

## A rare condition secondary to aberrant right subclavian artery syndrome: Aphagia lusoria

*Aberran sağ subklaviyen arter sendromuna sekonder gelişen nadir bir durum: Afajî lusoria*

Fatih Ada<sup>1</sup>, Salih Güler<sup>1</sup>, Fatmagül Demir<sup>2</sup>, Ekber Şahin<sup>2</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Sivas Cumhuriyet University School of Medicine, Sivas, Türkiye

<sup>2</sup>Department of Thoracic Surgery, Sivas Cumhuriyet University School of Medicine, Sivas, Türkiye

### ABSTRACT

Dysphagia lusoria is a well-described clinical entity caused by aberrant right subclavian artery. Herein, we present a 42-year-old male case with aberrant right subclavian artery-associated aphagia and progressed to aphagia lusoria presentation. To the best of our knowledge, this is the first associated case presented with aphagia.

**Keywords:** Aberrant right subclavian artery syndrome, aphagia lusoria, COVID-19, dysphagia lusoria, surgical treatment.

Aberrant right subclavian artery syndrome (ARSA) is frequently seen among aortic arch anomalies and the incidence of the disease is 0.5 to 1.8%.<sup>[1,2]</sup> As 60 to 80% of the patients are asymptomatic, the diagnosis is usually established in incidental or postmortem examinations.<sup>[3]</sup> Symptomatic patients usually present with solid food intolerance, postprandial bloating, or chest pain. Symptoms often vary depending on the posture. Cough, thoracic pain, and Horner's syndrome are rare symptoms of ARSA. Dysphagia lusoria due to vascular compression of the esophagus is a well-known clinical entity. Patients are usually admitted to internal medicine and gastroenterology departments initially and get diagnosed. Upper endoscopies of patients are usually normal. A typical external esophageal compression appearance is seen in the barium esophagram. However, the definitive diagnosis is made by computed tomography angiography (CTA) and magnetic resonance angiography (MRA).

### ÖZ

Aberran sağ subklaviyen arter sendromunun neden olduğu disfaji lusoria, iyi tanımlanmış bir klinik tablodur. Bu yazıda, aberran sağ subklaviyen arter sendromu ile ilişkili afajili ve afajî lusoria tablosuna ilerleyen 42 yaşında bir erkek olgu sunuldu. Bildiğimiz kadarıyla, bu olgu afajî ile birlikte seyreden ilk ilişkili olgudur.

**Anahtar sözcükler:** Aberran sağ subklaviyen arter sendromu, afajî lusoria, COVID-19, disfaji lusoria, cerrahi tedavi.

Aphagia, the most advanced form of dysphagia lusoria, has not been described in the literature. As patients usually undergo surgery after diagnosis, dysphagia almost never progresses to aphagia. Herein, we present the management of a male ARSA patient who postponed the treatment due to novel coronavirus disease 2019 (COVID-19) pandemic and progressed to aphagia lusoria.

### CASE REPORT

A 42-year-old male patient was admitted to our clinic with the diagnosis of ARSA. His medical history revealed that he had a dysphagia which was present for a long time and worsened about one year before. A surgical operation was planned after his admission to the outpatient clinic at the beginning of March 2020. However, the patient did not attend to the hospital for operation, due to the concerns about COVID-19 pandemic. He was admitted to the

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**Correspondence:** Fatih Ada, MD. Cumhuriyet Üniversitesi Tıp Fakültesi Kalp ve Damar Cerrahisi Anabilim Dalı, 58140 Sivas, Türkiye.

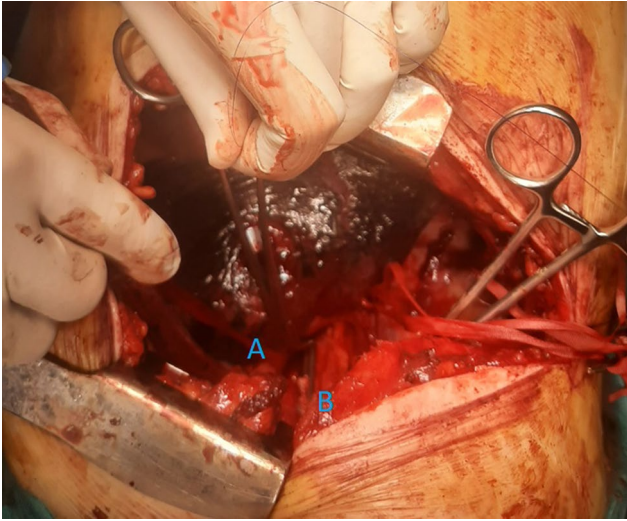
Tel: +90 346 - 219 10 10 e-mail: drfatihada@gmail.com

