Dysphagia Lusoria: An Uncommon Cause of Dysphagia

Digital Signatures

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Abstract

Dysphagia is a relatively common and increasingly prevalent clinical problem, with prevalence of nearly 22% in the adult primary care population and of 13.5% in the general population. We report a case of an uncommon type of dysphagia, due to esophagus compression by an abnormal right subclavian retroesophageal artery. This condition was first described by Bayford in 1794 as lusus naturae, meaning a freak or jest of nature, the so called “dysphagia lusoria”.

Keywords: Dysphagia; Arteria lusoria; Aberrant right subclavian artery; Dysphagia lusoria; Esophagus compression

Introduction

The Aberrant Right Subclavian Artery (ARSA), also known as “Arteria Lusoria” (AL), is the most common intra-thoracic embrylogic anomalies involving main arteries, with an incidence of 0.4% to 2% [1]. However, it is generally asymptomatic.

In less than 10% of patients, it may manifest with respiratory symptoms or dysphagia. The formers are more common in childhood, whereas the latter results to be the most frequent (91%) in elderly people.

This difference seems to be related to the absence of tracheal rigidity in children and the development, with the aging, of physiologic and anatomic changes such as increased stiffness of esophageal and vessel walls [2].

Dysphagia itself could determine serious problems related to malnutrition, therefore, it is important to recognize this condition as well as to understand the causes, to exclude possible mechanic obstructions due to tumor masses.

Case Report

A 73-year-old male presented with prolonged history of intermittent dysphagia. The initial presenting symptom was discontinuous dysphagia to solids, which worsened and became more progressive in nature.

In addition, the patient reported a subjective sensation of something blocked in his left chest. Physical examination was unremarkable. Initial investigations, including full blood count and chest plain film, were within normal limits.

Due to the persistent symptom, an upper endoscopy was performed and revealed only a mild antral gastritis. He subsequently underwent oral contrast swallow study, which showed smooth extrinsic indentation along posterior esophageal wall at the level of the aortic arch (Figure 1).

A contrast enhanced Computed Tomography (CT) of the chest was then acquired and showed a collapsed esophagus enclosed between the trachea on the right, the carotid arteries anteriorly, and the aberrant right subclavian artery posteriorly (Figure 2 and 3).

Therefore, a diagnosis of dysphagia lusoria was made. Since the patient's symptoms were mild and intermittent, without significant effect on his nutritional status, he was managed conservatively with dietary modifications and medical therapy with proton pump inhibitor and prokinetic agent.

The patient experienced a noticeable improvement of his symptoms.

Figure 1: Frontal esophagram shows an impression (black arrow) on the left side of the esophagus (black star) at the level of the aortic arch, caused by the aberrant right subclavian artery.
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% [1]. Embryologically, a left aortic arch with aberrant right subclavian artery results from the interruption of the right arch between the right common carotid artery and 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, there are four basic types of anomalous patterns of the ARSA ramification (Figure 4) [4]:

- The right subclavian artery arises from the distal portion 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 G-1);
- The right subclavian artery is anomalous (as in type G) and the left vertebral artery originates itself directly from the aortic arch (Type CG-1);
- The right 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 arise (Type H-1);
- There is a right aortic arch and the left subclavian artery origin succeeds both corotid arteries and the right subclavian artery (Type N-1).

The case presented refers to type G of Adachi’s classification.

If the artery compresses the esophagus, it may produce a condition called “dysphagia lusoria” [1,5]. The term “dysphagia lusoria” was first used by Bayford in 1794 to describe a type of dysphagia secondary to a right subclavian retroesophageal artery (aberrant) [4]. This abnormality is usually asymptomatic (90% to 93%) and most of the times is an incidental finding at necropsy [4,5]. Rarely, this malformation has been responsible for dysphagia, as in our case [1]. It is unknown why most patients with ARSA presenting with dysphagia are middle aged or older. According to the current literature this could
be related to many possible co-existing conditions: increased rigidity of trachea and or of AL because of atherosclerotic changes, which both lead to easy compression of esophagus, or aneurysm formation in the presence of Kommerell's diverticulum, elongation of the aorta and the coexistence of an ARSA with a truncus bicaroticus or the presence of a close origin of carotid arteries from the aorta, which limits the anterior displacement of trachea and esophagus [2,5]. In our case, we suppose that dysphagia resulted from atherosclerosis, which made the artery wall stiffer and then causing compression on the esophagus.

When evaluating patients with dysphagia, the primary and preferred imaging modality is fluoroscopy [6]. Actually, contrast swallow studies have been recognized for diagnostic screening of dysphagia lusoria [7]. In this case, esophagogram may show an indentation on the postero-lateral wall of the esophagus at the level and above the aortic arch. To better analyze the causes of this pathological appearance at fluoroscopy, cross-sectional imaging, such as CT and Magnetic Resonance (MR), particularly with vascular reconstruction, will be useful. In fact, they show the vascular lesion and the relationship of the various mediastinal vessels and structures without the need of conventional catheter angiography [1,3]. This latter is not routinely required and may be useful for exact demonstration of vascular anatomy when a surgery therapy is planned [2,8].

In addition they aid to exclude other possible causes of extrinsic compression, such as tumor masses involving lung or mediastinal adenopathies [6].

Among more invasive diagnostic procedure, upper endoscopy usually shows no significant signs and it is performed to exclude malignant lesion in case of persistent dysphagia. Meanwhile, esophageal manometry frequently reveals nonspecific findings and is unhelpful for the diagnosis [2,6,7,9]. In the case we presented, at fluoroscopy there was a quite suspicious narrowing on the posterior-left side of the esophagus that was confirmed at CT scan, which was necessary to exclude the presence of malignant causes of ab-extrinsic compression.

The management of patients with “dysphagia lusoria” depends on the degree of symptoms and the impact on the ability of the patients to maintain their weight and nutrition [9]. If there is insignificant impact on the nutritional status, in case of mild to moderate symptoms, this condition can be managed conservatively with dietary modification including chewing well and eating slower in smaller bites. Otherwise, medical treatment with proton pump inhibitor, with or without prokinetic drug, has been used to improve symptoms, as in our case. At last, for patients with severe symptoms, which not benefit from medical strategies, surgical repair and reconstruction of the aberrant vessel should be considered [2,9].

In conclusion, 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. The presence of the ARSA should be taken into consideration to distinguish from other causes of dysphagia. In our case the hypothesis that clinically overt artery lusoria was due to atherosclerotic wall and vessel stiffness supported by older age of symptomatic patients.

Dynamic oral contrast swallow studies with confirmatory CT or MRI imaging of vascular lesion remain useful diagnostic imaging tools, with subsequent medical or surgical management relating to the severity of symptoms.

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References