dissection.²⁾ Dissection of a right-sided aortic arch is even rarer with only a handful of cases being documented so far.²⁾ Here we report a successful endovascular repair of KD dissection associated with right-sided aortic arch and ALSA.

Case

A 76-year-old woman was admitted to our hospital for the treatment of a right common iliac artery aneurysm. The patient had been previously diagnosed with a right-sided aortic arch with ALSA originating from KD. On physical examination, we found that her blood pressure was 112/60 mmHg in both arms, and her heart rate was

regular at 90 beats per minute. Furthermore, no carotid bruits or any other abnormal findings were observed. Concomitantly, the laboratory examinations were unremarkable. Chest X-ray revealed a tortuous aorta with the absence of the left first aortic arch. While transthoracic echocardiography demonstrated no congenital anomaly, computed tomography (CT) revealed the right-sided aortic arch and KD with the maximal diameter 33 mm (Figs. 1A and 1B). Three-dimensional CT scanning showed the proximal-to-distal order of the branches of the aortic arch to be as follows: the left common carotid artery, right common carotid artery, right subclavian artery, and ALSA originating from the KD (Fig. 1C). Moreover, a right common iliac artery aneurysm with a maximal diameter of 48 mm could be recognized.

On the day before the scheduled surgery for iliac aneurysm, the subject complained of sudden back pain and numbness of the left arm. CT analysis revealed a retrograde type A aortic dissection and occlusion of ALSA. The false lumen had thrombosed, and an intimal tear was found in the KD (Fig. 2A). The ascending aorta was 39 mm in maximal diameter, and the thrombosed false lumen was 10 mm thick (Fig. 2B). ALSA originating from the KD was obstructed due to compression of the false lumen. Axillo-axillary bypass was immediately performed with an 8-mm ringed expanded polytetrafluoroethylene (ePTFE) graft, which caused the blood flow to the left arm to resume. After antihypertensive therapy for 2 months, CT showed an occlusion of the axillo-axillary bypass graft, but the false lumen was significantly diminished in size, and ALSA was well enhanced by increased antegrade flow in the true lumen. CT showed no abnormalities of the cervical and intracranial arteries and the circle of Willis. Because there was a possibility that the KD could rupture or recanalize to the false lumen, endovascular treatment was chosen to exclude the KD, including the intimal tear of the dissection. The endovascular procedure with debranching of ALSA was performed under general anesthesia in the operating room 2 months after the onset of aortic dissection. Two GORE TAG Thoracic Endoprostheses (WL Gore and Associates, Flagstaff, AZ, USA) were inserted through the

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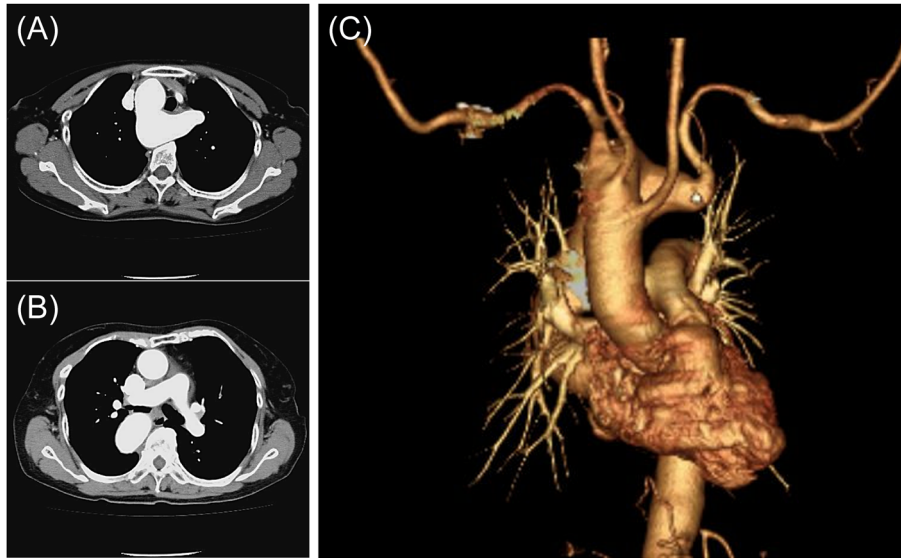


Fig. 1 Computed tomography (CT) analysis upon admission. (A, B) Kommerell's diverticulum and the aorta appear to be on the right side of both the esophagus and trachea. (C) Three-dimensional CT scanning shows a right-sided aortic arch with an aberrant left subclavian artery originating from the Kommerell's diverticulum.

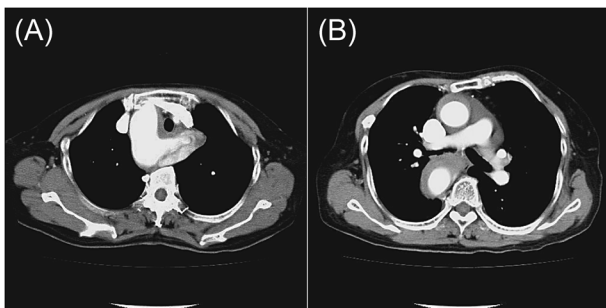


Fig. 2 (A, B) Computed tomography (CT) scan showing Stanford type A acute aortic dissection with an entry at the Kommerell's diverticulum.

left femoral artery and deployed in the descending aorta according to the diameter at the proximal and distal landing zone. A 34-mm-diameter, 200-mm-long GORE TAG[®] stent graft was first deployed at the distal descending thoracic aorta. Second, a 37-mm-diameter, 150-mm-long GORE TAG[®] stent graft was deployed at the level of the ALSA coverage. Percutaneous coil embolization of the proximal segment of ALSA was then performed to ensure exclusion of the KD through the left brachial artery. After deployment of the devices, angiography showed successful exclusion of the KD and a small type 2 endoleak. Finally, a blood pressure gradient of 13 mmHg was observed between the arms.

