Dysphagia lusoria caused by aberrant right subclavian artery associated with truncus bicaroticus in an 8-month-old girl. Case report and review of literature

Melpomeni Bizhga,[1*] Virtut Velmishi,[2*] Lorena Sila,[3] Albert Koja,[4] Stiljan Hoxha[5]

[1]Service of Pediatric Pneumology, Mother Teresa Hospital, Tirana, Albania; [2]Service of Pediatric Gastroenterology, Mother Teresa Hospital, Tirana, Albania; [3]Pediatric Department, Mother Teresa Hospital, Tirana, Albania; [4]Service of Pediatric Cardiology, Mother Teresa Hospital, Tirana, Albania; [5]Division of Cardiac Surgery, Department of Surgery, Dentistry, Pediatrics and Gynecology, University of Verona, Italy

*These authors contributed equally.

Correspondence: Virtut Velmishi, Service of Pediatric Gastroenterology, Mother Teresa Hospital, Dibra Street Nr 372, Tirana, Albania.
E-mail: tutimodh@yahoo.com

Key words: dysphagia lusoria, case report, aberrant right subclavian artery.

Contributions: VV performed the upper digestive endoscopy and drafted the manuscript; MB performed a bronchoscopy and conceived the manuscript; LS provided images and drafted the manuscript; AK approved the manuscript; SH performed surgical procedure, provided images during the intervention, and performed critical revision of the manuscript. All authors read and approved the manuscript.

Funding: none.

Conflict of interest: the authors declare no potential conflict of interest, and all authors confirm accuracy.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient included in this study.

Patient consent for publication: the patient's parents gave their written consent to use the patient's personal data for the publication of this case report and any accompanying images.

Availability of data and materials: all data underlying the findings are fully available.

Received: 27 January 2024.
Accepted: 2 April 2024.

Publisher's note: all claims expressed in this article are solely those of the authors and do not necessarily represent those of their affiliated organizations, or those of the publisher, the editors and the reviewers. Any product that may be evaluated in this article or claim that may be made by its manufacturer is not guaranteed or endorsed by the publisher.

Licensee PAGEPress, Italy
La Pediatria Medica e Chirurgica 2024; 46:332
doi:10.4081/pmc.2024.332

Abstract

Dysphagia lusoria is a rare pediatric condition caused by extrinsic compression of the esophagus by an abnormal subclavian artery. The most common congenital abnormality in aortic arch development is an aberrant right subclavian artery. The retroesophageal right subclavian artery is typically symptomatic in 10-33% of cases. The patient, an 8-month-old girl with a history of early dysphagia and stridor, was diagnosed with an abnormal right subclavian artery. She was admitted to the pneumology service multiple times due to stridor, vomiting, and failure to thrive. During hospitalization at the gastroenterology service, a barium swallow and an upper digestive endoscopy indicated an abnormal right subclavian artery, which was confirmed by an Angiography CT scan. She underwent surgery at the age of sixteen months. All symptoms are resolved following surgical intervention, and the patient is still asymptomatic and in good clinical condition 12 months later. Every physician should be aware of abnormal right subclavian arteries and their clinical symptoms in children and adults in order to recognize and diagnose them early. Only an early evaluation may reduce complications such as delayed physical growth, dysphagia, and recurrent respiratory infections.

Introduction

Dysphagia lusoria is a rare finding in children caused by extrinsic compression of the esophagus by the aberrant subclavian artery. An aberrant right subclavian artery is the most frequent congenital abnormality in the development of the aortic arch. The retroesophageal right subclavian artery is usually found to be symptomatic in 10-33% of the cases.[1] Various symptoms might be induced if this artery compresses the nearby structures, the trachealesophagus axis. In the case described herein, we also found another anatomic aortic arch variation in association with ARSA which is the bicarotideus trunc.

