Clinical Case Report

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A case report of frozen elephant trunk combined with endovascular treatment for acute aortic dissection of Kommerell's diverticulum involving right aortic arch and descending aorta

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Abstract

Rationale: Acute aortic dissection of Kommerell's diverticulum in the right aortic arch is extremely rare. There are several different procedures for the disease. With advances in endovascular treatment, hybrid surgical and endovascular management may provide a treatment of choice for this kind of disease.

Patient concerns: A 43-year-old man was admitted to the hospital with intermittent pain of left arm, chest, and back.

Diagnoses: Computed tomography demonstrated an aberrant left subclavian artery originating from Kommerell's diverticulum in the right aortic arch, acute aortic dissection of Kommerell's diverticulum involving arch and descending aorta.

Interventions: Total arch replacement combined with frozen elephant trunk was performed to create an adequate landing zone through a median sternotomy by circulatory arrest. Thoracic endovascular aortic repair was performed to isolate Kommerell's diverticulum from descending aorta completely, extending from the frozen elephant trunk to the distal descending aorta at the same time.

Outcomes: The patient got an uneventful postoperative course.

Lessons: Hybrid surgical and endovascular management is a safe and effective procedure for this rare disease.

Abbreviations: CT = computed tomography, KD = Kommerell's diverticulum, LCCA = left common carotid artery, LSCA = left subclavian artery, RCCA = right common carotid artery, RSCA = right subclavian artery.

Keywords: dissection, endovascular repair, frozen elephant trunk, Kommerell's diverticulum, right aortic arch

1. Introduction

A right-sided aortic arch is an extremely rare congenital defect. According to autopsy studies, it affects 0.04% to 0.1% of the population. Type II right aortic arch has an aberrant left subclavian artery (LSCA) originating from Kommerell's diverticulum (KD), which is a remnant of the left arch and the branches originate from the aortic arch in the following order: left common carotid artery (LCCA), right common carotid artery (RCCA), right subclavian artery (RSCA), and aberrant LSCA.[1] Aneurysm or dissection often originates from KD resulting in Kommerell's aneurysm or dissection located at the origin of the aberrant LSCA.[2] Because of the rarity as well as the heterogeneity of right aortic arch aneurysms and dissections, surgical treatment remains to be challenging and so there has been no accepted gold standard of treatment. We herein describe a case of right aortic arch complicated by acute aortic dissection of KD involving aortic arch and descending aorta, and was successfully treated with total arch replacement, frozen elephant trunk installation and endografting procedure.

2. Case report

A 43-year-old male with a history of hypertension requiring medication was admitted to our hospital with intermittent pain of left arm, chest, and back. The left arm turned black and cold at the time of onset, and recovered later after half an hour. Computed tomography (CT) revealed acute aortic dissection of KD involving right arch and descending aorta with an aberrant LSCA originating from KD (Fig. 1A–C).

The surgical procedure was performed under deep hypothermic circulatory arrest through a median sternotomy. Cardiopulmonary bypass was established with right atrium and right femoral artery cannulation, and left heart venting was achieved through a right superior pulmonary vein cannulation. The ascending aorta was cross-clamped and cardioplegia was infused through the aortic root. After the nasopharyngeal temperature was dropped to 20°C, the right aortic arch was opened under circulatory arrest with selective cerebral perfusion through RCCA. The orifice of KD in the arch and descending aorta was too deep to manipulate surgically. A frozen elephant trunk (CRONUS 28 mm × 120 mm) was installed into the descending aorta to cover the orifice of KD and create a landing zone for thoracic endovascular aortic repair (TEVAR), (Fig. 2C). Four

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The authors have no conflicts of interest to disclose.

