

Surgical and endovascular treatment of dysphagia lusoria in right-sided aortic arch with aberrant left subclavian artery with Kommerell diverticulum: Literature review and two case reports

Morteza Noaparast¹, Faramarz Karimian¹, Alireza Namvari^{1,*}

¹Department of Vascular Surgery, Imam Khomeini Hospital Complex, Tehran University of Medical Sciences, Tehran, Iran

Corresponding author: Dr. Alireza Namvari

Department of Vascular Surgery, Imam Khomeini Hospital Complex, Tehran University of Medical Sciences, Tehran, Iran
Email: alireza.namvari2001@gmail.com; Tel/Fax: +98 913 2646650; <https://orcid.org/0000-0002-9329-0494>

Abstract

Aberrant Subclavian Artery (ASA) is a rare anomaly within the aortic arch. In some cases, an aneurysm can also develop in the proximal ostium of the ASA which is called Kommerell diverticulum. In this article we report two symptomatic patients with a rare anatomical anomaly caused by a right sided aortic arch and left aberrant subclavian artery with Kommerell diverticulum causing dysphagia lusoria. In the first patient with hemoptysis we conducted surgical excision of the left subclavian artery origin along with bypass end-to-side bypass grafting. In the second patient we transected and ligated the left subclavian artery and with an end-to-side bypass transposition graft, followed by thoracic endovascular repair with simultaneous coil embolization of the Kommerell diverticulum.

Keywords: Endovascular, Dysphagia lusoria, Kommerell diverticulum

Introduction

Aberrant subclavian artery, as a rare anatomical variant of the origin of the subclavian artery, can occur in right or left artery [1]. This anatomical abnormality is the most common congenital vascular anomaly of the aortic arch, occurring in approximately 1% of individuals [2, 3].

An aneurysm in the proximal of ASA may also be developed, which is called Kommerell diverticulum [4, 5]. The earliest description of an anomalous right subclavian artery was reported by Bayford in 1735[4]. Also in 1936, Burckhard Friedrich Kommerell described the aortic diverticulum. It is an embryologic remnant of the fourth dorsal aortic arch, which occurs in a number of anomalies of the aortic arch system [6, 7].

In clinical manifestations, this condition is usually asymptomatic. This artery usually arises just distal to left subclavian artery and crosses in the posterior part of the mediastinum on its way to the right side upper extremity [8, 9]. In 80% of cases behind the esophagus it crosses. Some cases of this aberrant artery may cause a ring around the esophagus and trachea [10]. As a result of an aberrant right subclavian artery, dysphagia occurs, that is termed as dysphagia lusoria, this is condition

as a rare complication, occur in our cases [11]. In addition to dysphagia, aberrant right subclavian artery may cause dyspnea, stridor, fever or chest pain [12]. Also aberrant right subclavian artery can compress recurrent laryngeal nerve leading to a palsy of that nerve, as Ortner's syndrome [13, 14].

In addition, aberrant right subclavian artery arises from a dilated segment of proximal of descending section of aorta, as Diverticulum of Kommerell, it is alternatively known as a lusorian artery [12, 15]. Some of techniques of surgery such as direct surgical repair or endovascular treatment, have been described, in this study surgical and endovascular treatment of dysphagia lusoria in right-sided aortic arch with aberrant left subclavian artery with kommerell diverticulum in two cases have been described.

Case presentation #1: Surgical repair

A 36-year-old female was admitted to our hospital with chest pains and hemoptysis evolving and progressing within 3 days, with complaints of intermittent dysphagia for several years. After ruling out cardiac problems, chest Computed Tomography Angiography (CTA) was conducted, and a bronchoscopy was requested from the

pulmonologist. The bronchoscopy assessment resulted in no pathologic finding, but the CTA showed a Right Aortic Arch (RAA) with Left Aberrant Subclavian Artery (LASA), which had an aneurysmal origin (Kommerell diverticulum) (Figure 1 and 2). Therefore, emergent surgery was planned.