Case Report

The patient is an 8-month-old girl who was referred to gastroenterology service because of dysphagia and poor weight gain. She was born at term, 39 weeks, by vaginal delivery. The perinatal period was uneventful. Her birth weight was 2300 gr. She was the first child of an Albanian couple without consanguinity. At the age of

one month, she was admitted to pneumology service because of stridor and failure to thrive. According to her mother her daughter had multiple episodes of vomiting and choking. Her blood count was normal and her oxygen saturation was 99%. Chest x-rays showed no abnormality. Due to her swallowing difficulties, temporary feeding was provided via a nasogastric tube as the best solution to nourish her. At that time, pulmonologists thought that the symptoms were due to congenital laryngeal stridor. In this context, the patient was put on omeprazole therapy and was closely followed up. At the age of 8 months, she was admitted again to pneumology service because of the persistence of stridor and swallowing difficulties. Feeding was provided by a permanent nasogastric tube since the age of 4 months. Given the persistent stridor, she received oral prednisolone (1 mg/kg/day) and inhalator steroids for five days. No clinical improvement was noticed during corticosteroid therapy. She was transferred to a gastroenterology service for further procedures. A barium esophagogram showed an esophageal filling defect suggesting an extrinsic compression (Figure 1A, B). Our patient underwent an upper digestive endoscopy and laryngotracheoscopy simultaneously. During esophagoscopy was noticed an extrinsic compression in the posterior part of the esophagus, which was pulsatile and suggested an aberrant artery (Figure 2). Flexible bronchoscopy revealed laryngomalacia with a prolapse of aryepiglottic folds and mucosa. The trachea has no stenosis despite a rotation of 90 degrees anticlockwise because of external compression (Figure 3A, B).

CT-angiography was required, which showed an aberrant right subclavian artery originating from the distal aortic arch compressing the posterior thoracic oesophagus. An additional finding was the presence of a common carotid trunk (the common origin of the carotid arteries). No aneurysmal dilatation indicative of Kommerell's diverticulum was seen (Figure 4A, B). The persistency of the clinical conditions with severe esophageal compression indicated an early surgical repair. At the age of 16 months, she was transferred to the Division of Cardiac Surgery at the University of Verona School of Medicine, Italy. She underwent a successful elective procedure that consisted of surgical ligation and division of the ARSA via a postero-lateral thoracotomy in the IV intercostal space (Figure 5A, B). The aberrant vessel was controlled between two vascular clamps, one applied at the base of the vessel and the other to a distance of approximately 1 cm. The vessel was cut with Pots forceps and the two segments were sutured with 5-0 prolene running suture. After careful control of the hemostasis, the clamps were released and the two segments separated far enough from each other (the proximal segment retracted behind the esophagus) to prevent reattach-

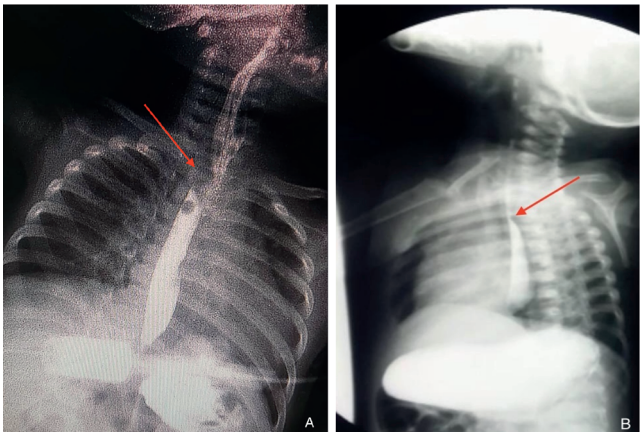

Figure 1. Barium swallow. A) Antero posterior view shows an esophageal filling defect (red arrow) B) Lateral view.

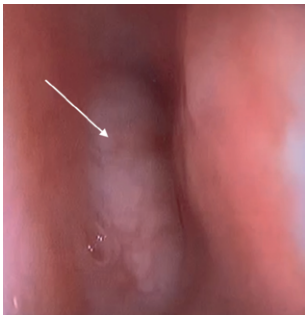

Figure 2. Esophagoscopy shows a bulging mass in the posterior wall of esophagus (white arrow).

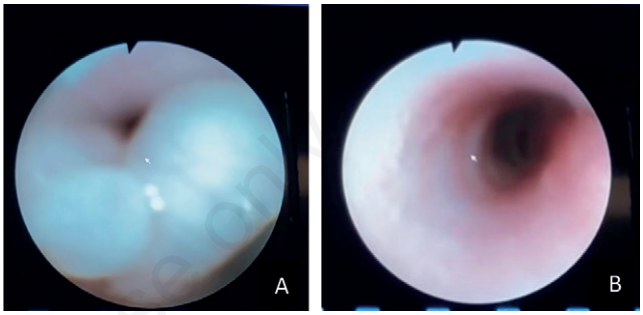

Figure 3. Flexible bronchoscopy showing A) laringomalacia B) tracheomalacia.

