Surgical Outcomes of Kommerell Diverticulum

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Background: We aimed to assess the clinical outcomes of patients who underwent surgical repair of Kommerell diverticulum (KD) with individualized surgical methods.

Methods: A retrospective analysis was performed of adult patients (aged ≥17 years) who underwent surgery to treat KD between June 2008 and October 2019.

Results: Nine patients (median age, 45 years; range, 19–67 years; 7 men) underwent surgical repair. The indications for surgical therapy were acute aortic dissection in 2 patients, the presence of compressive symptoms due to dilated KD in 4 patients, and aneurysm growth in 3 patients. Various surgical techniques were used: (1) resection of the diverticulum stump and revascularization of the aberrant subclavian artery (n=3), (2) one-stage total-arch replacement including the diverticulum segment (n=3), and (3) hybrid repair (n=3). Early mortality occurred in 1 case of hybrid repair. Transient paraparesis occurred in a patient who underwent total arch repair as part of complicated acute aortic dissection. During follow-up (median duration, 30 months; range, 7–130 months), no late death or associated aortic complications were documented. All survivors were free from symptoms and had no abnormal findings on follow-up computed tomography.

Conclusion: With a customized surgical approach and appropriate consideration of patient-specific anatomy and associated comorbidities, KD can be repaired with favorable outcomes.

Keywords: Aorta, Anomaly, Kommerell, Surgical operation, Outcomes

Introduction

Kommerell diverticulum (KD) is a rare congenital anomaly of the aortic arch resulting from the failed involution of the fourth primitive dorsal arch. It is a segmental aortic aneurysmal dilatation associated with aberrant subclavian arteries (ASAs) and can occur on either the left or the right side. The natural history of this disease is still not well understood due to its rarity, and issues surrounding the surgical treatment of KD remain controversial. Compressive symptoms, such as dysphagia lusoria, stridor, or compressive chest pain, may require therapeutic interventions when the surrounding structures (the esophagus or trachea) are mechanically compressed by the enlarged diverticulum. In contrast, most patients with KD are asymptomatic and are diagnosed incidentally. In such patients, a specific threshold for therapeutic intervention based on aneurysm size has not yet been established because of the unclear natural history of this disease. The levels of risk of catastrophic aortic events associated with KD, including rupture or dissection, vary widely across reports, from 4% to 19% and from 11% to 53%, respectively [1,2]. Although some authors have advocated generous prophylactic interventions for this condition, the technically demanding surgical procedures attributable to the complex anatomy involved have complicated the issue [3].

While various techniques have been proposed for the treatment of KD, including open repair and endovascular or hybrid approaches, the majority of reports are limited to case series with small sample sizes, and a knowledge gap still exists regarding the optimal management of this rare disease. Although we were also limited by a small number of cases, we sought to share our experiences with the surgical treatment of KD over the past 12 years.
Methods

Study subjects

We retrospectively reviewed the records of adult patients at least 17 years old who underwent open surgical repair of KD or a hybrid operation between June 2008 and October 2019 at Asan Medical Center, Seoul, Korea. A total of 9 patients were identified in the institutional cardiac surgical database. The data collected included details regarding each patient’s preoperative symptoms, comorbidities, operative profiles, and follow-up. This study was approved by the institutional ethics committee and review board of Asan Medical Center (approval no., 2020-0026). The requirement for informed consent was waived due to the retrospective nature of the study.

Imaging data

Preoperative computed tomography (CT) scans, including aortic CT angiography, were performed for all patients. By reanalyzing the CT scans, the orientation of the aorta, the origin of the ASA, the associated aortic anomaly, and the size of the KD were identified. Two measurements of KD size were made: the maximal distance from the tip of the aneurysm to the opposite aortic wall and the diameter of the diverticular orifice (Fig. 1). Postoperative CT scans were also performed for all patients during follow-up to assess the aortic anatomy after surgery.

Surgical profile

The indications for surgical repair included compressive symptoms associated with KD, aortic rupture, acute or chronic aortic dissection, and distal arterial obstruction by the diverticulum. Patients without symptoms or catastrophic aortic events were not treated surgically.

Four surgeons performed the surgical management of KD during the study period. The surgical approaches and techniques were determined at the operating surgeon’s discretion in consideration of the patient’s anatomy and associated comorbidities. Concomitant procedures were performed when needed.

