Asian Journal of Research and Reports in Gastroenterology

Volume 6, Issue 1, Page 98-102, 2023; Article no.AJRRGA.102917

Co-occurrence of the Rare Case of Dysphagia Lusoria with Achalasia

Nahla Ahmed Khalaf [a*] and Asem ElFert [a]

[a] *Tanta University Hospitals, Egypt.*

Authors' contributions

This work was carried out in collaboration between both authors. Both authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/102917

Case Report

Received: 18/05/2023
Accepted: 21/07/2023
Published: 29/07/2023

ABSTRACT

Dysphagia is a subjective feeling of difficulty of swallowing that has many causes. Of them, achalasia in which there is failure of relaxation of lower esophageal sphincter (LES) together with aperistalsis of the esophagus. Dysphagia lusoria a rare case of dysphagia is secondary to an aberrant right subclavian artery (ARSA) originating from left aortic arch compressing the esophagus. We found both conditions in a case presented with dysphagia and weight loss.
Aims: It is important to emphasize the importance of appropriate use of diagnostic modalities and the possibility of co-occurrence of different etiologies of dysphagia.

Keywords: Achalasia; dysphagia lusoria; aberrant right subclavian artery; lower esophageal sphincter.

1. INTRODUCTION

Dysphagia is a common gastrointestinal complaint, which vary in etiology and severity and impose a negative impact on the quality of life of patients [1].

It is usually underreported by clinicians, so it needs standardized evaluation including history, selection of appropriate diagnostic workup, assessment of complications, and providing a road map for the plan of management [2].

**Corresponding author: Email: nahla.khalaf@med.tanta.edu.eg, dr_nohnoh86@yahoo.com;*

"Achalasia is a rare disorder characterized by impaired relaxation of the lower esophageal sphincter (LES) and absent or spastic contractions in the esophageal body. The key pathophysiological mechanism is loss of inhibitory nerve function that probably results from an autoimmune neurodegeneration of the esophageal myenteric plexus" [3].

"Upper GI endoscopy and timed barium esophagogram are the initial investigations to rule out mechanical obstruction. High resolution manometry (HRM) is diagnostic and helps to classify achalasia" [4].

Achalasia can be classified into three subtypes: Type I, there is no esophageal contractility with a dilated, atonic esophagus, Type II, characterized by pan-esophageal pressurization caused by disorganized esophageal neuromuscular activity, and Type III, characterized by premature contractions and segmental pressurization of the distal esophagus [5].

"Dysphagia lusoria (DL) is a rare vascular anomaly identified in a small number of patients being evaluated for dysphagia. It is estimated that in a general population, the prevalence of DL is 0.4%-0.7%, and the incidence of aberrant right subclavian artery (ARSA) is 0.5%-1.8%" [6].

"This may manifest clinically with dysphagia or reflux. However, a low percentage of aberrant arteries result in trachea-esophageal symptoms, so we must exclude co-morbid esophageal motility disorders" [7].

2. CASE PRESENTATION

A 48-year-old Egyptian lady presented with intermittent dysphagia mainly to solids with occasional regurgitation for the last 10 years. Barium meal and EGD were performed at another hospital 5 years ago reporting no abnormal findings. She came to our endoscopy unit with dysphagia mainly to solids and marked weight loss (42 kgs in 7 years). Barium esophagogram revealed indentation of the upper esophagus, with free flow of barium (Fig. 1). Computed tomography (CT) of the chest showed aberrant right subclavian artery, prominent aortic knuckle and dilated lower esophagus (Fig. 2). We performed EGD, the pediatric endoscope passed the upper esophageal stenosis, but failed to pass the pinpoint lower esophageal stenosis (Olympus GIF-160) (Fig. 3). Achalasia was diagnosed as the cause of significant dysphagia and weight loss, and pneumatic dilatation was done with Hercules 18-19-20mm balloon (Cook, USA) (Figs. 4,5). The patient symptoms improved after two sessions of pneumatic balloon dilatation with some dietary modifications.