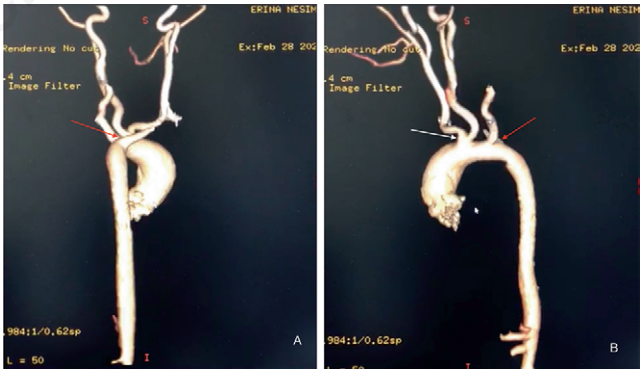

Figure 4. CT angiography A) lateroposterior view showing ARSA (red arrow), B) anterior view showing ARSA (red arrow), truncus bicaroticus (white arrow).

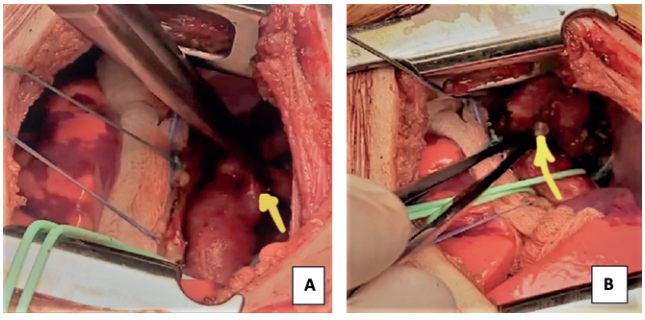

Figure 5. During intervention A. Detection of arteria lusoria B. Stump remnant after resection of arteria lusoria.

ment and thus future problems in the area. Surgery was uneventful and recovery was progressive and fast with total regression of the symptoms. The child was extubated on the first post-operative day (POD) and transferred to the clinical ward on the second POD, no inotropes or specific drugs were needed to manage the post-operative course. Discharge from the hospital occurred in the tenth POD. No complications in terms of right arm function were noted.

After surgery, the dysphagia symptoms disappeared. She is nourished normally and is gaining weight according to her age. Another esophagogram showed normal anatomy of the esophagus.

Discussion

The aberrant right subclavian artery, known as arteria lusoria, is the most common congenital of the aortic arch with an incidence range from 0.4-1.8 % in the general population.[2-4]

ARSA originates as the last branch of the aorta on the left side of the left subclavian artery.

Embryologically, during the fourth and the seventh week of gestation, six pairs of aortic arches are developed. The aberrant origin of the right subclavian artery occurs by the regression of the right fourth aortic arch and proximal right dorsal aorta and the persistence of the seventh intersegmental artery and the distal segment of the dorsal aorta. The retroesophageal course can be explained by the fact that the abnormal subclavian artery is constituted from a portion of the right dorsal aorta.[10]

Due to the embryologic anomaly, ARSA follows this abnormal course behind the esophagus in 80% of cases similar to our case report. But is also described in 13% of cases it passes between the trachea and esophagus and in 5% in a peritracheal course.[5]

Although most of the cases of this condition remain clinically silent, when the symptoms are present, they vary depending on the patient's age. In infants, the trachea is less rigid, so generally, the compression causes respiratory symptoms like stridor, wheezing, and recurrent pneumonia. Several cases of ARSA associated with tracheomalacia have been described in the pediatric population, similar to our report case.[6] This tracheomalacia may be caused by the weakness of the trachea in the early years of life as well as the compression by the aberrant artery.

