An Uncommon Cause of Dysphagia in a 35 Year Old Male

by Carmelo Blanquicett, Terence Dunn, Arjun Nanda, Frederick Weber

Typical causes of intermittent esophageal dysphagia in a young person include eosinophilic esophagitis, esophageal dysmotility and esophageal rings. We report a 35-year-old male with a one year history of intermittent dysphagia to solid foods. After the endoscopic removal of a food bolus, a barium swallow revealed extrinsic compression of the proximal esophagus. Computed tomography angiogram revealed an aberrant right subclavian artery (ARSA) coursing behind the esophagus, suggesting the diagnosis of dysphagia lusoria. Although rare, dysphagia lusoria represents an important consideration in the differential diagnosis of intermittent esophageal dysphagia in a young adult.

INTRODUCTION

In young adults, esophageal dysphagia is most commonly attributed to eosinophilic esophagitis, strictures, motility disorders or neurological injury. Occasionally symptoms can result from extrinsic compression from mediastinal masses, vascular structures or surgical changes. We present the case of a 35-year-old male who presented with complaints of intermittent esophageal dysphagia and was found to have dysphagia lusoria. Although uncommon, it is important to consider this diagnosis when evaluating patients with dysphagia.

CASE REPORT

A 35-year-old Caucasian male was admitted to our institution after he presented to the emergency department with the sensation of food stuck in his chest for two days following the consumption of pork.

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He reported a one-year history of intermittent solid-food dysphagia with episodes typically resolving by "coughing up" and regurgitating the food bolus. He denied choking, aspiration, associated dyspnea, difficulty initiating a swallow or odynophagia. He denied tiring upon chewing or focal weakness. His weight had been unchanged, and he denied any prior tobacco use. He underwent an esophagogastroduodenoscopy (EGD) with esophageal biopsies two months prior to presentation at an outside facility and was placed on empiric proton pump inhibitor (PPI) therapy without clinical benefit. Biopsies did not reveal evidence of eosinophils. His past medical history included chronic tension headaches but he denied a history of atopy or asthma.

At the time of our evaluation, he was handling his oral secretions and had no complaints of dyspnea or choking. His vital signs and physical examination, including a comprehensive cardiovascular and head, eyes, ears, nose and throat (HEENT) exam were unremarkable. Laboratory studies were within normal limits. After glucagon was administered in the emergency room

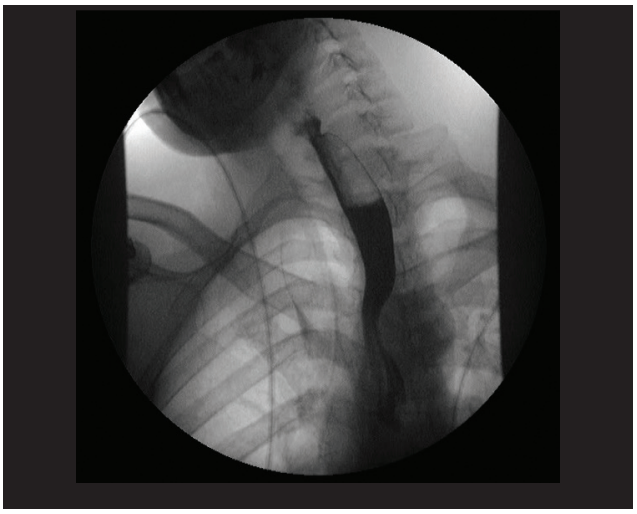

Figure 1. Barium swallow: revealed smooth narrowing at the pharyngoesophageal junction consistent with cricopharyngeal bar as well as extrinsic compression of the mid-esophagus from vascular structure.

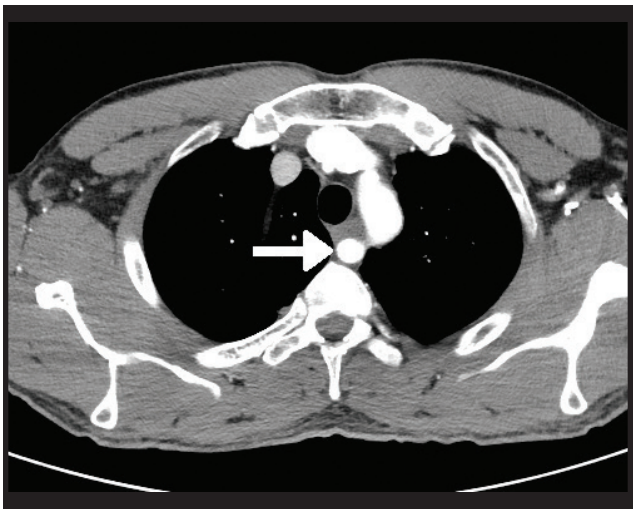

Figure 2. Computed tomography chest: demonstrates an aberrant, right subclavian artery (arrow) coursing posteriorly to the esophagus, causing mild compression of such structure. No aneurysm of the subclavian artery can be appreciated, with the remainder of the mediastinum being unremarkable.

without clinical improvement, a large food bolus was successfully removed endoscopically from the proximal esophagus. The underlying mucosa appeared friable but no strictures or rings were visualized.