Results

Baseline patient characteristics and preoperative findings

The median age of the 9 patients at operation was 45 years (range, 19–67 years), and 7 (77.8%) were men. Preoperative CT scans revealed a right aortic arch (RAA) with a left ASA in 7 patients (77.8%) and a mean maximal aneurysmal diameter of 6.1±2.3 cm.

The indications for surgical treatment were complicated acute aortic dissection in 2 patients (patients 4 and 8), the presence of compressive symptoms exerted by the dilated KD in 4 patients (patients 1, 2, 7, and 3), and aneurysm growth in the remaining patients (patients 5, 6, and 9). Three patients presented typical compressive symptoms requiring surgical treatment. In patient 3, KD repair was performed as a concomitant procedure during the surgical excision of a left atrial myxoma, since the patient exhibited dysphagia attributed to KD.

Perioperative profiles depending on surgical approach

The surgical approaches and techniques were determined at the operating surgeon’s discretion in consideration of the patient’s anatomy and associated comorbidities. Diverticular stump resection and revascularization of the ASA were chosen for young patients with relatively small diverticula. For patients who required concomitant ascending aorta replacement, such as those with aortic dissection, sternotomy was preferred, and one-stage total-arch replacement was performed. However, a hybrid opera-
tion combined with endovascular vascular repair was an alternative for those deemed to be otherwise at high risk associated with extensive surgery.

Resection of diverticulum stump with aberrant subclavian artery revascularization

Resection of the diverticulum stump and revascularization of the ASA were performed in 3 patients (33.3%). Graft interposition of the proximal ascending aorta and supra-aortic vessels was not performed in these patients. The mean age of these 3 patients was 25.0±7.1 years, and the mean maximal diameter of the ascending aorta was 2.6±0.2 cm. The mean diameter of the diverticulum was 3.9±0.3 cm in these patients. All 3 patients had RAAs associated with left ASAs. The distal left ASAs were transferred to the left common carotid arteries, after which simple stump closure of the diverticulum was performed (Fig. 2). The approach for the stump closure was made through a median full sternotomy in 2 patients (patients 1 and 3), while patient 2 underwent this procedure through a left posterolateral thoracotomy (Table 1). Cardiopulmonary bypass (CPB) was used in 2 patients (patients 1 and 3).

One-stage total-arch replacement including the diverticulum segment

Proximal segments of the aorta were replaced in 3 patients. Graft interposition was performed on the ascending aorta, aortic arch, and proximal descending thoracic aorta including the diverticulum segment under CPB via median full sternotomy. Arch vessels were reimplanted to graft branches of a 4-branch graft. The mean age of these patients was 50.0±16.1 years, and the mean maximal diverticular size was 7.1±0.6 cm. The indications for surgery were acute aortic dissection (in patient 4), chronic aortic dissection (in patient 5), and symptomatic aortic aneurysm involving the aortic arch (in patient 6). The mean pump time was 310.0±113.8 minutes, and all 3 patients underwent surgery under moderate hypothermia (mean lowest body temperature, 23.4°C±3.3°C). Patient 4 presented with acute type B aortic dissection complicated by left upper-limb malperfusion. Detailed information regarding the anatomic profile and surgical procedure was described previously [4]. The intimal tear was located at the mid-arch involving

![Fig. 2. Resection of the diverticulum stump and revascularization of the aberrant subclavian artery. (A) All 3 patients had a right aortic arch with an aberrant left subclavian artery. The aortic arch vessels branched in the following order: left carotid artery, right carotid artery, right subclavian artery, and aberrant left subclavian artery. (B) The aberrant left subclavian artery was divided and anastomosed to the left common carotid artery with the end-to-side technique. The proximal stump of the left subclavian artery was repaired with an oversewing stitch. LCCA, left common carotid artery; RCCA, right common carotid artery; RSCA, right subclavian artery; ALSA, aberrant left subclavian artery.](image)

<table>
<thead>
<tr>
<th>Table 1. Patient characteristics</th>
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<tr>
<td>Patient</td>
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<td>9</td>
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ASA, aberrant subclavian artery; KD, Kommerell diverticulum; M, male; F, female; RAA, right aortic arch; LAA, left aortic arch; ALSA, aberrant left subclavian artery; ARSA, aberrant right subclavian artery; MCA, middle cerebral artery; AD, aortic dissection; DTA, descending thoracic aorta.
dissection flaps spanning from the opening of the right common carotid artery to the renal arteries and including the KD and the left ASA. Emergency total-arch replacement was performed through median full sternotomy in this patient.