Figure 1. Right Aortic Arch (RAA) with Left Aberrant Subclavian Artery (LASA)



Figure 2. Right Aortic Arch (RAA) with Left Aberrant Subclavian Artery (LASA)

performed and the aortic arch and descending aorta was circumferentially freed. The proximal origin of left subclavian artery aneurysm was found adjacent to the descending aorta. Therefore, dissection from all surrounding tissues was carried out in order to partially clamp the aorta. The aneurysm was then successfully excised; the exit site of subclavian artery was repaired with 4/0 Prolene stitches in continuous formation. Subsequently, resection was continued to the left side of esophagus and the subclavian artery ligated. The position was then changed to supine, the base of the neck exposed, the left subclavian artery and common carotid artery was explored, and using an ePTFE bypass graft, the left common carotid artery was bypassed to the left subclavian artery with 5/0 Prolene sutures. Finally, distal pulses of superior limbs were checked, and finally the site of incision repaired and closed.

Case presentation #2: Endovascular repair

A 27-year-old male was admitted to our hospital with complaints of inability to swallow for 45 days, with otherwise healthy general condition. A chest CTA was ordered which showed a RAA with a LASA and a 22mm Kommerell diverticulum (KD) noted in its proximal ostium (Figure 3 and 4). The KD was severely compressing the esophagus, and so dysphagia lusoria was diagnosed. Therefore, elective Thoracic Endovascular Aortic Repair (TEVAR) was planned.



Figure 3. RAA with a LASA and Kommerell diverticulum

Procedure

Under general anesthesia and left lateral position, right posterolateral thoracotomy was



Figure 4. RAA with a LASA and Kommerell diverticulum

Procedure

The repair was conducted in two stages. In the first stage, and under general anesthesia, the patient received transverse supraclavicular cervical incision, and the surgical dissection was conducted between the two heads of the sternocleidomastoid muscles. The carotid and subclavian arteries were dissected circumferentially deep into the mediastinum, the proximal subclavian artery transected and the proximal stump ligated, and finally an end-to-side anastomosis completed without tension. Subsequently on day seven of the post-operative recovery, and under spinal anesthesia, right femoral artery arteriotomy was conducted and percutaneous access achieved on the left femoral artery (which was used to advance a 5F Cobra2 angiographic catheter in the Kommerell diverticulum within the Left Aberrant Subclavian Artery), and the right radial artery (which was used to advance a 5F pigtail angiographic catheter in the ascending aorta). An extension cuff stent-graft with a size of 26-80 (Cook Medical, Bloomington, IN, USA) was then inserted from the right femoral artery and deployed from the level of the Right Subclavian Artery (which was superior to the LASA), while jailing the Cobra catheter in the KD. Subsequently, via the jailed Cobra catheter, two large pushable Nester coils (Cook Medical, Bloomington, IN, USA) were deployed behind the stent-graft and within the KD (Figure 5). One month post-procedural CTA showed no endoleak of any type, no migration of the stent, patency of the cerebral vessels, and a mild shrinkage of the KD. The patient's symptoms related to his dysphagia were completely resolved



Figure 5. CTA after stent graft placement

Discussion

Most of abnormalities which was congenital in supra-aortic trunk are explained by of the fourth aortic arch interruption between descending aorta and right subclavian artery [16]. This condition as the aberrant subclavian artery is reported approximately in 0.5 to 1.8 percentage of populations [6, 8] which in comparison was more common on right rather than left side and an aberrant left subclavian artery in right aortic arch is less common [17, 18]. 60% of cases with aberrant subclavian artery influenced with aneurysm as Kommerell's diverticulum [19]. Left aberrant subclavian artery from right-sided aortic arch passes behind esophagus to left arm [20].

In normal development of aortic arches, right dorsal aorta regresses caudal to the origin of the 7th intersegmental artery which gives rise to the right subclavian artery [21]. In formation of an aberrant right subclavian artery, the regression occurs instead between the 7th intersegmental artery and the right common carotid so that the right subclavian artery is then connected to the left dorsal aorta via the part of the right dorsal aorta which normally regresses [22]. During growth, the origin of the right subclavian artery migrates until it is just distal to that of the left subclavian [3, 23].