Post-endoscopy esophagram revealed a smooth narrowing at the pharyngoesophageal junction consistent with a cricopharyngeal bar. More significantly, there was extrinsic compression of the proximal esophagus distally (Figure 1). A computed tomography (CT) angiography of the chest demonstrated an aberrant right subclavian artery (ARSA) passing posteriorly to the esophagus resulting in compression of the posterior aspect of the thoracic esophagus (Figure 2). These findings, in conjunction with the patient's history, suggested the diagnosis of dysphagia lusoria. The patient was referred to vascular surgery clinic for consideration of surgical correction. A reconstruction of the CT images was performed as shown in Figure 3.

DISCUSSION

Dysphagia lusoria results from a congenital abnormality in the development of the aortic arch and its branches causing extrinsic compression of the esophagus. In the majority of cases, the causative vessel is an ARSA originating from a left-sided aortic arch, but persistent right-sided aortic arch with aberrant left subclavian artery has also been described.[1] The ARSA originates from the proximal portion of the descending thoracic

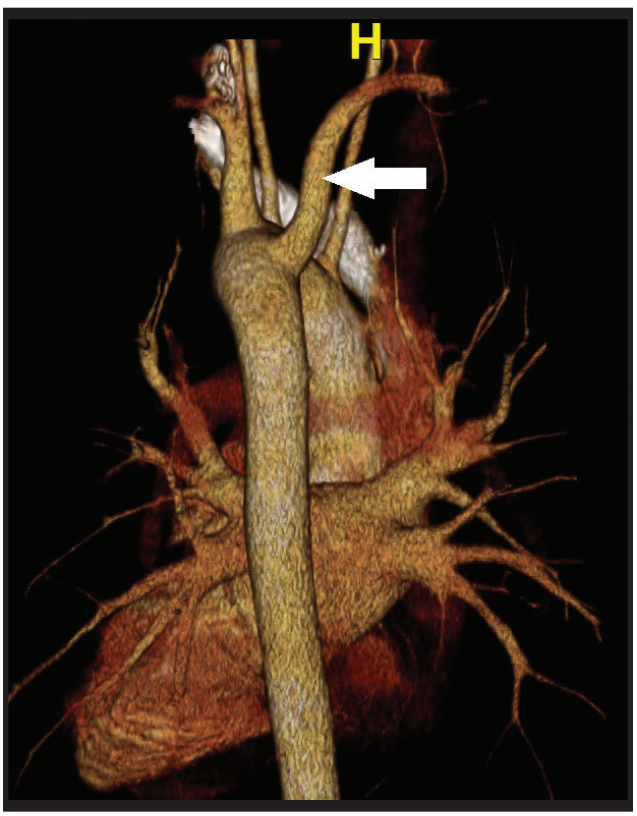

Figure 1. Barium swallow: revealed smooth narrowing at the pharyngoesophageal junction consistent with cricopharyngeal bar as well as extrinsic compression of the mid-esophagus from vascular structure.

aorta with three possible anatomic positions: posterior to the esophagus (80%), between the esophagus and trachea (15%) and anterior to the trachea (5%).[2] This abnormal course led to the term lusorian artery from the Latin lusus naturae or "freak of nature". The prevalence of a lusorian artery has been estimated at 0.4% to 0.7% among the general population.[1] Patients with Down Syndrome commonly have vascular anomalies, and the incidence of ARSA in these patients has be reported in up to 39%.[3] Based on retrospective data, only 30-40% of individuals with ARSA develop symptoms of dysphagia in their lifetime.[1] Without intervention, progressive worsening of symptoms may occur via proposed mechanisms such as age-related loss of esophageal or arterial compliance and aortic elongation. [2]

Depending on the severity of the symptoms, the approach to management varies. In mild cases, behavioral modification strategies, including smaller bites with more thorough chewing, can be adopted.[4] Adjunctive PPI therapy may also improve symptoms. In more severe cases, a surgical approach is warranted. Options include open surgical repair with ligation of the aberrant subclavian artery and anastomosis to the ipsilateral carotid artery or a combined surgical and endovascular (hybrid) approach using stenting as well as carotid-to-subclavian bypass grafting.[5] High success rates, with complete resolution of symptoms, have been reported in patients who underwent surgery.[5,6] We recommended our patient undergo surgical evaluation, given his history of recurrent dysphagia with impaction, in order to address the degree to which his symptoms had progressed.

In summary, dysphagia lusoria represents a rare clinical manifestation of a somewhat uncommon vascular anomaly and should be considered in the evaluation of intermittent esophageal dysphagia in a young adult.

References

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Answers to this month's crossword puzzle:

1 C	R	2 I	S	3 P	R	■	4 S	5 S	B	6 E	■	7 E	N	8 D
O	■	S	■	E	■	9 P	E	T	■	D	■	N	■	E
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30 O	■	■	■	N	■	I	■	■	■	T	■	31 V	■	O
32 L	A	33 B	■	S	■	34 A	S	35 P	L	E	N	I	36 C	■
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39 F	40 A	R	N	E	S	41 O	I	D	X	■	42 S	C	A	N
43 I	D	S	■	C	■	R	■	A	■	44 I	■	U	■	U
45 N	D	A	■	46 T	A	B	U	L	A	T	E	S	■	S