Dysphagia, looking further than the usual suspects, a case report

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European Journal of Medical Case Reports

Volume 7(1):01–05 https://doi.org/10.24911/ejmcr/173-1644152189



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ABSTRACT

Background: Vascular ring is a rare diagnosis in adults as the majority is diagnosed in symptomatic infants or young children. Only a few cases are documented in the literature of dysphagia in adults caused by a vascular ring. It is not uncommon for an adult diagnosed with vascular rings to have asthma in his medical history.

Case Presentation: A 61-year-old woman presented with dysphagia with a medical history of slight asthma and left hemithyroidectomy. After extensive workup, it was determined to be the result of a right-sided aortic arch with a left-sided arteriosum ligament, a left aberrant subclavian artery with Kommerell's diverticulum, and a ventral left common carotid artery.

Conclusion: A right-sided aorta with a left-sided arteriosum ligament is a vascular ring and should be recognized as such. Although rare in adults, one should be triggered to think of a vascular ring in patients with dysphagia, especially with mild asthma and reflux complaints.

Keywords: Right-sided aorta, case report, dysphagia, vascular ring, aberrant left subclavian artery.

Received: 06 February 2022
Accepted: 15 October 2022
Type of Article: CASE REPORT
Specialty: Anatomy

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Background

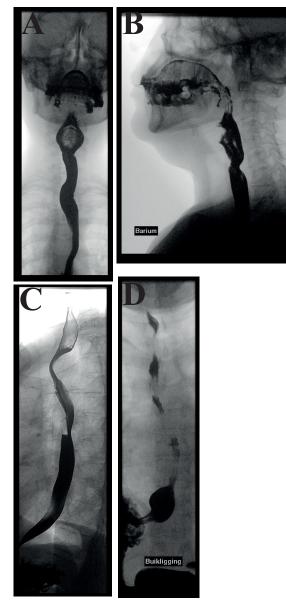
Vascular rings are anatomic variations in which the aortic arch and its branching vessels enclose the trachea and esophagus. The vascular ring is not a common diagnosis in the elderly. They have been well-documented in children to cause digestive signs and symptoms and respiratory problems like obstructive flow or infections. Only a few reports are made of vascular ring symptoms in the elderly, mostly having symptoms of dysphagia. Recognition in adults is difficult and often delayed because sometimes a different diagnosis is thought of.

Case Presentation

A 61-year-old woman underwent a hemithyroidectomy on the left side in 2019 elsewhere. After surgery, she was an outpatient and the internist referred her to surgery in our clinic with complaints of dysphagia. She further had a history of asthma, diabetes, and pre-existing left-sided vocal cord paralysis according to her medical status. The medication she uses is omeprazole and vitamin D. Her complaints were dysphagia and a slight cough. Duration and when complaints arise were not mentioned in the status.

A barium swallow test was performed and the following observations were made: a plump cricopharyngeal muscle without complete relaxation (marginal normal function), a right-sided impression due to goiter, a dorsal impression of a right-sided aorta, and a small hiatus hernia esophagus (Appendix I). Further findings were normal. The test was done with liquids and solids; no stasis, reflux, or aspiration was found during fluoroscopy. Esophageal manometry was performed and showed elevated pressure proximal, at the time thought to be a hypertrophic cricopharyngeal muscle causing her complaints. Not the thyroid or any other diagnosis was thought to be the reason for her complaints. The slight cough was explained as a result of the small HD, not reacting to omeprazole.

The patient was referred to a dedicated/interdisciplinary swallow team for treatment and potentially off-label Botox treatment of the cricopharyngeal muscle. She was referred back to the surgeon as the swallow team did not think the cricopharyngeal muscle was the culprit.



Appendix I. Barium swallow test. (B) Plump cricopharyngeal muscle. (C) Impression of struma. Severe impression dorsal of esophagus by right-sided aortic arch. (D) = fig 4 Small hiatus hernia esophagus

The surgeon ordered a thyroid uptake single-photon emission computed tomography (SPECT-CT) to see if the patient could qualify for radioactive iodine therapy. The following findings were made: right-sided thyroid (estimated 55 g), extension dorsal of the esophagus with slight impression on the esophagus, in this dorsal part of the right thyroid no iodine uptake, almost no impression on the trachea, and known vascular anomaly of the rightsided aorta with compression on the esophagus (Appendix II). With the suggestion of a vascular ring, the patient was referred to a cardiothoracic surgeon for treatment options. See timeline (appendix IV). (See timeline (appendix IV)).

Discussion

The differential diagnosis of dysphagia is extensive, and although sometimes the diagnosis is obvious, in some cases it is difficult to pinpoint the cause, especially, when it is an anatomic cause in an elder patient and if there are multiple anatomical explanations for the dysphagia. Normal work-up of dysphagia include a thorough anamnesis (and elicit symptoms in patient history), physical examination, barium swallow, manometry, and endoscopy. Several questionnaires have been made to aid the interpretation of dysphagia [1].