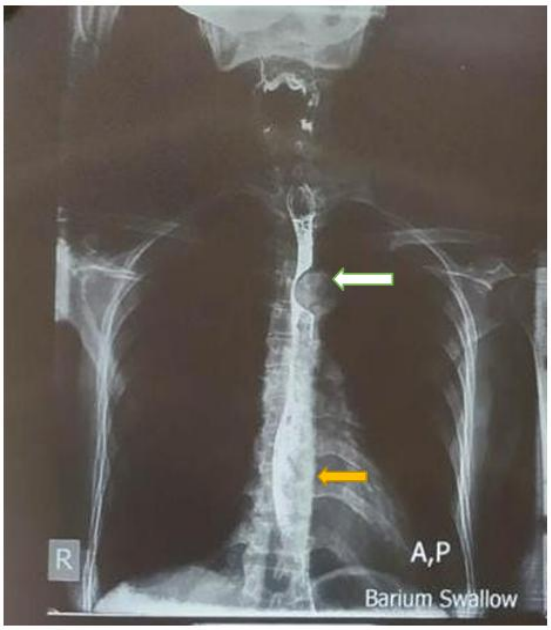

Fig. 1. Barium esophagogram showing indentation of esophagus andprominent aortic knuckle (white arrow) and fusiform dilation of esophageal body with narrow tapering at lower esophagus (yellow arrow)

