

Dysphagia Lusoria: Uncommon Cause of Complete Esophageal Obstruction – Case Report

Dysphagia lusoria – neuobičajen uzrok kompletne opstrukcije jednjaka – prikaz slučaja

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Abstract. Aim: Vascular anomalies only rarely cause dysphagia, as patients are predominantly asymptomatic. The aberrant right subclavian artery (ARSA), the most common anomaly of the aortic arch, may cause esophageal compression, resulting in dysphagia known as *dysphagia lusoria*. Although the majority of patients require no treatment or only conservative management, symptomatic cases necessitate surgical intervention or a combination of surgical and endovascular procedures. The aim of this article is to present one such symptomatic case. **Case report:** We present a patient with a rare cause of dysphagia due to ARSA (arteria lusoria), which led to complete esophageal obstruction. Following surgical resection and transposition of the right subclavian artery, combined with endovascular sealing using an Amplatzer™ Vascular Plug II (Abbott Medical, Plymouth, MN, USA), the patient experienced complete resolution of symptoms. **Conclusion:** The literature on dysphagia lusoria is largely limited to case reports. Given the small number of patients requiring treatment, it is difficult to establish a single optimal therapeutic approach. Therefore, each case should be evaluated by a multidisciplinary team, with an emphasis on minimally invasive options.

Keywords: deglutition disorders; endovascular aneurysm repair; endovascular procedures; subclavian artery

Sažetak. Cilj: Vaskularne anomalije iznimno rijetko uzrokuju disfagiju jer su bolesnici uglavnom asimptomatski. Aberantna desna arterija subklavija (ARSA), najčešća anomalija luka aorte, kada komprimira jednjak, uzrokuje disfagiju poznatu kao *dysphagia lusoria*. Iako većini pacijenata nije potrebno liječenje ili je ono samo konzervativno, simptomatski bolesnici trebaju operaciju ili kombinaciju operacije i endovaskularnog postupka. Cilj je ovog članka prikazati jedan od simptomatskih slučajeva. **Prikaz slučaja:** Prikazujemo bolesnika s neuobičajenim uzrokom disfagije, uzrokovane aberantnom desnom arterijom subklavijom (*arteria lusoria*) koja je dovela do potpune opstrukcije jednjaka. Nakon kirurškog zahvata resekcije i transpozicije desne arterije subklavije i endovaskularnog brtvljenja pomoću uređaja Amplatzer™ Vascular Plug II (Abbott Medical, Plymouth, MN, SAD), pacijentu su svi simptomi nestali. **Zaključak:** Literatura o disfagiji luzoriji ograničena je uglavnom na prikaze slučajeva. Zbog vrlo malog broja pacijenata koji trebaju liječenje, teško je istaknuti jednu opciju liječenja kao optimalnu. Iz tog razloga, multidisciplinarni tim trebao bi odlučiti o svakom slučaju imajući u vidu minimalnu invazivnost.

Ključne riječi: arterija subklavija; endovaskularni popravak aneurizme; endovaskularni postupak; poremećaj gutanja

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INTRODUCTION

Vascular anomalies are rare cause of dysphagia¹. Aberrant right subclavian artery (ARSA or arteria lusoria), which arises as the last branch of the aortic arch, coursing posterior to the esophagus, is the most common developmental anomaly of the aortic arch. It occurs in approximately 0.5% to 2.5% of humans^{2,3}, and may be associated with other vascular anomalies⁴. In most cases, it is asymptomatic². Only an extreme minority of patients have symptoms of esophageal compression – dysphagia lusoria, due to ARSA itself, or aneurysmal dilatation of its origin from descending thoracic aorta, referred to as Kommerell's diverticulum. The latter may also compress trachea, causing cough as a symptom. In symptomatic patients, the diagnosis may be suspected by esophageal x-ray with barium contrast, digital subtraction angiography of the aortic arch, or esophagogastroduodenoscopy (EGD), and confirmed by computed tomography angiography (CTA) or magnetic resonance imaging^{1,5-7}. Conservative medical treatment (consisting of lifestyle modifications, such as dietary changes) may be sufficient for patients with minimal symptoms¹, but recurrent or persistent symptoms and failure of conservative treatment require surgical correction^{1,2,6-10}.