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**Figure 1.** (a) Esophagus, (b) Clamped right aberrant subclavian artery.

hospital with impaired metabolic values and a weight loss approximately 35 kg at the end of May 2020. He could only drink water for the last three days and could not even swallow saliva on the day of admission to hospital. His routine laboratory tests showed that he was slightly anemic (hemoglobin: 13.2 g/dL), his total protein and albumin was decreased (total protein: 5.7 g/dL, albumin: 3.1 g/dL), and creatinine was

slightly elevated (creatinine: 1.32 mg/dL). Coagulation parameters and thyroid function tests were found to be normal. Intravenous support was initiated after the patient was hospitalized. Following hemodynamic recovery, the patient underwent ligation and division of the aberrant right subclavian artery with right anterolateral thoracotomy (Figure 1). Also, the right carotid artery and the right subclavian artery bypass was done using a polytetrafluoroethylene (PTFE) graft with a supraclavicular approach simultaneously (Figure 2). The patient was drinking water easily on postoperative Day 1 and tolerated juicy foods on Day 3. His diet was adjusted predominantly as juicy foods and discharged uneventfully one week after operation. During follow-up, he could consume solid foods without any problem (Figure 3).

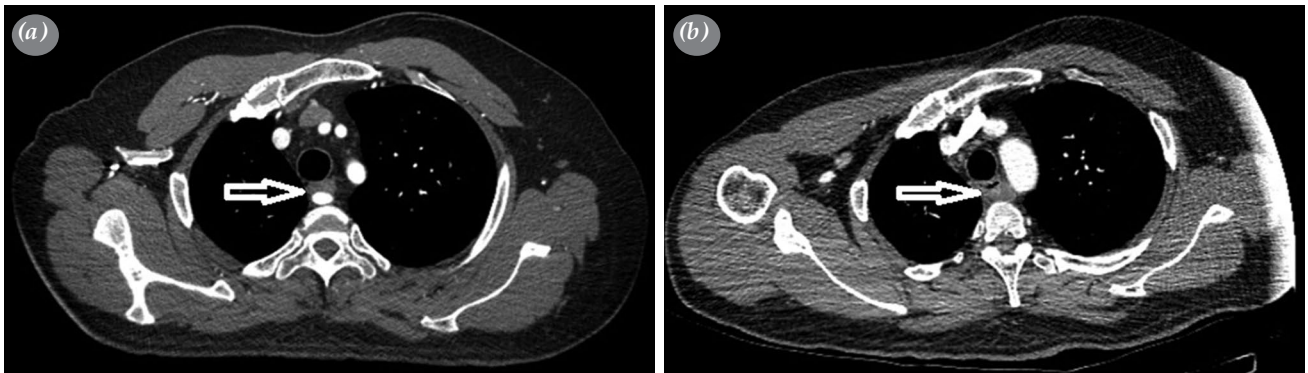
## DISCUSSION

Aberrant right subclavian artery syndrome was first described by Hunauld<sup>[4]</sup> in 1735. Approximately five decades later, Bayford defined dysphagia lusoria and, thereafter, it is also called Bayford-Autenrieth dysphagia.<sup>[5,6]</sup>

Symptoms are usually dysphagia, dyspnea, retrosternal pain, cough, and weight loss in dysphagia lusoria. In a review consisting of 141 reports, the symptoms of female patients appeared earlier than men.<sup>[6]</sup> In our patient, the main symptoms were



**Figure 2.** (a) Preoperative appearance (3-dimensional computed tomography). (b) Postoperative appearance (3-dimensional computed tomography).



**Figure 3.** (a) Esophageal lumen is totally closed. (b) Esophageal lumen is open.

initiated with solid food dysphagia, cough, and weight loss. Due to delay in treatment, aphagia was developed and surgery was done under this condition. Barium esophagram is an important test for diagnosis of dysphagia lusoria. However, CTA, MRA and aortography are more valuable in definitive diagnosis. In our case, the diagnosis was made by CTA. Esophageal manometric measurements are used to predict the surgical outcome and follow-up after surgery.<sup>[7]</sup> In addition to direct complications of ARSA due to vascular ring, there are also late complications such as aneurysm and tracheoesophageal fistula.<sup>[8,9]</sup> Different congenital anomalies may accompany to ARSA. For instance, ARSA association with aortic coarctation has been described in the literature.<sup>[10]</sup>

Treatment depends on severity of the symptoms of disease. Diet adjustment may be sufficient in patients who have relatively slight compression, but most patients are candidates for surgical treatment.<sup>[11]</sup> In our case, surgical treatment was applied, as it was impossible to apply diet therapy.