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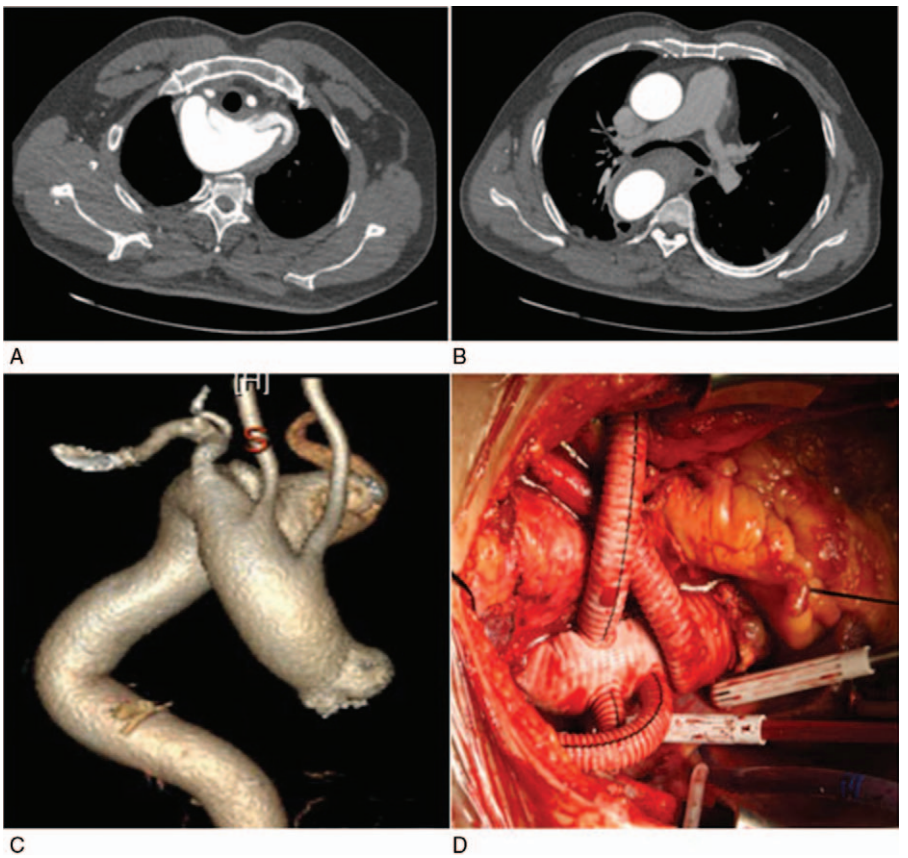

Figure 1. (A–C) Preoperative CT scan showing an aortic dissection of KD involving right arch and descending aorta with an aberrant LSCA originating from KD; (D) total arch replacement combined by reconstructing the 4 branches in the following order: LCCA, RCCA, RSCA, LSCA. CT = computed tomography, KD = Kommerell's diverticulum, LCCA = left common carotid artery, LSCA = left subclavian artery, RCCA = right common carotid artery, RSCA = right subclavian artery.

branch prosthetic vessel (Terumo Gelweave 26/10/8/8 × 10 mm) was used to replace the arch and reconstruct the four branches of the arch in following order: LCCA, RCCA, RSCA, LSCA (Fig. 1D). Distal anostomosis of LSCA was performed through an infraclavicular incision (Fig. 2D). Ligation of LSCA was performed before vertebral artery enters LSCA and reconstruction of the branch vessel to LSCA was performed through a neck incision (Fig. 2D). TEVAR (Medtronic VAMF3434C 150 TE) was performed through right femoral artery incision, extending from the frozen elephant trunk to the distal descending aorta (Fig. 2C). The cardiopulmonary bypass time was 232 minutes, cross-clamping time was 112 minutes, and circulatory arrest time was 28 minutes.

The time of mechanical ventilation support was 20.55 hours. The length of time in the intensive care unit was 62.8 hours. The length of postoperative hospital stay was 14 days. The postoperative course was uneventful. Postoperative CT before discharge showed thrombus formation in the KD, with no residual leakage of the elephant trunk and hybrid endograft (Fig. 2A–C).

3. Discussion

A right aortic arch is a rare congenital defect of the aorta, and was first described 250 years ago by Fioratti and Aglietti. Type II right arch has a retroesophageal aberrant LSCA originating from KD.[1] Symptoms of right aortic arch might occur due to either compression of mediastinal structures or due to aneurysmal disease or dissection. In this case, the patient was diagnosed by acute dissection of KD involving right aortic arch, descending aorta, and aberrant LSCA. He presented with intermittent pain in his left arm, chest, and back. The left arm turned cold and black due to ischemia of aberrant LSCA.

