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Aberrant Subclavian Artery: A Neglected Cardiovascular Pathology

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Abstract

Objectives: The aberrant subclavian artery is among the congenital vascular anomalies of the aortic arch. Aberrant subclavian arteries can be seen in different types and on the right or left side. Aneurysmal dilatation, named Kommerell's diverticulum, may be seen in the descending thoracic aorta at the origin of the right aberrant subclavian artery. The aim of this study aimed to present our experience with aberrant subclavian artery pathology.

Materials and Methods: This was a retrospective review of our experience from two instutitions with 14 patients who had an aberrant subclavian artery between 2015-2021. Seven patients (62.5%) were male, and the median age at the time of surgery was 38.4 months (range: 6 months to 75 years). Patients were either asymptomatic, incidentally diagnosed, or presented with dyspnea and/or dysphagia lusoria.

Results: Asymptomatic patients were not interfered. There was no early or late mortality in 6 patients who underwent surgery. Three (50%) patients underwent aberrant right subclavian artery repair through right thoracotomy, two received a hybrid approach due to Kommerell's diverticulum, and one patient underwent aberrant left subclavian artery repair from left thoracotomy. The median postoperative duration of the intensive care unit and hospital stays were 1 and 6 days,

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respectively. Patients were followed for a median of 3.6 years (range, 1.2-6.8 years) after intervention. No endoleak was observed in the follow-up of patients who underwent endovascular repair.

Conclusion: The data suggest that symptomatic aberrant subclavian artery pathology can be treated safely in selected patients with good early and mid-term results. The right or left side of the aberrant subclavian artery may change the surgical approach. Patients with Kommerell's diverticulum with a right aberrant subclavian artery can be treated with a hybrid approach using endovascular techniques.

Keywords: Aberrant subclavian artery, Kommerell's diverticulum, surgical approach, hybrid techniques

Introduction

The aberrant subclavian artery (ASCA) is a congenital aortic anomaly that was first described by Hunauld[1] in 1735[2]. The incidence of the left ASCA is approximately 0.05%, and the right ASCA is 0.5-1%[2]. Kommerell's diverticulum, a descending aortic aneurysm at the origin of an ASCA, occurs in approximately 60% of right ASCA patients[3].

ASCAs uncommonly cause symptoms such as chest pain, dysphagia, and dyspnea[4]. However, dysphagia due to ASCA is also referred to as "dysphagia lusoria", hassle in swallowing solid food is a pathognomonic sign fort he disease[5].

The right ASCA and left ASCA develop embryologically in different ways. The right ASCA occurs when the right fourth arch involute cranially to the seventh intersegmental artery. Left ASCA occurs by the anomalous regression of the left fourth arch and left dorsal aorta[6]. The right ASCA is more constant than the left ASCA, and it is usually related to other congenital cardiac anomalies[7].

It has been presented that ASCA and Kommerell's diverticulum may be predisposed to engage in aneurysmal degeneration, so compose important risk factors for distal embolization, compression of adjacent structures, aortic dissection, and rupture[8]. Several surgical and endovascular techniques have been suggested for the treatment of these aneurysms[9,10]. The aim of this study was to present our experiences with ASCA with different surgical techniques.

Materials and Methods

Medical records of 14 consecutive patients who were diagnosed with ASCA between January 2015 and December 2021 were retrospectively. Patients with different aortic arches or vascular ring pathologies were excluded from the study. The data were collected retrospectively from patients' previous hospital records. Our study was approved by the Institutional Ethics Committee of İstanbul Medipol University (approval no: 24/11/2022, date: 1011) and was conducted in accordance with the principles of the Declaration of Helsinki.

A total of 14 patients met these criteria, including 7 males and 7 females. The median age of the patients was 38.4 years (range, 6 months to 75 years), and the median weight of the patients was 48.6 kg (range: 5 to 82 kg).

Preoperative thoracic computed tomography (CT) angiography (Figure 1) was performed in all patients to define aortic arch anatomy and to detect Kommerell's diverticulum or aneurysm if present. Thirteen patients had a right ASCA and one patient had left ASCA. Two of the 14 patients had Kommerell's diverticulum concomitant to the right ASCA.