Surgical mortality was reported between 16 and 25% in previous publications, but this rate has reached almost 0% with surgical and technological developments in recent years.<sup>[3,12]</sup> There is no standard approach for surgical treatment. It depends on the location of ARSA, complications, presence of other cardiovascular anomalies and experience of center.<sup>[10,13]</sup> Surgical approach may be right supraclavicular, right anterolateral thoracotomy, and median sternotomy.<sup>[14]</sup> The main goal of surgical treatment is ligation alone or ligation and division of the aberrant subclavian artery. Reimplantation or bypass of the subclavian artery is essential for upper limb perfusion. In our case, ligation and division of ARSA were performed from a right anterolateral thoracotomy approach. Also, right carotid subclavian bypass operation with a

PTFE graft was performed simultaneously. It should be kept in mind that this approach can be used in eligible patients, owing to its excellent exposure and promising surgical results.

In conclusion, in addition to the direct effects of the COVID-19 pandemic, its indirect effects have caused many different clinical pictures. One of these is aphagia lusoria symptom that has not been described before in the literature. As a matter of fact, our case was planned to be treated before proceeding to this situation, but delayed admission caused this result. The fight against the COVID-19 pandemic continues actively all over the world. However, this is only the visible side of the iceberg, and it would take a long time for the consequences of invisible side to appear.

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**Data Sharing Statement:** The data that support the findings of this study are available from the corresponding author upon reasonable request.

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## REFERENCES

1. Carrizo GJ, Marjani MA. Dysphagia lusoria caused by an aberrant right subclavian artery. *Tex Heart Inst J* 2004;31:168-71.
2. Meher R, Sabharwal A, Singh I, Raj A. Dysphagia due to a rare cause. *Indian J Surg* 2004;66:300.

3. Levitt B, Richter JE. Dysphagia lusoria: A comprehensive review. *Dis Esophagus* 2007;20:455-60.
4. Hunauld PM. Examen de quelques parties d'un singe. *Histoire de l'Académie Royale des Sciences* 1735;2:516-23. [Abstract]
5. Bayford D. An account of a singular case of obstructed deglutition. London: Medical Society of London; 1787.
6. Polguy M, Chrzanowski Ł, Kasprzak JD, Stefańczyk L, Topol M, Majos A. The aberrant right subclavian artery (arteria lusoria): The morphological and clinical aspects of one of the most important variations--a systematic study of 141 reports. *ScientificWorldJournal* 2014;2014:292734.
7. Janssen M, Baggen MG, Veen HF, Smout AJ, Bekkers JA, Jonkman JG, et al. Dysphagia lusoria: Clinical aspects, manometric findings, diagnosis, and therapy. *Am J Gastroenterol* 2000;95:1411-6.
8. Naqvi SEH, Beg MH, Thingam SKS, Ali E. Aberrant right subclavian artery presenting as tracheoesophageal fistula in a 50-year-old lady: Case report of a rare presentation of a common arch anomaly. *Ann Pediatr Cardiol* 2017;10:190-3.
9. Kamiya H, Knobloch K, Lotz J, Bog A, Lichtenberg A, Hagl C, et al. Surgical treatment of aberrant right subclavian artery (arteria lusoria) aneurysm using three different methods. *Ann Thorac Surg* 2006;82:187-90.
10. Karakuş E, Akyüz M, Işık O. Surgical planning of aberrant right subclavian artery and aortic coarctation co-existence. *Turk Gogus Kalp Dama* 2018;26:163-4.
11. Şahin Y, Aydın Şahin D, Kervancıoğlu M. Aberrant sol subklavyen arter basısına bağlı yutma güçlüğü olan bir olgu. *ACU Sağlık Bil Derg* 2018;9:327-30.
12. Ota T, Okada K, Takanashi S, Yamamoto S, Okita Y. Surgical treatment for Kommerell's diverticulum. *J Thorac Cardiovasc Surg* 2006;131:574-8.
13. Kouchoukos NT, Masetti P. Aberrant subclavian artery and Kommerell aneurysm: Surgical treatment with a standard approach. *J Thorac Cardiovasc Surg* 2007;133:888-92.
14. Atay Y, Engin C, Posacioglu H, Ozyurek R, Ozcan C, Yagdi T, et al. Surgical approaches to the aberrant right subclavian artery. *Tex Heart Inst J* 2006;33:477-81.