Meanwhile in adults mostly occurs dysphagia that has been related to increased wall thickness and rigidity of the esophagus or atherosclerotic alteration in the wall of the artery.[8]

According to the literature, significant symptoms occur when there is a common origin of the carotids (truncus bicaroticus) at the front and the aberrant artery at the back, which can cause compression of the trachea and limits the trachea's mobility.[7] An interesting finding in our case remains early presentation and severity of dysphagia and stridor making the diagnostic more than a challenge. Initially, we have to make a differential diagnosis with congenital laryngeal stridor which has a similar presentation: stridor, vomiting, and poor gain weight. Taking into account that stridor and dysphagia were so reluctant to treatment makes us think about a vascular abnormality. Diagnostic keys were barium esophagogram and upper digestive endoscopy.

ARSA is generally diagnosed incidentally based on imaging techniques. Barium esophagogram can often show an accurate compression in the posterior wall of the esophagus, while CT or MRI angiography remains the gold standard not only for the diagnosis but also for the surgery planning as a result of the detailed visualization of the anatomical structure.[9] In our case, CT angiography not only concluded our suspicion about ARSA but revealed another abnormality such as truncus bicaroticus making even rare our finding.

After diagnosis, another challenge was surgery repair that was performed with a left thoracotomy approach. The simplest and "less invasive" strategy was chosen based on the age and fragility of the tiny patient. Surgery consisted of ligation of ARSA and surgical resection of the vessel origin from the arch (Figure 5A, B) to open up space and thus abolish esophagus compression. Mobility and perfusion of the right arm were adequate after the operation and problems were not noted. This is because in small patients retrograde perfusion from the Willis circle and collateral arteries that are present is sufficient to perfuse the divided right subclavian artery.

Ligation and resection were preferred over re-implantation because is simple and less invasive compared to the same operation associated with reimplantation of the ARSA to the right carotid artery.[11] When you manipulate head vessels in small infants there is a concern and risk associated with cerebrovascular events.

Conclusions

Every physician should be aware of the aberrant right subclavian artery and its clinical symptoms in the pediatric population or adults, to recognize and have an early diagnosis. Only an early evaluation might reduce the complication such as retarded physical growth, dysphagia, and recurrent respiratory infections

References

1. Natsis K, Didagelos M, Gkiouliava A, et al. The aberrant right subclavian artery: cadaveric study and literature review. Surg Radiol Anat 2017;39:559–65. Erratum in: Surg Radiol Anat 2017 Sep 14.
2. Kieffer E, Bahnini A, Koskas F. Aberrant subclavian artery: surgical treatment in thirty-three adult patients. J Vasc Surg 1994;19:100e11.
3. Molz G, Burri B. Aberrant subclavian artery (Arteria lusoria): sex differences in the prevalence of various forms of the malformation. Evaluation of 1378 observations. Virchows Archiv A: Pathological Anatomy and Histology 1978;380:303–15.
4. Myers PO, Fasel JHD, Kalangos A, Gailtoud P. Arteria lusoria: Developmental anatomy, clinical, radiological and surgical aspects. Ann Cardiol Angeiol 2010;59:147–54.
5. Natsis K, Didagelos M, Gkiouliava A, et al. The aberrant right subclavian artery: cadaveric study and literal review. Surg Radiol Anat 2017;39:559–65.
6. Pantvaidya GH, Mistry RC, Ghanekar VR, et al. Injury of an aberrant subclavian artery: a rare complication of video assisted thoracoscopic esophagectomy. Ann Thorac Cardiovasc Surg 2005;11:35–7.
7. Janssen M, Baggen MGA, Veen HF, et al. Dysphagia lusoria: clinical aspects, manometric findings, diagnosis, and therapy. Am J Gastroenterol 2000;95:1411–6.
8. Ulger Z, Ozyurek AR, Levent E, et al. Arteria lusoria as a cause of dysphagia. Acta Cardiologica 2004;59:445–7.
9. Karcaaltincaba M, Haliloglu M, Ozkan E, et al. Non-invasive imaging of aberrant right subclavian artery pathologies and aberrant right vertebral artery. Br J Radiol 2009;82:73–8.
10. Moore KL, Persaud TVN. The developing human: clinically oriented embryology. 6th Ed. Philadelphia: WB Saunders Co.; 1998. pp. 384–394.
11. Nelson JS, Hurtado CG, Wearden PD. Surgery for Dysphagia Lusoria in Children. Ann Thoracic Surg 2020;109:e131–3.