Dysphagia Lusoria - A Rare Cause of Prolonged Dysphagia

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SUMMARY

A 64-year-old man presented with prolonged history of intermittent dysphagia with sensation of food sticking at his upper chest. Physical examination was unremarkable, and an upper endoscopy did not reveal the underlying cause. On computed tomography scan of thorax, an aberrant right subclavian artery was seen coursing posterior to the esophagus resulting in external compression, which is a typical radiological feature of Dysphagia Lusoria. The pathophysiology, clinical features, imaging features and updated treatment modalities of this rare disease are discussed.

KEY WORDS: Dysphagia, dysphagia lusoria, aberrant subclavian artery

INTRODUCTION

Dysphagia lusoria is a rare diagnosis to describe association of dysphagia with vascular compression of the esophagus. The term was first introduced in 1761 by David Bayford as "dysphagia lusus naturae" (meaning "freak of nature" in Latin) when he discovered an aberrant right subclavian artery compressing esophagus in an emaciated female patient who died of long-standing dysphagia.¹

CASE REPORT

A 64-year-old man presented with two-year history of intermittent dysphagia, worse with solid food and described as postprandial chest discomfort with sensation of food sticking at mid-upper chest. He had no odynophagia, regurgitation or weight loss. No other history suggestive of infective or drug-related cause of dysphagia. Physical examination was unremarkable. Initial investigations including full blood count and chest radiograph were within normal limits. Due to the persistent symptom, an upper endoscopy was done and revealed only mild antral gastritis. He subsequently underwent barium swallow study (Figure 1) and computed tomography (CT) scan of thorax (Figure 2). The left oblique (Fig. 1a) and frontal images (Fig. 1b) of barium swallow study showed smooth extrinsic indentation along posterior esophageal wall (white arrow). This is seen just above aortic arch level (black star). Axial images from contrast enhanced CT scan (Fig. 2) showed aberrant right subclavian artery (thick arrow), arising from distal aortic arch and coursing posterior to the esophagus (thin arrow). A

diagnosis of dysphagia lusoria was made. Owing to moderate symptoms that the patient has without significant impact on his nutritional status, he was managed conservatively with dietary modifications, and started on proton pump inhibitor and prokinetic agent. He reported symptomatic improvement since then.

DISCUSSION

From the published literature, the prevalence of dysphagia lusoria in the general population is estimated at 0.4% to 0.7%, with majority of lesions being right subclavian artery originating from the left-sided aortic arch.² This is thought to be resulted from an embryologic abnormality from involution of the fourth vascular arch along right dorsal aorta, with persistent 7th intersegmental artery. Interestingly, this anomaly is often asymptomatic, and only 30% to 40% of patients may develop symptoms such as solid bolus dysphagia during young adulthood, with or without requiritation or thoracic pain, and rarely Horner syndrome. Late-onset presentation in elderly patient, as in our case, is uncommon. The exact mechanism behind this is unclear, but a few explanations have been proposed such as age-related esophageal or arterial stiffening, aortic elongation or progressive aneurysmal dilatation of the aberrant artery.³

Barium swallow study remains an excellent tool for diagnosing this rare condition. The vascular lesion can be subsequently defined by CT or magnetic resonance imaging of chest with vascular reconstruction. Upper endoscopy is usually normal, however it was often done to exclude malignant lesion in the case of persistent dysphagia. Esophageal manometry frequently reveals nonspecific findings and is not helpful in diagnosis.^{2,3} The management of patients with dysphagia lusoria mainly depends on the severity of symptoms. For mild to moderate symptoms without significant impact on nutritional status, this condition can be managed conservatively with dietary modification through chewing well and eating slower in smaller bites. Medical treatment with proton pump inhibitor, with or without prokinetic drug, has been used to improve symptoms in some patients. For patients with severe symptoms not amenable to medical strategies, surgical repair and reconstruction of the aberrant vessel should be considered. Older method such as endoscopic dilation of esophageal narrowing may only result in temporary relief and its role is primarily palliative in patients who are poor

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Fig. 1: The left oblique (Fig. 1a) and frontal images (Fig. 1b) of barium swallow study showed smooth extrinsic indentation along posterior esophageal wall (white arrow). This is seen just above aortic arch level (black star).



Fig. 2: Axial images from contrast enhanced CT scan (Fig. 2) showed aberrant right subclavian artery (thick arrow), arising from distal aortic arch and coursing posterior to the esophagus (thin arrow).

surgical candidates.³ A variety of surgical approaches have been proposed in the literature with different vascular reconstruction methods. More recently, endovascular approach is emerging as promising treatment. Novel methods via mediastinoscopic or supraclavicular approach have been described with good short term outcomes^{4,5}.

In summary, our case describes a late-onset presentation of dysphagia lusoria in an elderly man, which is a rare but important diagnosis and should be considered in cases with prolonged dysphagia, which can be overlooked with upper endoscopy. Barium study with confirmatory CT or MRI imaging of vascular lesion remain useful diagnostic tools, with subsequent medical or surgical management depending on severity of symptoms.

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