There are mostly case reports found in literature describing various treatment options for dysphagia lusoria, from a maximal surgical approach (sternotomy or thoracotomy with ligation, transposition or bypass procedure of ARSA to common carotid artery) to less invasive thoracic endovascular aortic repair (TEVAR) combined with some type of surgery⁶⁻¹¹. Given the small number of symptomatic patients requiring treatment and the variety of treatment options, it is difficult to single out one as optimal. This highlights the importance of multidisciplinary approach, in which vascular surgeon and interventional radiologist decide every case of this pathology, and minimally invasiveness should be the goal.

This case describes a successful, minimally invasive, two-stage procedure without complex thoracic surgery – a combination of ARSA resection and transposition to the right common carotid

artery via supraclavicular incision and endovascular Amplatzer™ Vascular Plug II (Abbott Medical, Plymouth, MN, USA) sealing with immediate symptom relief as an excellent result.

CASE REPORT

A 67-year-old female, with persistent progressive dysphagia to solid food since childhood, was referred to the emergency department because of sudden inability even to drink. This severe symp-

A multidisciplinary team approach in this pathology is of utmost importance, addressing each case. The goal should be minimal invasiveness leading to resolution of dysphagia.

tom lasted for two days. EGD showed complete esophageal obstruction with extraesophageal pulsatile compression. CTA revealed ARSA, emerging as the last branch of the aortic arch, after the joint orifice of both common carotid arteries and the left subclavian artery, coursing posterior to the esophagus and compressing it with complete esophageal obstruction, with an aneurysm on its orifice – the Kommerell's diverticulum (KD). All this was consistent with the diagnosis of dysphagia lusoria (Figure 1). The patient was admitted to the Department of Vascular Surgery for total parenteral nutrition and urgent treatment.

There were three treatment options available for this pathology^{6,10,11} – classic surgical transthoracic correction consisting of suturing the KD and reinsertion of ARSA in its anatomically correct location; thoracic aortic endovascular repair (TEVAR) covering the orifices of both subclavian arteries with bilateral carotid to subclavian bypass; and right-sided subclavian to carotid transposition and endovascular plugging of the KD using Amplatzer™ Vascular Plug II. The multidisciplinary team, formed of vascular surgeon and interventional radiologist, determined the last-mentioned hybrid procedure as most appropriate and as minimally invasive approach as possible for this patient.

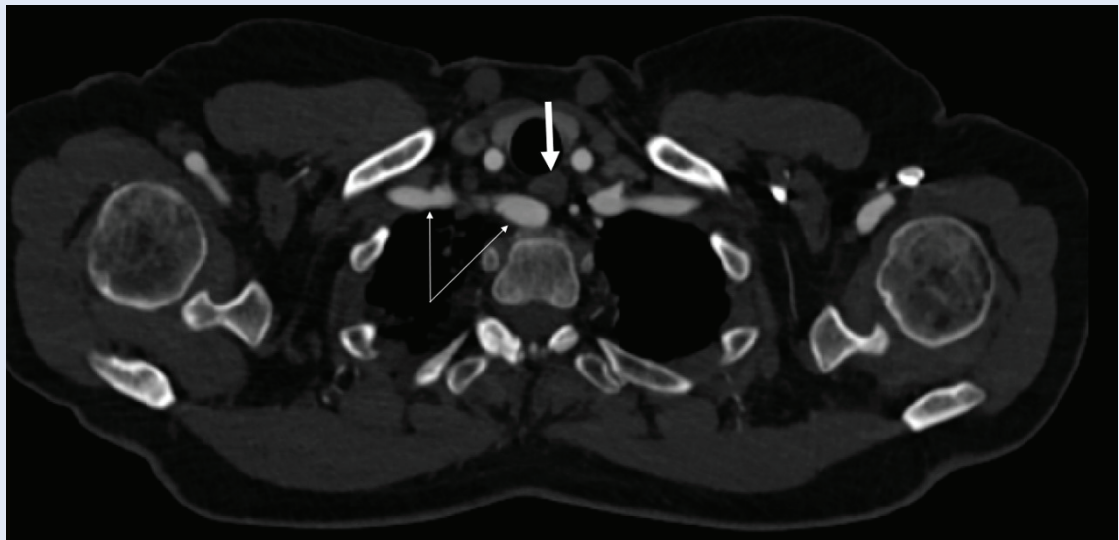


Figure 1. Arteria lusoria (thin arrows) leading to complete esophageal obstruction (thick arrow).

