A CASE REPORT OF UNUSUAL ANATOMICAL ANOMALY CAUSING:
“DYSPHAGIA LUSORIA”

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ABSTRACT

Intermittent esophageal compression by an aberrant right subclavian retropharyngeal artery (RSRA) is a rare cause of dysphagia, termed as “Dysphagia Lusoria”. This is a congenital anomaly with the right subclavian artery (lusorian artery), originating from the dorsal part of the aortic arch and coursing through the mediastinum between the esophagus and the vertebral column.

In this article, we report a case of a patient with atypical chest pain and chronic dysphagia resulting from this condition, and further discuss its clinical features and options of management.

Keywords: Dysphagia, aberrant right subclavian artery, atypical chest pain, aortic anomalies, barium swallow.
INTRODUCTION

David Bayford, in 1794 first introduced the term “lusus nature” translated from Latin as “difficulty in swallowing, freak of nature”, to refer to an extraordinary disposition of the right subclavian artery (lusorian artery) causing an esophageal obstruction, which was accidentally discovered, after the necropsy of a 62-year old lady, suffering from chronic dysphagia [1]. The aberrant right subclavian retropharyngeal artery (RSRA) is the most common intra-thoracic embryologic anomaly of the aortic arch, with an incidence ranging from 0.4% to 2% [2]. People with this anomaly are generally asymptomatic; however, some may present with dysphagia. In this case report, we describe a lady with chronic history of atypical chest pain and dysphagia resulting from Dysphagia Lusoria, and further discuss its clinical features, investigations and treatment options of this rare entity.

CASE REPORT

A 71-year-old woman presented with prolonged history of atypical chest pain, intermittent dysphagia and sensation of globus hystericus without presenting weight loss. Her initial presenting symptom was discontinuous dysphagia to solids, which worsened and became more progressive in nature. Her past medical history includes hypertension, dyslipidemia, and chronic obstructive pulmonary disease.

On examination, her vitals were normal, was not cachectic and was not jaundiced. Initial investigations, including full blood count, electrolytes, renal function tests, liver function tests and chest films were within normal limits. Her neurological assessment was also unremarkable.

Due to the persistent symptom, an upper endoscopy was performed, which revealed chronic antral gastritis and hiatus hernia with no any intraluminal lesions or strictures. She subsequently underwent barium swallow study, which revealed smooth extrinsic indentation at the proximal third of the esophagus, situated superior to the arch of the aorta at approximately the level of T4 and T5 vertebrae (Figure 1), suggestive of an anomalous right subclavian artery.
Figure 1: A barium-swallow examination shows posterior oblique indentation of the proximal esophagus.

A contrast enhanced computed tomography (CECT) of the chest was performed to further delineate the aorta and its branches, which showed a collapsed esophagus enclosed between the trachea on the right, carotid arteries anteriorly, and the aberrant right subclavian artery posteriorly (Figure 2 and 3). Based on the above findings, a diagnosis of dysphagia lusoria was made. Since the patient’s symptoms were mild and intermittent without significant effect on her nutritional status, she was advised to undergo some dietary modifications to include more semisolid foods and was also prescribed proton pump inhibitors (PPIs) along with prokinetics to help her relieve her reflux symptoms from hiatus hernia. She was counseled for the possibility of surgical treatment in the event of intractable symptoms should medical therapy and lifestyle modification fails along with indepth discussion on risks and benefits of such surgical interventions.

Figure 2: Contrast enhanced CT shows the aortic arch with an aberrant subclavian artery and compressed esophagus.
Figure 3: Coronal contrast enhanced CECT shows the aortic arch with an aberrant subclavian artery.

On her subsequent follow up, the patient experienced a noticeable improvement on her symptoms and is currently still undergoing conservative management with regular follow up in gastroenterology outpatient clinic.