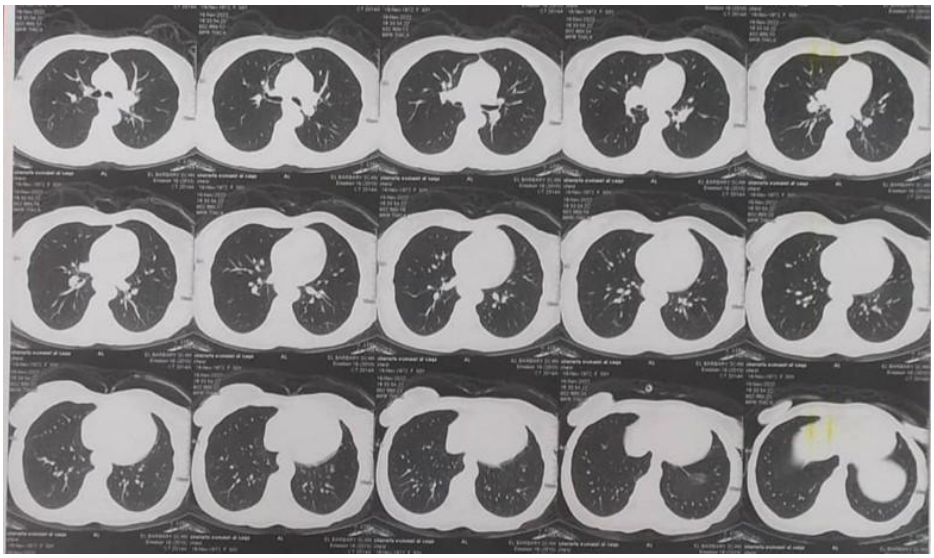

Fig. 2. CT chest showing aberrant right subclavian artery

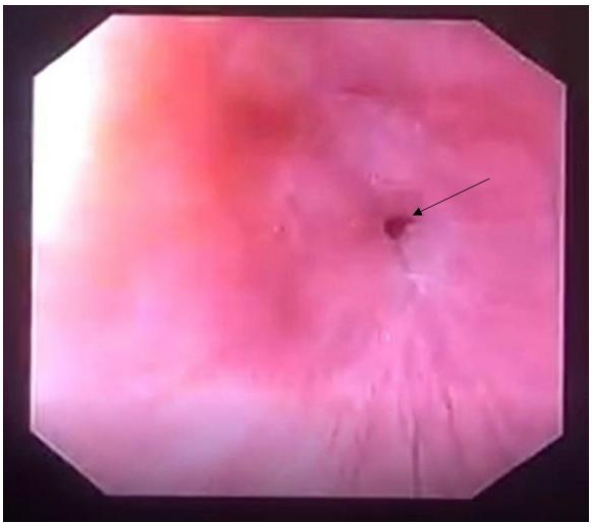

Fig. 3. EGD showing pinpoint GEJ not passing the scope

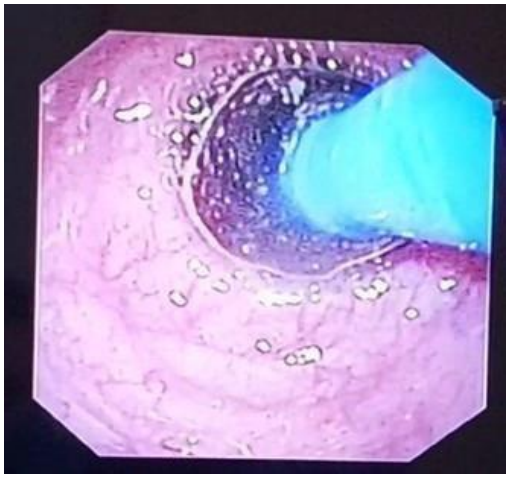

Fig. 4. Balloon dilatation of LES

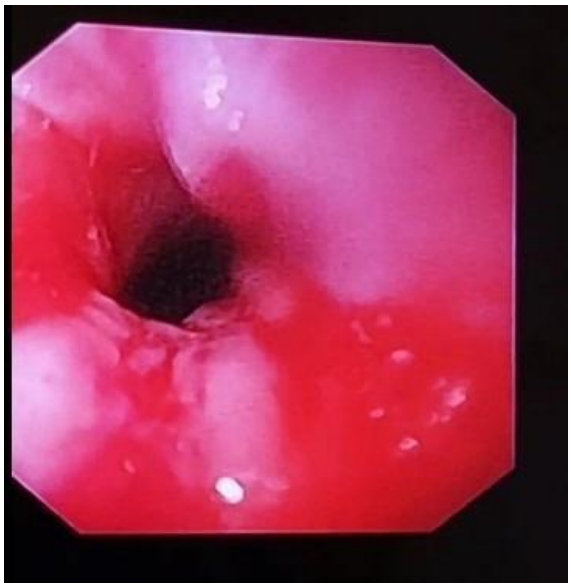

Fig. 5. Dilated LES after balloon dilatation

3. DISCUSSION

We did not find a published association between dysphagia lusoria and achalasia.

“Achalasia is a primary esophageal motility disorder, characterized by progressive degeneration of ganglion cells in the esophageal myenteric plexus. It results in impaired relaxation of LES on swallowing and aperistalsis in the distal smooth muscle segment of the esophagus” [8].

“Patients most commonly present with dysphagia to solids and liquids, regurgitation, and occasional chest pain with or without weight loss, diagnosis is suggested by barium swallow and endoscopy, and confirmed by manometry” [9].

Endoscopy, barium esophagogram and esophageal manometry are complementary in diagnosing achalasia and according to high resolution manometry (HRM) it is graded into three subtypes [10].

All 3 subtypes have impaired esophagogastric junction (EGJ) relaxation, but the distinguishing features are the pattern of esophageal pressurization and contraction; in achalasia type I (second most common; 20%–40% of cases) is characterized by 100% failed peristalsis (aperistalsis) with the absence of panesophageal pressurization to more than 30 mm Hg, achalasia type II (most common; 50%–70% of cases) is characterized by 100% failed peristalsis (aperistalsis) with pan esophageal pressurization to greater than 30 mm Hg, and achalasia type III (least common; 5% of cases) is characterized by spastic contractions because of abnormal lumen obliterating contractions with or without periods of pan esophageal pressurization [11].

‘Treatment modalities available for this purpose include pneumatic dilation, laparoscopic Heller myotomy and recently peroral endoscopic myotomy or POEM” [12].

