Failure to Thrive and Severe Malnutrition due to a Jest of Nature: Dysphagia Lusoria

Kevin K Yu1; Sruthi K Subramanian2; Asmeen Bhatt2

1University of Texas Health Sciences Center Houston, McGovern Medical School, Department of Internal Medicine, USA.
2University of Texas Health Sciences Center Houston, McGovern Medical School, Division of Gastroenterology, Hepatology, and Nutrition, USA.

Abstract

Dysphagia Lusoria is a rare condition in which dysphagia results from external vascular compression of the esophagus. We describe a 69-year-old man with longstanding intermittent solid-food dysphagia with failure to thrive who was found to have an aberrant right subclavian artery resulting in dysphagia lusoria. This caused a mass effect upon the posterior esophagus resulting in dilatation of the mid and upper esophagus. A diagnosis of dysphagia lusoria was made and surgery was consulted for surgical correction and total parental nutrition started. Definitive diagnosis of dysphagia lusoria involves imaging modalities such as barium esophagram or a CT/MRI. Upper endoscopy is often normal though a pulsatile extrinsic mass can sometimes be seen. There are no pathognomonic manometric findings. Given the rarity of the condition, a high index of suspicion is often required for diagnosis. Definitive management of patients with dysphagia lusoria often involves surgical correction of the anatomical aberrancy. In patients who decline to undergo surgical correction, have mild-moderate symptoms, or are poor surgical candidates, eating slower, chewing well, taking smaller bites, and taking PPIs could be effective therapy options.

Keywords: Dysphagia lusoria; Esophageal dysphagia; Extrinsic compression; Aberrant congenital vascular development.

Introduction

Dysphagia Lusoria is a rare condition in which dysphagia results from external vascular compression of the esophagus. Named after lusus naturae, jest of nature, dysphagia lusoria was first documented in 1794 [1]. It is most commonly due to an aberrant retroesophageal right subclavian artery that affects around 0.5-2% of the population. Here, we describe a case of longstanding dysphagia lusoria that resulted in severe malnutrition and failure to thrive.

Case Description

A 69-year-old man with HIV on antiretroviral therapy with normal CD4 count and undetectable viral load, chronic iron deficiency anemia, gastroesophageal reflux disease, chronic kidney disease, and malnutrition with associated unintentional weight loss presented for sepsis secondary to pyelonephritis. During his hospitalization, he was evaluated for solid-food dysphagia and underwent Fiberoptic Endoscopic Evaluation of Swallowing (FEES). Patient was found to have normal pharyngeal phase of swallowing but with substantial backflow from the upper esophageal sphincter resulting in significant aspiration. Gastroenterology was consulted for dysphagia evaluation. Patient denied odynophagia but reported longstanding intermittent solid-food dysphagia. He had undergone esophagogastroduodenoscopies previously with evidence of reflux esophagitis and gastritis and was on proton pump inhibitors. Repeat endoscopic evaluation was attempted for evaluation of intrinsic disease 3 hours after the FEES study, during which the patient swallowed 2 oz of water and 3 tablespoons of applesauce colored with green food coloring. The swallowed FEES material was found throughout the length of the esophagus and food particles were seen in the mid-esophagus, obstructing the view of the scope preventing safe advancement. The procedure was terminated for patient safety. A CT chest was obtained post-procedure to rule-out extrinsic compression and showed an aberrant right subclavian artery passing behind the upper intrathoracic esophagus (Figure 1). This caused a focal mass effect upon the posterior esophagus resulting in dilatation of the mid- and upper esophagus above the level of the aberrant right subclavian artery (Fig 2A-D). A diagnosis of dysphagia lusoria was established and cardiothoracic surgery was consulted for surgical correction and total parental nutrition started for nutritional augmentation.
Discussion

The pathogenesis of dysphagia lusoria is established early during embryonic development due to a sporadic abnormal involution of the embryonic fourth aortic arch [2]. Rarely, an aberrant left subclavian artery can also result in a similar clinical entity [3]. Patients are often asymptomatic; however, intermittent dysphagia can develop as patients age.

Definitive diagnosis often involves imaging modalities such as barium esophagram or a CT/MRI [2]. Upper endoscopy is often normal though a pulsatile extrinsic mass can sometimes be seen. While patients may have abnormal findings on esophageal manometry, there are no pathognomonic manometric findings [4]. Given the rarity of the condition, a high index of suspicion is often required for diagnosis.

Definitive management of patients with dysphagia lusoria often involves surgical correction of the anatomical aberrancy [2, 4]. In patient’s who decline to undergo surgical correction, have mild-moderate symptoms, or are poor surgical candidates due to co-morbidities, lifestyle modifications such as avoiding exacerbating foods, eating slower, chewing well, taking smaller bites, sipping liquids, and taking proton pump inhibitors could be effective therapy options.