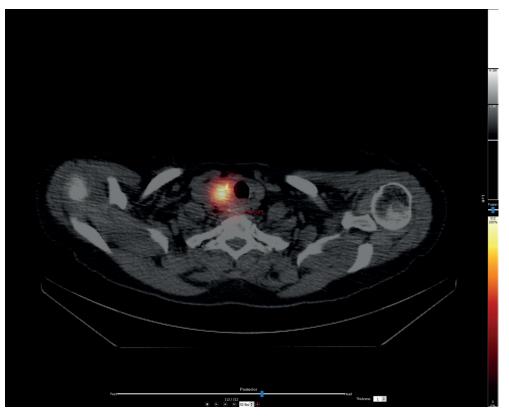
This case presents an uncommon cause not always thought of during work-up. Our patient had a right-sided aortic arch with a left ligament arteriosum (Appendix III).

Symptoms of vascular rings in children are: noisy breathing and barky cough (seal-bark cough), recurrent upper respiratory tract infections, wheezing, dyspnea on exertion, and dysphagia. Some infants can have apnea or even an apparent life-threatening event. Dysphagia is usually a symptom until solid foods are given to the child [2]. Most of the time, a vascular ring is diagnosed in children since it is symptomatic. In adults, it is considered rare and may be misdiagnosed as asthma. This case represented a medical history of asthma, and her reflux did not respond to omeprazole. It is possible that in this case the reflux complaints and asthma are symptoms of vascular ring.

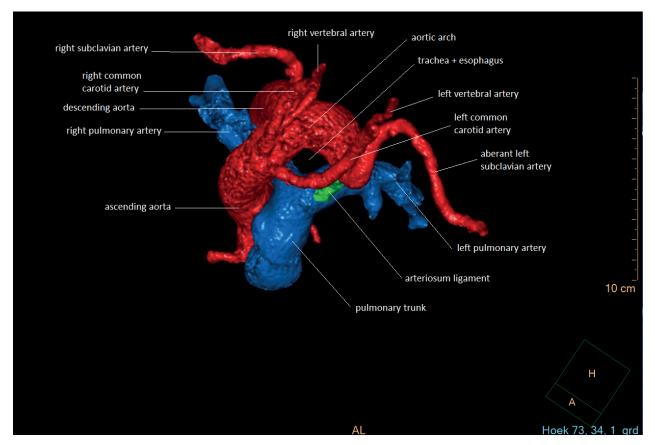
Understanding the embryology of the aorta helps to understand vascular rings. A right-sided aorta is formed when the fourth left branchial arch involutes, resulting in a right-sided arch. Of the right-sided aorta (approximately 0.1% in adults) there are three types of the right-sided aortic arch:

- Type 1: right-sided aortic arch with mirror image branching (59% of all right-sided arches).
- Type 2: right-sided aortic arch with the aberrant left subclavian artery (39.5%).
- Type 3: right-sided aortic arch with isolation of the left subclavian artery (rarest type, 0.8%) [3,4].

In the right aortic arch, a vascular ring is completed with a left-sided ligament arteriosum. It remains unclear why some patients become symptomatic or have slowly progressing symptoms in adulthood. It had been postulated that progressive aortic arch elongation and aortic atherosclerosis are contributing factors [5]. Or maybe the



Appendix II. I-123 SPECT-CT: status after left-sided hemithyroidectomy. Right thyroid 55 g, extension of right thyroid dorsal of esophagus. No uptake in thyroid tissue dorsal from the esophagus, and hardly any compression on the trachea.



Appendix III. (A) Right-sided aortic arch with left-sided ligament arteriosum and Kommerell's diverticulum with aberrant left subclavian artery. (B) The trachea and esophagus pass through the vascular ring.

Timeline				
	Medical History:			
	ASTMA			
	Diabetes			
	Acid related	l complaints	5	
	2019 Left hemithyroidectomy			
Current illness:				
Dysphagia		Jan 2019		
		Jun 2019	Barium swallow test	
work diagnosis:		Oct 2019	Manometry	
- Plump cricopharyngeal muscl	e			
DDx:				
- Goiter				
- right sided aorta				
- hiatus hernia esophagus				
		Feb 2020	Referral to dedicated	
			/interdisciplinary swallow team	
			,,	
Work diagnosis:				
- Goiter				
DDx:				
- right sided aorta				
- hiatus hernia esophagus				
		Dec 2021	Thyroid uptake SPECT CT	
Final diagnosis:				
Vascular ring				
			Referral to cardiothoracic	
			surgeon	

Appendix IV. Timeline.

dilation of Kommerell's diverticulum may be a contributing factor.

Conclusion

The purpose of this case report is to understand and identify relevant clinical anatomy. A right-sided aorta with a left-sided arteriosum ligament (and a ventral left common carotid artery) is a vascular ring and should be recognized as such. Although it is rare in adults, one should be triggered to think of a vascular ring in patients with dysphagia, especially with mild asthma and reflux complaints.

What is new?

Dysphagia in infants and young children based on anatomical variations (such as right-sided aorta variations) is well documented. Only a few reports are made of dysphagia in adults based on a right-sided aorta.

Conflict of interest

The author declares that there is no conflict of interest regarding the publication of this case report.

Funding

None.

Consent for publication

Obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	Patient (gender, age)	61yr, female
2	Final diagnosis	Vascular ring
3	Symptoms Dysphagia (ASTMA and acid related complaints in medical history)	
4	Medications	Omeprazole
5	Clinical procedure	Clinical work-up dysphagia complaint.
6	Specialty	Radiology, Surgery