Incidence of right side reported as 59 percentages in type I, 39.5 percentage in type II, and 3.08 percentage in type III of aortic arches [24]. In our cases, the second case whom had a type II aortic arch, left carotid artery was highest superior branch,

followed by right carotid artery then the right subclavian artery, and finally the Left Aberrant Subclavian Artery (RAA + LASA) [25]. Kommerell diverticulum, most common, occurs in origin of Left Aberrant Subclavian Artery, and been reported to occur in all of RAA + LASA cases [3], sometimes RAA in adults are detected as atherosclerotic consequence such as dissection, dilatation, and/or compression effects in adjacent organs. Clinical manifestations may be included as dyspnea, dysphagia, wheezing, stridor, recurrent pneumonia, cough, chest pain, choking and obstructive emphysema [26].

Hanault et al. reported first case of LASA in 1735 [27]. In addition, Byford et al. in 1789 clinically described LASA and invented the word “dysphagia lusoria” as a description [28]. Histopathologic studies recently show intara cystic necrosis in a kommerell diverticulum wall which made the diverticulum vulnerable to dissection and rupture [29]. Compression effect to other organs and also surgical complexity may be cause high rate of morbidity and mortality [29].

Some approaches of surgery for right aberrant subclavian artery in addition to Kommerell’s diverticulum have been described, classically as a surgical repair approach which are two-staged [15]. After completion of bypass from carotid to subclavian artery, Kommerell’s diverticulum using cardiopulmonary bypass excluded by a direct approach [30]. Recently, endovascular procedures have been suggested, which in one of our cases, elective case, have been used [8, 31].

For our patients, treatment was indicated since both were symptomatic. In patient #1, emergent surgical repair was chosen due to active hemoptysis and unavailability of endovascular repair as a rapid option. In patient #2, TEVAR was a viable option since the patient was hemodynamically stable. As part of the pre-procedural planning phase for patient #2, both options were discussed with the patient, and the clinical team in close collaboration with the patient decided on the less invasive endovascular repair, with the option to proceed to a surgical bailout if necessary. Since the endovascular approach did not preclude the patient to any future surgical intervention, it was deemed an appropriate treatment modality. Both treatment options proved effective in our two patients in follow up imaging.

In following some others studies in this field, have been discussed. In some other studies mentioned that, asymptomatic cases and without an aneurysm not have any indication of surgery [32].

Two cases, which was discussed in this study were symptomatic and have compression effect to near organs. In addition, in younger cases, some other anomalies such as right aortic arch with vascular ring ductus arteriosus which encompass the esophagus, may introduce similar clinical manifestations [33]. Also in children and infants because of trachea flaccidity, respiratory manifestations will be more common [34]. So indication of surgery will be depending on symptoms and age of cases. In 1946, Gross suggested simple isolation and ligation of the aberrant subclavian artery [35], however this approach were acceptable in children, and in adults it causes Steal Syndrome or upper limb ischemia with high probability [36, 37].

Kommerell diverticulum dissection and rupture is reported, it is still underreported because of its rarity. Austin et al. estimated rate of diverticulum rupture in 32 patients (19%) [38]. Cina et al. in dissection and rupture rate of diverticulum reported in 32 cases (53%) with RAA and LASA [12]. In addition, Nicholas et al. reported 2 dissections in 10 cases. In documents there are not any definite threshold of surgery in asymptomatic cases [36]. Cina et al. suggested surgery in cases with diameters higher than 3cm [12], however Ota et al. suggested 5cm as its threshold [39], however in general elective intervention is widely recommended [40]. In addition, Lococo et al. presented a case with a 30mm Kommerell diverticulum in an asymptomatic case; however, general condition of patient was not appropriate for surgery, in 12-month follow-up, their case was remained asymptomatic [41], but two cases of our study was symptomatic and needs to surgery.

Right Aortic Arch with a Left Aberrant Subclavian Artery is an extremely rare anatomical anomaly which may results in Kommerell diverticulum and cause dysphagia lusoria. Treatment is indicated in symptomatic cases or where the size exceeds 3cm. We treated two symptomatic patients, one emergently with surgical techniques, and another electively with hybrid surgical and endovascular techniques. The symptoms of both patients has subsided in post-operative follow up imaging and continue to function well in the mid-term. Further randomized studies comparing surgical and endovascular treatment modalities would be required to determine superiority of each method.

Author contribution

All authors contributed equally and approved the final version of manuscript.

Conflict of Interest

Authors declare no conflicts of interest.

Consent for publication

Written informed consent was obtained from the patients for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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