**Hybrid repair of Kommerell diverticulum**

Hybrid repair of KD was performed in the other 3 patients. Three distinct hybrid approaches were used in the respective patients: (1) graft interposition of the total aortic arch just before the diverticular segment of the descending thoracic aorta, followed by retrograde thoracic endovascular aorta repair (TEVAR) (in patient 9), (2) type 2 zone 0 debranching and elephant trunk insertion followed by retrograde TEVAR (in patient 8) (Fig. 3), and (3) zone 2 debranching through a neck incision and subsequent retrograde TEVAR (in patient 7) (Fig. 4). The mean age of the 3 patients was 51.3±4.6 years, and the size of the diverticulum ranged from 4.0 to 9.3 cm. Hybrid procedures were performed due to compressive symptoms attributable to a huge diverticulum (in patients 7 and 9) and acute type A aortic dissection in patient 8. Two patients underwent sur-

**Fig. 3.** Hybrid repair of Kommerell diverticulum. Endovascular repair is a potential alternative in patients with aortic diverticulum complicated by aortic dissection. (A, B) A computed tomography scan of patient 8 shows a type A aortic dissection with an acute angle of aortic arch curvature. (C) Illustrates aortic disease. (D) Ascending aorta replacement with the elephant trunk technique, along with zone 0 debranching surgery, was performed to create a proximal landing zone. (E) The procedure was followed by thoracic endovascular aortic repair for the exclusion of aortic dissection in the descending thoracic aorta. LCCA, left common carotid artery; RCCA, right common carotid artery; RSCA, right subclavian artery; ALSA, aberrant left subclavian artery; LVA, left vertebral artery.

**Fig. 4.** Minimally invasive hybrid surgery to treat Kommerell diverticulum with bypass of the common carotid artery to the subclavian artery and endovascular repair. (A) Bypass of the left common carotid artery to the aberrant subclavian artery was performed with a ring-reinforced polytetrafluoroethylene graft through a cervical approach. The aneurysm of the Kommerell diverticulum was covered with a stent graft. This technique made it possible to repair the Kommerell diverticulum with a minimal incision and without cardiopulmonary bypass. (B) A computed tomography scan of patient 7 shows a ringed polytetrafluoroethylene graft (arrow) connecting the left common carotid artery and left subclavian artery and stent graft (star) in the aorta. LCCA, left common carotid artery; RCCA, right common carotid artery; RSCA, right subclavian artery; ALSA, aberrant left subclavian artery; LVA, left vertebral artery.
gery under CPB (patients 8 and 9), while the procedure for patient 7 did not require CPB.

Clinical outcomes

One in-hospital death (14.2%) occurred. This death occurred after a hybrid procedure in patient 9, who underwent surgery due to an extensive aneurysm compressing the aorta. A type 3 endoleak had led to persistent growth of the diverticulum and the aneurysmal sac after the endovascular repair. The patient could not be weaned from the ventilator and eventually died of sepsis attributed to a tracheoesophageal fistula, a complication that arose within 2 weeks of the hybrid procedure.

One patient (patient 4) experienced temporary paraparesis but recovered without sequelae. This patient also experienced mediastinitis. Several rounds of debridement and irrigation followed by omental and pectoralis muscle flap interposition were performed. On follow-up, he showed a progressively enlarging thoracoabdominal aorta (maximal diameter, 5.1 cm) due to remnant distal aortic dissection and consequently underwent thoracoabdominal aorta replacement a year after the index procedure.

No late mortality occurred, and all patients were free from relevant symptoms of KD during the follow-up period (median duration, 30 months; range, 7–130 months). Postoperative serial CT scans performed for the other 7 patients (with the exception of patients 4 and 9) showed no abnormal findings related to the repaired aorta (median time of CT scan, 6 months; range, 0–115 months) (Table 2).