The patient's post-procedure course was uneventful. Neither left arm claudication nor subclavian steal syndrome occurred. Although CT scanning revealed a small endoleak, the stent graft was correctly positioned, and the KD had been successfully excluded (Fig. 3). The



Fig. 3 Postoperative computed tomography (CT) showing the exclusion of Kommerell's diverticulum.

patient was successfully discharged from the hospital on the seventh day after the treatment. Three months later, endovascular repair of the right common iliac artery was performed. At that time, the endoleak remained. However, 1 year later, follow-up CT showed that the endoleak had disappeared. The most recent scan was performed 5 years after the operation and showed neither any endoleak nor any dilatation of the KD.

Discussion

A right-sided aortic arch is an anatomical variant occurring in approximately 0.1% of the general population.¹⁾ From the branch configuration of the aortic arch, the mirror image group accounts for 60%, while the group with ALSA accounts for 40%.³⁾ In the former, the branch is bilaterally symmetric with a normal left aortic arch. In the latter, the proximal-to-distal order of the branches of the aortic arch is as follows: left common carotid artery, right common carotid artery, right subclavian artery, and ALSA originating from the KD.³⁾ In embryologic terms, KD is a persistent remnant of the fourth primitive dorsal arch due to a failed regression.⁴⁾

Cina reviewed 32 cases of KD associated with a right-sided aortic arch, of which 13 were associated with aortic dissection, and two presented with rupture.²⁾ Esophageal or tracheal compressive symptoms are definitive indications for surgical intervention in patients with KD. In terms of size, KD with rupture or dissection has been reported regardless of its size. Cina suggested surgery for KD of over 30 mm in diameter.²⁾ Considering the normal diameter of a subclavian artery, 30 mm at the proximal portion is equivalent to twice the normal diameter. In addition, because KD is an embryological remnant due to failed regression,⁴⁾ it is supposed that the tissue is vulnerable. In our case, the KD size was over 30 mm in diameter, with an intimal tear of the dissection in the KD. Therefore, the risk of rupture or recanalization to the false lumen was high. Open surgeries in this condition have been reported, such as total arch replacement through a median sternotomy and descending aorta replacement through a right thoracotomy. These procedures involve careful surgery with a high degree of familiarity with anatomy, and, therefore, present a high degree of difficulty. In the past decade, owing to the widespread use of thoracic endovascular aortic repair, successful endovascular treatment with stent grafts for KD has been reported.⁵⁻⁹⁾ The right aortic arch has a shorter radius and more acute curvature than the gentle curve of the left aortic arch. Moreover, the severe aortic angulation at the point of transition of the thoracic to the abdominal aorta is an anatomical challenge in endovascular repair.^{9,10)} In our case, since the device could not pass the severe angulation of the aorta and type 1 proximal endoleak appeared, a pull-through technique and an additional device were applied to the proximal site. Fortunately, their events did not occur. While the evaluation of long-term results and careful observation is important, in future, endovascular therapy would be expected to improve treatment results with the modification of these devices.

Conclusion

Endovascular treatment of KD dissection associated with a right-sided aortic arch and an ALSA is feasible. Exclusion of the KD including the intimal tear of the dissection by stent graft placement and coil embolization is effective and serves as a less invasive therapy, avoiding second rupture and recanalization to the false lumen.

Disclosure Statement

The authors have no conflicts of interest.

Author Contributions

Conception of this case presentation: MH, TF

Writing: MH

Critical review and revision: all authors

Final approval of the article: all authors

Accountability for all aspects of the work: all authors

References

- 1) Hastreiter AR, D'Cruz IA, Cantez T, et al. Right-sided aorta. *Br Heart J* 1966; **28**: 722-39.
- 2) Cina 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.
- 3) Stewart JR, Kincaid OW, Titus JL. Right aortic arch: plain film diagnosis and significance. *Am J Roentgenol* 1966; **97**: 377-89.
- 4) van Son JA, Konstantinov IE, Burckhard F, Kommerell and Kommerell's diverticulum. *Tex Heart Inst J* 2002; **29**: 109-12.
- 5) Midorikawa H, Kanno M, Ishikawa K, et al. Endovascular repair of a Kommerell's diverticulum and aneurysmal right-sided aortic arch: a case report. *Ann Vasc Dis* 2009; **2**: 54-7.
- 6) Motoki M, Hattori K, Kato Y, et al. Endovascular repair of ruptured aberrant left subclavian artery with right aortic arch. *Ann Thorac Surg* 2013; **95**: 699-701.
- 7) Hsu HL, Huang CY, Chen JS. Total endovascular repair for acute type B dissection in the setting of right aortic arch with aberrant left subclavian artery and Kommerell diverticulum. *J Thorac Cardiovasc Surg* 2015; **150**: 409-11.
- 8) Gao P, Wang M, Dong D, et al. Endovascular repair of a Kommerell diverticulum anomaly. *Ann Thorac Surg* 2015; **99**: 1801-3.
- 9) Klonaris C, Avgerinos ED, Katsargyris A, et al. Endovascular repair of a right-sided descending thoracic aortic aneurysm associated with a right aortic arch and a left subclavian artery arising from a Kommerell's diverticulum. *Cardiovasc Intervent Radiol* 2009; **32**: 758-61.
- 10) Bodine JA, D'Souza VJ, Formanek AG. An unusual type of dissecting thoracic aneurysm in association with right aortic arch. *Vasa* 1982; **11**: 223-8.