Surgery

Under general anaesthesia a right-sided supraclavicular incision with anterior scalenotomy was made, taking care to avoid injury to the phrenic nerve. By dissecting the right subclavian artery, all visible branches were ligated and transected and retrojugular tunnel was created towards the right common carotid artery. Three minutes after an intravenous bolus of 5000 IU heparin sodium solution (Heparin, Belupo, Croatia), the ARSA was clamped, transected and double-ligated as far intrathoracic as possible, forming a stump. The distal end was then transposed retrojugularly to the right common carotid artery using 6-0 polypropylene monofilament running suture. The wound was closed in the usual manner, closing the platysma and skin, with a suction drain positioned near the anastomosis. After

awakening from anaesthesia, the patient no longer had dysphagia. The patient was transferred to the recovery room for monitoring.

Endovascular procedure

The next day, the second stage of the procedure was performed via right-sided transfemoral approach through a 90 cm long 8 Fr Destination™ Guiding Sheath (Terumo Interventional Systems, Terumo Medical Corporation, Somerset, NJ, USA). The lumen of the KD was selectively cannulated with a guiding catheter, and an 18 mm diameter vascular plug (Amplatzer™ Vascular Plug II) was placed into it. The femoral puncture site was closed with Angioseal® VIP vascular closure device (Terumo Interventional Systems, Terumo Medical Corporation, Somerset, NJ, USA).

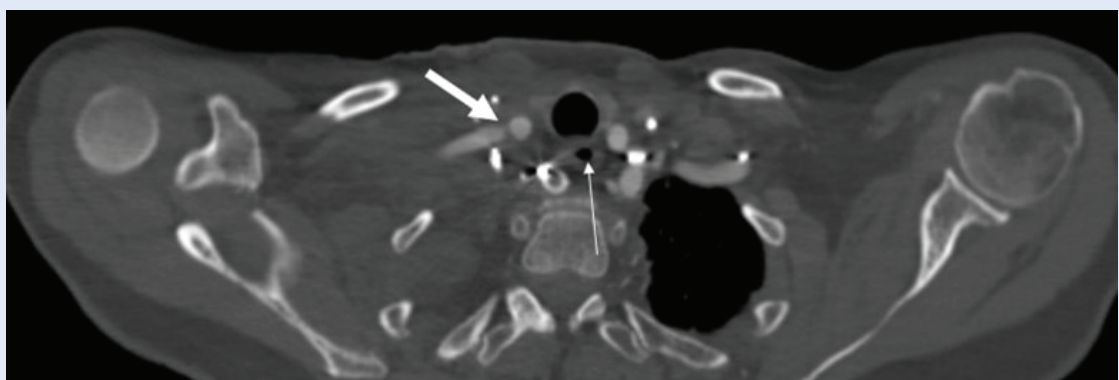


Figure 2. Re-expanded esophagus (arrow) after transection of ARSA.

Postoperative period

Control CTA showed re-expansion of the esophagus, complete occlusion of the remaining stump of the lusoria artery and KD, with regular patency of the transposed artery, supra-aortic arteries and aortic arch, and without signs of complications (Figures 2, 3).

The patient was discharged on the third postoperative day without dysphagia, able to eat solid food and drink, which was confirmed at follow-up examinations.