Discussion

KD is a congenital aortic anomaly that arises due to failed involution at the fourth primitive dorsal arch [1]. The prevalence of right ASA from the left aortic arch and left ASA from the right aortic arch has been reported to be 0.7%–2.0% and 0.04%–0.4%, respectively. Approximately 20%–60% of cases of ASAs are associated with KD. Only a small portion of patients with KD present symptoms. Most of these aortic conditions are identified incidentally on CT scans performed for reasons unrelated to KD. Due to its rare clinical presentation, the natural history of this disease is still not well known. The reported incidence of catastrophic aortic events associated with KD, such as aortic rupture and aortic dissection, has varied widely across re-

Table 2. Operative details and complications

<table>
<thead>
<tr>
<th>Patient</th>
<th>Procedure</th>
<th>Urgency</th>
<th>Approach(es)</th>
<th>CPB time (min)</th>
<th>ACC time (min)</th>
<th>Lowest BT (°C)</th>
<th>Outcome</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Resection of diverticulum stump and revascularization of ASA</td>
<td>Elective</td>
<td>Sternotomy</td>
<td>93</td>
<td>NA</td>
<td>33.6</td>
<td>F/U without symptoms over 100 months</td>
</tr>
<tr>
<td>2</td>
<td>Resection of diverticulum stump and revascularization of ASA</td>
<td>Elective</td>
<td>Thoracotomy</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>F/U without symptoms over 105 months</td>
</tr>
<tr>
<td>3</td>
<td>Resection of diverticulum stump and revascularization of ASA; cardiac myxoma excision</td>
<td>Elective</td>
<td>Sternotomy</td>
<td>128</td>
<td>30</td>
<td>22.4</td>
<td>F/U without symptoms over 106 months</td>
</tr>
<tr>
<td>4</td>
<td>Asc. aorta, total-arch and proximal DTA replacement</td>
<td>Emergency</td>
<td>Sternotomy</td>
<td>431</td>
<td>15</td>
<td>20.1</td>
<td>Mid-DTA replacement due to progressive remnant distal aortic dissection</td>
</tr>
<tr>
<td>5</td>
<td>Asc. aorta, total-arch and proximal DTA replacement</td>
<td>Elective</td>
<td>Sternotomy</td>
<td>205</td>
<td>132</td>
<td>26.8</td>
<td>F/U without symptoms over 7 months</td>
</tr>
<tr>
<td>6</td>
<td>Asc. aorta, total-arch and proximal DTA replacement</td>
<td>Elective</td>
<td>Sternotomy</td>
<td>294</td>
<td>197</td>
<td>23.4</td>
<td>F/U without symptoms over 8 months</td>
</tr>
<tr>
<td>7</td>
<td>Hybrid operation (LSCA to LCCA bypass, retrograde TEVAR)</td>
<td>Elective</td>
<td>Sternotomy</td>
<td>217</td>
<td>33</td>
<td>17.5</td>
<td>F/U without symptoms over 130 months</td>
</tr>
<tr>
<td>8</td>
<td>Hybrid operation (asc. aorta replacement, debranching surgery, retrograde TEVAR)</td>
<td>Urgent</td>
<td>Sternotomy, femoral</td>
<td>357</td>
<td>234</td>
<td>23.3</td>
<td>In-hospital death</td>
</tr>
</tbody>
</table>

CPB, cardiopulmonary bypass; ACC, aortic cross-clamping; BT, body temperature; ASA, aberrant subclavian artery; NA, not available; F/U, follow-up; asc. aorta, ascending aorta; DTA, descending thoracic aorta; LSCA, left subclavian artery; LCCA, left common carotid artery; TEVAR, thoracic endovascular aorta repair.
ports by different researchers. An early study by Austin and Wolfe [5] reported a 19% rate of aortic dissection in 32 KD patients, and Cina et al. [6] reported a 53% rate of aortic dissection or rupture among 33 patients with KD. Tanka et al. [1] reviewed 212 cases of KD and reported an 11% frequency of aortic dissection and a 4% frequency of diverticular rupture.

Compressive symptoms of the esophagus or trachea, including dysphagia, dyspnea, chest pain, and cough, are definite indications for the surgical treatment of KD. For asymptomatic patients, however, a generally accepted, specific threshold for prophylactic surgery based on diverticular size or baseline aortic dimensions has not yet been established. Several groups have suggested criteria based on diverticular size, but these size criteria and the associated measurement methods have been inconsistent. Cina et al. [6] advocated the aggressive surgical treatment for KD larger than 3 cm in diameter at the level of the orifice, and Ota et al. [7] suggested surgical management for symptomatic aneurysms larger than 5 cm. Considering the incremental complexities of surgery as the diverticulum grows, Vinnakota et al. [2] and Kouchoukos and Masetti [3] also advocated early surgery to eliminate the risk of aortic dissection or aneurysm rupture. In contrast, asymptomatic ASA and KD are generally thought to be benign anomalies of the aorta. According to a study by Erben et al. [8], the natural course of aneurysmal growth seems to be relatively minimal, at approximately 1.4–2.4 mm per year.