Six of the were symptomatic and had dysphagia lusoria or respiratory problems due to tracheal compression. The remaining 8 patients were asymptomatic and diagnosed incidentally.

Surgical Technique

The operations were performed through right thoracotomy in the 3 patients with right ASCA. Systemic heparinization (100 U/kg) was performed after the

preparation of ASCA and the descending aorta. The right ASCA was carefully resected from the descending aorta, and the stump on the descending aorta was repaired primarily. The ASCA was anastomosed directly or with the use of a graft to the ascending aorta or the right common carotid artery. We used near-infrared spectroscopy for neurological monitorisation in all patients.

The left thoracotomy approach was used in a patient who had left ASCA with a right aortic arch and ligamentum arteriosum. This complete vascular ring pathology was repaired by the division of the ligamentum arteriosum.

The hybrid approach was preferred in 2 patients who had Kommerell's diverticulum. In these patients bilateral infraclavicular incision were performed and bilateral axillary arteries were dissected. An 8 mm ringed PTFE greft was placed subfascially and anastomosed end to side to the bilateral axillary arteries. Then the patients were then transferred to the angiography unit and the right femoral artery was dissected surgically for the endovascular procedure. The proximal of the right ASCA occluded endovascularly to prevent endoleak. The stent greft was placed distal to the left subclavian artery orifice until the midportion the descending thoracic aorta to exluded the aneurysm. The femoral artery was reconstructed primarily after the delivery system removal.

Except for the patient who was operated for the left ASCA, the other 5 patients received 100 mg aspirin and 75 mg clopidogrel for 3 months. After 3 months, these patients received only one antiplatelet therapy for lifelong. Control CT angiography was performed on all patients who underwent surgery at the 3rd postoperative month. In patients who underwent the thoracic endovascular aortic repair (TEVAR) procedure, additional control CTA was performed in the postoperative 1st year.

Statistical Analysis

IBM SPSS Statistics Software 21 (SPSS Inc., Chicago, IL, USA) was used for statistical analysis. Continuous variables were reported as median ± range. Categorical variables were reported as n (%).

Results

Symptomatic patients were treated surgically, and asymptomatic patiens without Kommerell' diverticulum were followed up in this study. Three symptomatic patients who had right ASCA underwent ASCA surgical treatment through right thoracotomy and ASCAs were implanted into the ascending aorta (Figure 2) directly in one patient, with the use of a graft in another patient and directly to the right carotid artery in one patient. The two patients with Kommerell's diverticulum were treated with a hybrid approach and TEVAR procedures (Figure 3). The last symptomatic infant who had a complete vascular ring with left ASCA, right aortic arch, and left ligamentum arteriosum underwent vascular ring

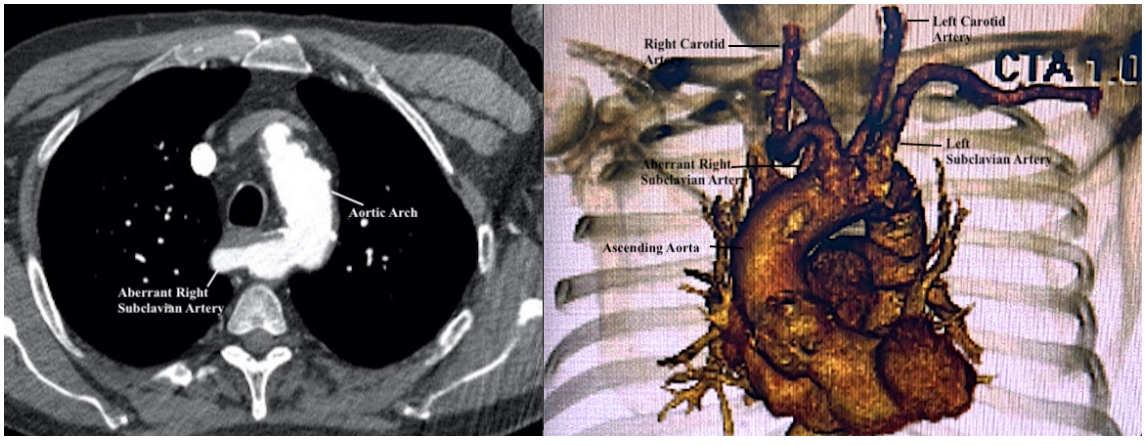

Figure 1. CT angiography view of the right ASCA
CT: Computed tomography, ASCA: Aberrant subclavian artery

division by the division of the ligamentum arteriosum through left thoracotomy.