Most cases are asymptomatic and symptomatic cases present after 45- years of age as non-progressive mechanical dysphagia [13].

Management of this condition depends on severity: mild cases are best managed by patient reassurance and lifestyle and dietary modification while severe cases are best managed with surgery aimed at moving and fixing the aberrant vessel in its appropriate position [14,15]. Our case responded to dietary modification and pneumatic dilatation without need for endovascular intervention. We believe that the magnitude of the problem was due to severe lower esophageal stenosis caused by achalasia, so it improved after pneumatic dilatation, but needed dietary modification to manage the moderate upper esophageal stenosis.

4. CONCLUSION

A high index of suspicion and wise choice of appropriate diagnostic measures and putting in

mind the co-occurrence of different etiologies are the key for proper management of dysphagia.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Mari A, Sweis R. Assessment and management of dysphagia and achalasia. Clin Med (Lond). 2021;21(2):119-123.
2. Le KHN, Low EE, Yadlapati R. Evaluation of esophageal dysphagia in elderly patients. Curr Gastroenterol Rep. 2023;25(7):146-159.
3. Savarino E, Bhatia S, Roman S, Sifrim D, Tack J, Thompson SK, Gyawali CP. Achalasia. Nat Rev Dis Primers. 2022;8(1):28.
4. Mohan Ramchandani, Partha Pal. EMJ Gastroenterol. 2020;9[1]:106-17.
5. Kahrilas PJ, Bredenoord AJ, Fox M, Gyawali CP, Roman S, Smout AJ, Pandolfino JE. International High Resolution Manometry Working Group. The Chicago Classification of esophageal motility disorders, v3.0. Neurogastroenterol Motil. 2015;27(2):160-74.
6. Maier W. Dysphagia lusoria: anomalous origin of the right subclavian artery. Demonstration by computed tomography. Eur J Radiol. 1981;1(3):229-31
7. Coles M, Sharma A. Dysphagia Lusoria: Is the Dysmotility Connection Illusory or Real? Dig Dis Sci. 2020;65(4):942-945.
8. Oude Nijhuis RAB, Zaninotto G, Roman S, Boeckxstaens GE, Fockens P, Langendam MW, Plumb AA, Smout A, Targarona EM, Trukhmanov AS, Weusten B, Bredenoord AJ. European guidelines on achalasia: United European Gastroenterology and European Society of Neurogastroenterology and Motility recommendations. United European Gastroenterol J. 2020;8(1):13-33.
9. Moonen A, Boeckxstaens G. Current diagnosis and management of achalasia. J Clin Gastroenterol. 2014;48(6):484-90.
10. Vaezi MF, Pandolfino JE, Yadlapati RH, Greer KB, Kavitt RT. ACG clinical guidelines: diagnosis and management of achalasia. Am J Gastroenterol. 2020;115(9):1393-1411.
11. Katsumata R, Manabe N, Sakae H, Hamada K, Ayaki M, Murao T, Fujita M, Kamada T, Kawamoto H, Haruma K. Clinical characteristics and manometric findings of esophageal achalasia-a systematic review regarding differences among three subtypes. J Smooth Muscle Res. 2023;59:14-27.
12. Cappell MS, Stavropoulos SN, Friedel D. updated systematic review of achalasia, with a focus on POEM therapy. Dig Dis Sci. 2020;65(1):38-65.
13. Irakleidis F, Kyriakides J, Baker D. Aberrant right subclavian artery - a rare congenital anatomical variation causing dysphagia lusoria. VASA. 2021;50(5):394-397.
14. Dranseika V, Erdil T, Schweiger M, Balmer C, Pretre R, Dave H. Dysphagia, and an aberrant subclavian artery: more than just a coincidence. Interact Cardiovasc Thorac Surg. 2020;31(2):228-231.
15. Tenneti VJD TA, Reddy A. Dysphagia lusoria: a rare cause of adult dysphagia. Cureus. 2022;14(8):e28648.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/102917