Given this knowledge gap regarding the natural history of KD and the divergent points of view on the surgical treatment of KD, close outpatient follow-up has been conducted for asymptomatic patients at our center. Elective surgery is planned for patients with: (1) newly developed symptoms, (2) evidence of aneurysm growth, or (3) newly discovered aortic rupture/dissection in serial CT angiography. Various surgical techniques have been introduced for the repair of KD. We attempted to employ these techniques in a customized fashion, with consideration of the underlying anatomy and associated comorbidities of each patient.

Resection of the diverticular stump and subclavian-to-carotid bypass are standard procedures for patients with relatively small aneurysms. Various approaches, including left or right thoracotomy, median sternotomy, and a combination of thoracotomy and sternotomy, have been reported with favorable clinical outcomes [9-11]. Median full sternotomy provides exposure for the treatment of concomitant heart disease, as in the case of patient 3. When the patient’s anatomy allows, ASA revascularization and repair of the diverticulum through a left thoracotomy can be performed without using CPB (as in the case of patient 2).

When aortic dissection involves the aortic arch, graft interposition of the aorta encompassing the diverticular segment may be more suitable. Recent studies have shown satisfactory surgical outcomes in patients who underwent one-stage total-arch replacement including the diverticular segment with a branched vascular graft [12]. We conducted one-stage graft interposition using a 4-branched graft for 3 patients in whom aortic dissection involved the aortic arch.

In patients with huge diverticula or a greatly dilated aorta, surgical procedures can be more demanding, especially if the patient has multiple comorbidities. Endovascular repair can be an alternative to surgical repair, minimizing postoperative complications and maximizing postoperative recovery. The pressure of the aneurysmal sac decreases after successful sealing of the aneurysm, relieving the compressive symptoms of KD. For patients with steep aortic arch angles and limited proximal landing zones, partial aorta graft interposition with a debranching procedure provides easier access and an adequate landing zone for a stent graft.

However, KD repair using TEVAR could be unsuitable for patients with compressive symptoms. The literature includes a case in which compressive symptoms did not improve after endovascular repair. Reoperations due to endoleak have also been reported [2]. In our study, patient 9 had a type 3 endoleak from the ASA after exclusion via an endovascular graft. Even after subclavian artery occlusion using a device, the remnant aneurysmal sac compressed the surrounding organs. This eventually induced pneumonia and pulmonary abscess, which caused the death of the patient. Previous studies have shown medial attenuation of resected diverticular segments on histologic analysis [10,13]. This could explain the vulnerability of the aorta to endoleak after endovascular repair.

A right aortic arch with an ASA frequently creates a vascular ring when it is accompanied by ligamentum arteriosus. However, not all KD patients have a vascular ring that encircles the trachea and esophagus. Cases of recurrent compressive symptoms have been reported in patients with KD who previously underwent ligamentum division surgery [13,14]. This suggests that KD alone usually compresses the surrounding organs independently of ligamentum arteriosus, and decompression of the enlarged aneurysm can release the compressive symptoms of KD in the vast majority of cases. In cases with segmental aortic resection via sternotomy in our series, the total arch and proximal
descending aorta were replaced, a procedure that inevitably involves the removal/release of the ligamentum arteriosus. For other cases involving KD resection or a hybrid approach, the ligamentum arteriosus was left unidentified to avoid recurrent laryngeal nerve injury; however, we did not find any residual structure compressing adjacent organs in either symptom-based or imaging-based (CT) evaluations in all 8 successful cases.

One limitation of this study is that it was a retrospective review of a limited number of cases. As KD is a congenital aortic anomaly, each patient has distinctive anatomy; accordingly, 4 different surgeons applied modified and individualized surgical techniques for each patient. A cohesive strategy to treat this aortic disease should be established on the basis of further research.

In conclusion, among 9 patients, 1 case of early mortality (11.1%) occurred after surgery. Because KD is associated with vulnerability to severe aortic disease, a certain degree of mortality and morbidity after surgical repair is expected. However, with modified and individualized surgical techniques designed in consideration of each patient’s anatomy and comorbidities, KD can be treated with minimal morbidity and mortality and a reasonable rate of symptom resolution.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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