Patients who underwent an operation had an uneventful postoperative course. No vascular or respiratory complications were observed. The median duration of intensive care unit stay after the operation was 1 day (range: 1-2 days) and the median duration of hospital stay after the operation was 6 days (range: 4-8 days). Postoperative early mortality, upper extremity ischemia, and wound problems were not observed, and all patients were discharged from the hospital uneventfully.

The median of follow-up for all patients was 3.6 years (range: 1.2-6.8 years). No endoleak, native, or bypass graft occlusion were observed in patients who received treatment. Significant regression in symptoms was observed in all patients immediately as wel as during the follow-up.

Figure 2. Conventional surgical treatment of right ASCA. Division from the descending aorta and re-implantation to the ascending aorta with an estension graft

ASCA: Aberrant subclavian artery

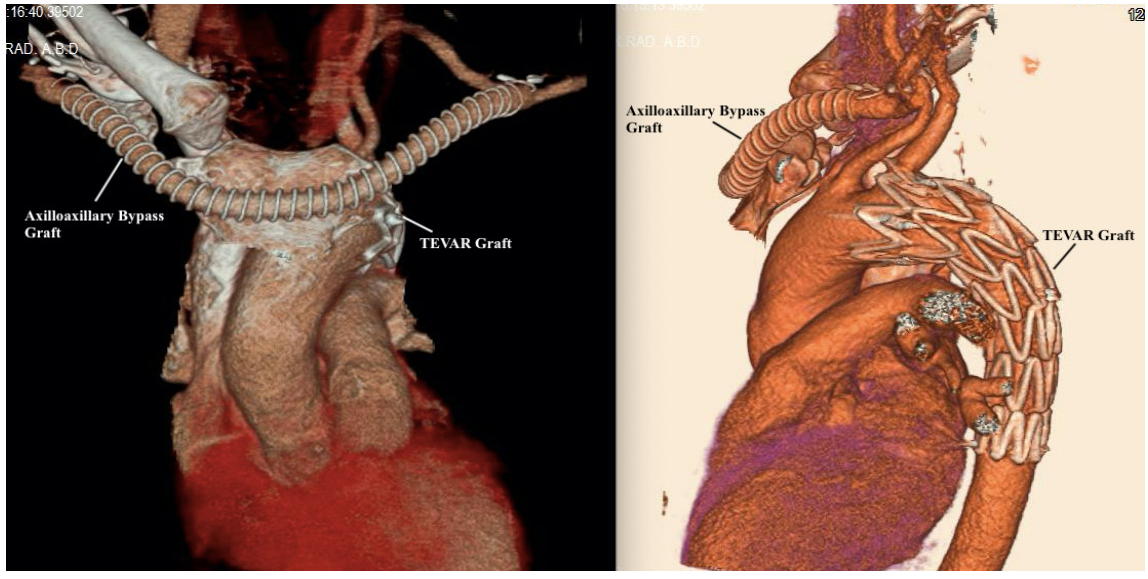

Figure 3. Hybrid treatment for Kommerell's diverticulum. Axilloaxillary bypass followed by TEVAR

TEVAR: Thoracic endovascular aortic repair

Discussion

This report presents our surgical experience and medical follow-up of the 14 patients who had ASCA. The primary goal of our surgical management in patients with ASCA is to relieve symptoms and prevent aortic dissection or rupture in patients with Kommerell's diverticulum.