Surgery remains to be challenging for KD aneurysm or dissection due to its rarity, retroesophageal anatomy, and difficulty in exposure of KD. A number of operative approaches used were as follows: median sternotomy, right thoracotomy, left thoracotomy, median sternotomy with thoracotomy, bilateral thoracotomy, and bilateral thoracotomy with transverse sternotomy.[3,4] According to the previous studies, median sternotomy remained to be hard to expose the whole KD.[5–7] With advances in endovascular treatment, hybrid surgical and endovascular management provided a treatment of choice for this kind of disease.[8] In this case, we used a frozen elephant trunk to create an adequate landing zone through a median sternotomy, while TEVAR was performed by extending from the frozen elephant trunk to the distal descending aorta to isolate the KD and dissection. LSCA was ligated before the entry of vertebral artery to prevent backflow of blood to KD through a neck incision. Hybrid surgical and TEVAR management could avoid additional right thoracotomy, and lessen the trauma to the patient. CT scan showed thrombus formation in the KD, with no residual leakage of the elephant trunk and hybrid endograft before discharge.

The case reported here corroborates evidence that the hybrid surgical and endovascular management was successful for treating acute aortic dissection of Kommerell's diverticulum involving right aortic arch and descending aorta. Also the study

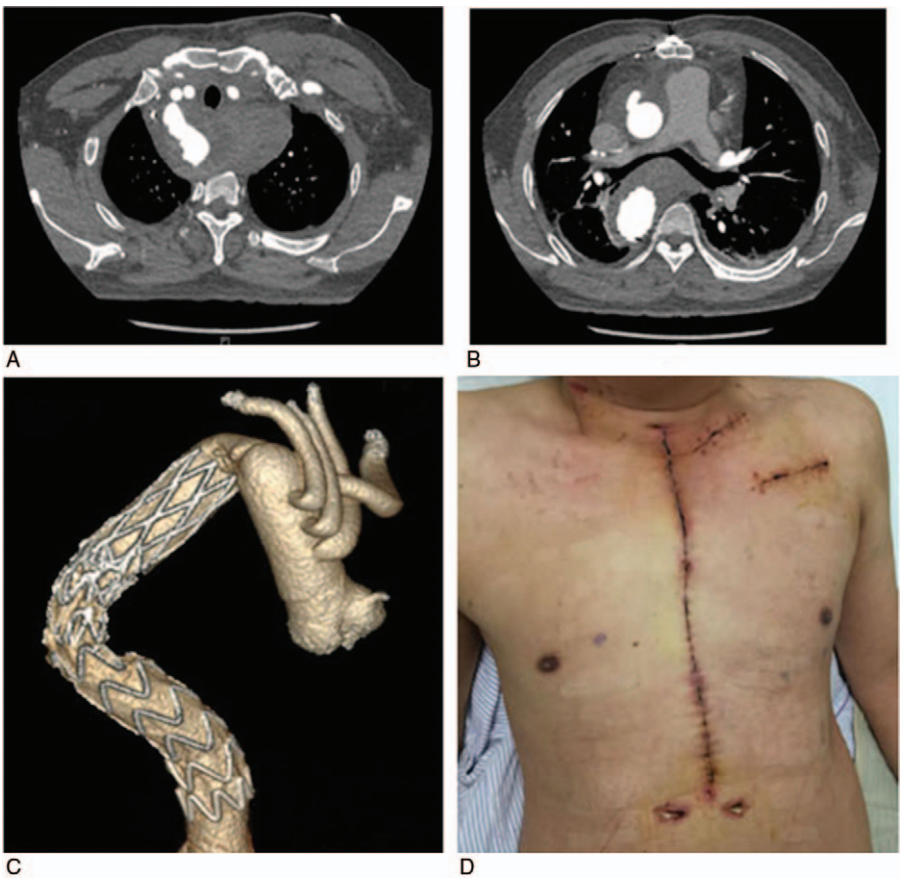

Figure 2. (A–C) Postoperative CT scan showing thrombus formation in the KD, with no residual leakage of the elephant trunk and hybrid endograft; (D) median sternotomy incision, neck incision, and infraclavicular incision. CT = computed tomography, KD = Kommerell's diverticulum.

demonstrated good short-term results, while follow-up is required for long-term results.

Author contributions

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Project administration: H. Guo.
Resources: H. Guo.
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Writing – original draft: H. Guo, X. Sun.
Writing – review & editing: C. Shu, C. Yu, H. Guo, X. Sun.

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