DISCUSSION

Due to the low prevalence of symptomatic patients with ARSA, there is no standard treatment procedure or unique optimal approach. Classical surgical procedures usually involve extensive procedures consisting of thoracotomy or sternotomy, repositioning of the ARSA in an anatomically normal position with partial clamping of the aortic arch in a deep hypothermic circulatory arrest, and suturing of ARSA orifice on the descending thoracic aorta, with all possible complications and a high mortality rate^{6, 10–12}. A less invasive technique is TEVAR with unilateral or bilateral subclavian to common carotid artery bypass, or subclavian to subclavian cross-over bypass, which is also far more complex and more expensive procedure than used here. The approach used in this patient is a minimally invasive procedure that has resulted in complete resolution of dysphagia and is far more patient-friendly than sternotomy or thoracotomy in terms of the extent of the surgery, postoperative pain, and perhaps not least, the size of the postoperative scar. The point of subclavian to carotid transposition is to transect the ARSA and all of its available branches, resulting in retraction of the proximal part of the ARSA and the disappearance of compression on the esophagus, and thus dysphagia. None of the case reports about dysphagia lusoria found in the literature describe the final stage of dysphagia – a complete obstruction of the esophagus to even water and saliva. Complete obstruction of the digestive tract constitutes an urgent indication for decompression and reconstruction, in this case resolving external esophageal compression to avoid long-term nasogastric feeding tube

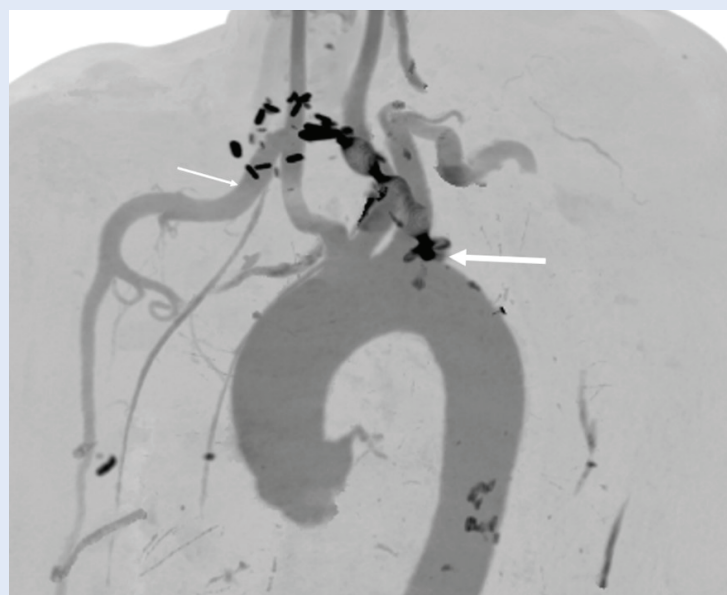


Figure 3. Transposed ARSA (thin arrow) and Amplatzer™ Vascular Plug II (thick arrow) without blood flow in ARSA stump on control CTA.

ARSA transection is the key point for re-dilation of the esophagus.

or parenteral nutrition. Baring that in mind, the multidisciplinary team concluded that this aforementioned procedure is optimal for this patient. Only for technical reasons, it was divided into two days as a two-step procedure, but it could be easily done in a hybrid operating room as a single-stage, which would be the recommendation. If TEVAR had been used, it would have also covered the joint orifice of the common carotid arteries, which was near the KD. That approach would require more extensive surgery in the form of revascularization of both arms. For this reason, in this case, the Amplatzer™ Vascular Plug II proved to be the best and most elegant solution to stop the blood flow in the KD and ARSA stump, without the need for TEVAR, as minimally invasive option.

Postprocedural CTA showed an excellent result – a complete resolution of esophageal lumen without any external compression, properly patent transposition, no blood flow from aorta to the ARSA stump, and no signs of any complication.

Even though every patient should be evaluated for itself, the authors recommend this relatively

simple procedure as the first choice for treatment of symptomatic dysphagia lusoria.

CONCLUSION

Even though only minority of patients with arteria lusoria are symptomatic, this pathology presents an important cause of extraluminal esophageal compression and dysphagia with variety treatment options, none of which proven as optimal strategy, but the goal is minimal invasiveness. For this reason, our suggestion for treatment is a combination of endovascular procedure (Amplatzer™ Vascular Plug II) and surgical resection of subclavian artery with its transposition to carotid artery, because without the resection of ARSA, the extraluminal compression remains unsolved.

Conflicts of Interest: Authors declare no conflicts of interest.

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