The left ASCA originating from the right aortic arch is less common than the right ASCA. Kommerell's diverticulum may be found in up to 60% of patients with this variant[10,11]. Consistent with the literature in our study, only 1 (8.3%) patient had left ASCA, whereas 13 patients had right ASCA. However, in our study, unlike the literature, 14.2% of patients (2 patients) had Kommerell's diverticulum, which may be attributed to the small cohort size.

The course of aneurysms associated with Kommerell's diverticulum is not known precisely because of the rarity of the anomaly[12]. Nonetheless, rupture, or dissection of these aneurysms have been reported in the series[10,12]. Cinà et al.[10] reported that the rate of dissection or rupture was 53% among 32 patients, and Kouchoukos and Masetti[12] reported that the rate of dissection was 20% in 10 patients. In our series, only 2 patients had aneurysm related to Kommerell's diverticulum and none had rupture or dissection; however, patient patients were treated to prevent such complications.

A number of surgical approaches such as ASCA repair via right or left thoracotomy, and open surgical repair of Kommerell's diverticulum or hybrid approach with endovascular repair have been defined in the literature[9,10,13]. The thoracotomy has to be on the same side with the ASCA, ie through right thoracotomy for the right ASCA and through left throacotomy in the left ASCA with right aortic arch; otherwise, re-implantation attempts would end up with additional abberancy or when it is ligated and divised, the corresponding arm will receive pulseless nategrade flow. We preferred right thoracotomy approach in the right ASCA without Kommerell's diverticulum. We resected the right ASCA from the descending aorta and anastomosed the right subclavian artery to the right common carotid artery or the ascending aorta. We preferred left thoractomy approach in the left ASCA with a right aortic arch and performed division of the ligamentum arteriosum. The hybrid approach was preferred in the right ASCA with Kommerell's diverticulum. Kommerell's diverticulum was excluded with a stent graft following axilloaxillary bypass to establish the right upper extremity circulation. We did not prefer open surgical repair due to comorbidities in patients who had Kommerell's diverticulum concominat to the right ASCA as well as the considerable risks of the conventional open surgery.

The indication for surgical intervention in asymptomatic or symptomatic patients has not yet been clearly determined. According to a recently published consensus guideline, surgical treatment was recommended (Class 1) in symptomatic patients with ASCA and/or Kommerell's diverticulum[13]. The same consensus guideline recommends treatment when aneurysmal ASCA is greater than 3 cm in diameter and Kommerell's diverticulum is greater than 5.5 cm because of the risks of rupture and dissection[13]. Backer et al.[14] suggested that the size of the Kommerell's diverticulum being 1.5 times larger than the subclavian artery might be a point to consider surgical or endovascular intervention. We preferred the hybrid approach with TEVAR and axilloaxillary bypass in two patients with aneurysmal dilatation.

While dysphagia as a symptom is more common in grown -up patients with ASC, respiratory problems are more common in infants[15]. In our study, only one infant patient was operated against respiratory and alimentation symptoms, and the remaining patients were treated for dysphagia lusoria and/or Kommerell's diverticulum.

Study Limitations

There are 2 major limitations of the study regarding with the retrospective nature of the design and limited cohort size despite the inclusion of two centers.

Conclusion

In conclusion, according to the results of the current research, symptomic patients with ASCA or patients with Kommerell's diverticulum may be treated with conventional surgical techniques or hybrid methods safely with good mid term results. Asymptomatic patients may be followed uneventfully. Studies with multiple centers, including an increased number of patients and long-term outcomes, are warranted to establish a consensus regarding the pathology.

Ethics

Ethics Committee Approval: Our study was approved by the Institutional Ethics Committee of İstanbul Medipol University (approval no: 24/11/2022, date: 1011).

Informed Consent: Retrospective study.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Önalan MA, Öztaş DM, Alpagut U, Uğurlucan M, Concept: Öztaş DM, Erdinç İ, Design: Öztaş DM, Alpagut U, Uğurlucan M, Data Collection and/or Processing: Önalan MA, Öztaş DM, Meriç M, Analysis and/or Interpretation: Bıçakhan B, Erdinç İ, Uğurlucan M, Literature Search: Önalan MA, Bıçakhan B, Writing: Önalan MA, Meriç M, Uğurlucan M.

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