DISCUSSION

The aberrant right subclavian artery (ARSA), also known as Arteria Lusoria (AL), is the most common intra-thoracic embryologic anomalies involving main arteries, with an incidence of 0.4% to 2% [2]. An indepth embryologic knowledge of the aorta and its vessels is pivotal in order to understand the pathogenesis of dysphagia lusoria. Embryologically, interruption of the right arch between the right common carotid artery and right subclavian artery results in a left aortic arch with aberrant right subclavian artery [3]. This vessel arises as the last great vessel of the aortic arch, from the dorsal margin of the aorta, and steers towards the right arm, crossing the middle line of the body and usually passing behind the esophagus. According to the Adachi-Williams' classification, four basic types of anomalous patterns of the ARSA ramification, exists (Figure 4).
Figure 4: Basic anomalous patterns of the aberrant right subclavian artery according to Adachi-Williams’ classification.

- **Type G-1**: The right subclavian artery arises from the distal position of the aortic arch as its last branch; there are no abnormalities of the other main branches (common right and left carotid arteries and the left subclavian artery).
- **Type CG-1**: the right subclavian artery is anomalous (as in type G) and the left vertebral artery originates itself directly from the aortic arch.
- **Type H-1**: the subclavian artery is anomalous (as in type G) and there is a unique trunk, named bicarotidic trunk, from which both the common right and left carotid arteries arises.
- **Type N-1**: there is a right aortic arch and the left subclavian artery origin succeeds both carotid arteries and the right subclavian artery.

Our patient has an ARSA referring to type G of Adachi-Williams’ classification.

A condition called “dysphagia lusoria” may be produced if the esophagus is compressed by the artery. This abnormality is usually asymptomatic (90% to 93%) and most of the times is an incidental finding at necropsy [4, 5]. In children, the most common presenting history would be respiratory problems. It is postulated that in children, the trachea is soft and flaccid contributing to airway compression with associated lower tract infections. In adulthood, patients become symptomatic in the later decades of life, may be due to...
increased rigidity of the esophagus and arterial thickening due to atherosclerosis as age progresses [6]. In our case, we suppose that dysphagia resulted from atherosclerosis, which made the artery wall stiffer and then causing compression on the esophagus.

Endoscopic findings of ARSA are usually normal but sometimes a pulsatile impression may be seen [7]. Barium-contrast swallow studies have been recognized as an excellent tool for screening this condition [6]. In this case, esophagogram may show a characteristic diagonal compression defect at the level of the 3rd and 4th vertebrae. When evaluating patients with dysphagia, the cause of this pathological appearance with fluoroscopy, cross sectional imaging, such as computed tomography (CT) and magnetic resonance (MR), particularly with vascular reconstruction can be analyzed better, as it shows the vascular lesions and the relationship of various mediastinal vessels and structures [2, 3].

Treatment options for ARSA mainly depend on the severity of symptoms and presence of aneurysm, and the impact on the ability of the patient to maintain their weight and nutrition [8]. Janssen and colleagues reported three out of six patients with dysphagia lusoria who became free of symptoms after dietary changes or acid inhibition or promotility agents, which is similar with our patient’s condition [6]. The dietary and lifestyle modifications include reducing exacerbating foods, eating slowly, adequate chewing, taking smaller bites and more liquids [7]. For patients with severe symptoms, who remain symptomatic despite conservative measures, surgical repair and reconstruction of the aberrant vessel should be considered [7, 8].

The main goal of the surgical repair is to remove the aberrant vessel causing the symptoms and restoring the circulation to the right upper limb. The surgical approach depends on the vascular abnormality. In a patient with a normal aortic arch without aneurysmal formation of the proximal aberrant right subclavian artery, a supraclavicular approach alone is suitable [9]. A combined cervical and thoracic approach, via a median sternotomy or lateral thoracotomy, would be suitable for patients with other associated lesions [10]. The perioperative mortality rate in the group of patients with aneurysmal disease of the lusorian artery or an aneurysm at the aortic origin of the lusorian artery was 23.5% [10].

Endovascular treatment of peripheral arteries or aneurysm of the thoracic or abdominal aorta has become increasingly popular as it seems safe and effective alternative for elective treatment [11].

CONCLUSION

In conclusion, our case describes a late-onset presentation of “dysphagia lusoria” in an elderly woman, which is a rare entity and needs to be considered in patients with unexplained chronic dysphagia with normal upper gastrointestinal endoscopy. Dynamic oral contrast swallow studies with confirmatory CT or MRI imaging is an investigation of choice, with subsequent medical or surgical management relating to the severity of symptoms.
REFERENCES