Unusual symptomatic late onset presentation of aberrant right subclavian artery: report of two cases and short literature review

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Abstract

Aberrant subclavian arteries are congenital vascular anomalies that usually do not cause any symptoms. When symptomatic they are considered as a rare cause of dysphagia. This presentation is known as dysphagia lusoria. They are diagnosed by barium swallow or contrast-enhanced computed tomography, although it may be an incidental finding. Management varies from life modifications and drug therapy to surgical intervention. We report two cases of the unusual form of late onset symptomatic presentation because of the presence of aberrant right subclavian artery. Main symptom was chest pain without dysphagia. Due to age and medical comorbidities both patients were managed conservatively.

Introduction

Dysphagia lusoria is a term used to describe dysphagia typically caused by an aberrant (retroesophageal) right subclavian artery (ARSA) and was introduced in 1794 by Bayford [1,2]. Dysphagia is a Greek word meaning “difficulty to eat”, however it is used to describe a situation when the food cannot be swallowed. Lusoria comes from the Latin “lusus naturae” meaning “freak nature” [2]. However, the first case of ARSA causing dysphagia was described in 1733 [3] and the first radiologic description was made in 1936 by Kommerell [4,5]. In this situation, the right subclavian artery does not originate from brachiocephalic trunk, but distally and dorsally of the left subclavian artery [3]. Therefore, it typically has retroesophageal course and proceeds between the esophagus and spine in the 80% of the cases [6-8]. This anomaly is attributed to involution of the 4th aortic arch during the 6th to 8th week of gestation. As a result, the 7th intersegmental artery remains attached to the descending aorta [6]. The root of the ARSA is formed by the persisting right aortic arch and as a result a diverticulum at the proximal descending aorta can be found. This broad base of the ARSA is known as Kommerell’s diverticulum [5,9]. Commonest symptoms include dyspnea during childhood and dysphagia mainly for solid food with aging [3,6] (Table 1). In this article, we report two cases of late onset presentation of symptomatic ARSA. Due to medical comorbidities, subjects were managed conservatively.

Case

Case #1

A 69-year-old male, heavy smoker (50 p/y) with a history of myocardial infarction 10 years ago was presented to our hospital complaining of chest pain. At that time, percutaneous coronary intervention had been performed and two coronary stents had been placed. He was taking medication for heart failure (NYHA II). Because of his history he was subjected to cardiological examination that was unremarkable for pathology. After careful history, it was elicited that pain was becoming worse when swallowing. A contrast enhanced CT scan revealed ARSA (Figure 1). Blood pressure was the same on both hands. Due to heart failure, an attempt for conservative management was made. Symptoms became better with dietary modifications such as smaller meals and longer meal-time.

Case #2

A 73-year-old woman presented to our hospital with dysphagia that was gradually deteriorated during the last year. Paradoxically, symptoms of dysphagia were more prominent in liquids. On the chest radiography mediastinum enlargement was suspected and as a result a contrast enhanced CT was performed to exclude a tumor or an aneurysm (Figure 2). Manometry or motility studies were not performed. The patient had many comorbidities including a stroke one month before. As a result, a conservative management was preferred.
Table 1. Etiology of dysphagia related to ARSA.

- Aging resulting in increased esophageal rigidity or aortic elongation
- Presence of bicarotid trunk (the two carotids have the same origin from the aortic arch)
- Aneurysm formation often related to Kommerel’s diverticulum
- Atherosclerosis resulting to rigidity of the aberrant artery
- Decreased tracheal rigidity during childhood

Discussion

The incidence of ARSA varies between studies from 0.4 to 2.3% [6,10]. However, most of the studies come up with an incidence of 0.4 to 0.7% [11]. An aberrant left subclavian artery with the right sided aortic arch has also been described [12]. Even though, almost 70% of the persons with ARSA remain symptom free [6]. Symptoms, when present usually start during childhood and there is a peak after middle

Figure 1. Chest CT scan of the patient No. 1 showing the aberrant right subclavian course (white arrow).

Figure 2. Chest CT scan of the patient No 2 showing the aberrant right subclavian course (white arrow).
age forming a bimodal distribution with an average age of 48 [6]. Most symptomatic patients with ARSA complain of dysphagia for more than 6 months. It is usually for solid food but it gradually becomes worse. Some authors question whether dysphagia is caused by aberrant artery itself or by a coexisting motility dysfunction caused by esophageal [6]. This of course could be the case in some patients. From a Bayesian point of view, dysphagia symptoms are much more common in persons with ARSA and surgical treatment is often beneficial, indicating not only that ARSA could cause dysphagia, but also that external pressure should be the reason in most cases [13]. On the other hand, as ARSA and dysphagia are both common in older patients, incidental (false positive) findings cannot be excluded. In our case dysphagia mainly for the liquids is a specific symptom for achalasia. Unfortunately, we were not able to exclude this diagnosis. Chest pain as a symptom is described in about 20% of the cases presented [6,14], though it is usually accompanied with dysphagia.

Even if chest radiography [15] and manometry as well as findings in upper endoscopy may show abnormalities, first diagnostic procedure which should reveal this abnormality is barium esophagogram [6]. However, if it is unremarkable and dysphagia has a chronic course, a CT scan with contrast or CT angiography should be made [9]. Multidetector computed tomographic (MDCT) angiography is considered the golden standard if a surgical approach is to be followed [16,17]. In addition, if an ARSA is diagnosed with a CT in a person with dysphagia, further procedures should be made in order to exclude esophageal disease.

In almost every patient a conservative management should be the first-line treatment and includes both lifestyle or dietary modification and drug therapy [6]. When symptoms persist then a surgical approach relieves symptoms in most cases [13]. Surgical technique should be personalised according to the anatomical variants of the ARSA [3,11,13].

Many different surgical accesses have been described such as median sternotomy [18], left or right thoracotomy [19], cervical [12] or right supraclavicular [3,11,12]. Lately endovascular or hybrid approach with promising early results [20]. However, long-term prognosis needs to be examined. The reconstructions of the aberrant vessel may include simple ligation and division, right subclavian to ascending arch [16] or carotid artery [3,11,12]. Stent grafting can be used in cases of aneurysmal disease [16].

In our article, we presented a late onset presentation of two patients with symptomatic ARSA. They presented with chest pain without dysphagia and dysphagia mainly for liquids respectively, which are both considered as non-typical presentations of the anomaly. Dysphagia becoming worse with liquids may indicate a motility problem. Unfortunately, motility studies such as barium swallow and manometry were not performed as the patient was managed conservatively and failed to attend for follow up. On the other hand, symptoms persisted for about a year which is typical in such cases [12]. In both patients conservative management was selected with lifestyle changes and dietary modifications.

References