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CASOS CLÍNICOS

Dysphagia Lusoria and Zenker's Diverticulum

Dysphagia Lusoria y Diverticulum de Zenker. Reporte de caso

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| Resumen |

Una mujer de 81 años fue remitida para evaluación de una disfagia que padecía desde hacía cuatro años. Una endoscopia gastrointestinal reveló un divertículo de Zenker y una compresión extrínseca del esófago. Un escaneo computadorizado del tórax confirmó la presencia de la compresión extrínseca, causada por una arteria subclavia derecha aberrada, que comprometía la pared esofágica posterior, lo que confirmó un diagnóstico de disfagia lusoria. Una asociación entre el divertículo de Zenker y la disfagia lusoria es tan poco común que no han sido publicados sus casos con anterioridad.

Palabras clave: Divertículo de Zenker, Trastornos de Deglución, Estudios de Casos (DeCS).

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Summary

An 81-year-old woman was referred for evaluation of dysphagia which she had suffered for four years. An upper gastrointestinal endoscopy revealed a Zenker's diverticulum and extrinsic compression of the esophagus. A thoracic CT scan confirmed the presence of extrinsic compression caused by an aberrant right subclavian artery, which compromised the posterior esophageal wall thus confirming a diagnosis of dysphagia lusoria. An association between Zenker's diverticulum and dysphagia lusoria is so uncommon that there have been no previously published cases.

Key words: Zenker Diverticulum, Deglutition Disorders, Case Studies (MeSH).

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Introduction

Dysphagia is the sensation that swallowing is difficult or impossible. The word derives from the Greek roots *dys*, which means difficulty, and *phago*, which means to eat. In the broadest sense, dysphagia includes any mechanical or functional alteration in the passage of solids and/or liquids between the mouth and the stomach. However, this can be differentiated into *odynophagia*, which is painful swallowing, and *globus pharyngis* which is the sensation of having a lump in one's throat (in the pharynx or cervical esophagus). Dysphagia is an important cause of morbidity and mortality that can cause dehydration, malnutrition, laryngeal spasms, bronchial spasms, pneumonia related to aspiration and death due to obstructed air passages. It occurs frequently: it presents in 10% of people over the age of 50 years, in 25% of elderly people hospitalized for any reason, and in 40% of the patients in geriatric institutions (1-3).

Case report

An 81-year-old woman came to the gastroenterology service after suffering from dysphagia for four years.

Dysphagia was initially associated with solid foods, but six months prior to consultation the patient began to feel as if a foreign object was lodged behind her sternum. Patient had halitosis and occasional difficulty breathing, but did not report weight loss.

After an upper gastrointestinal endoscopy performed at a local hospital, a Zenker's diverticulum was founded. The patient had been referred for endoscopic septotomy. An important factor reported in the patient's medical history was her systemic arterial hypertension managed with losartan. Physical examination showed oropharyngeal erythema but no other alterations.

At the Hospital Universitario de La Samaritana (HUS-Samaritan Teaching Hospital), an esophageal x-ray showed a diverticular formation 20mm x 17mm in the cervical esophagus, indicated by a white arrow in Figure 1. The red arrow indicates extrinsic compression in the posterior wall of the thoracic esophagus at the top of the aortic arch together with multiple tertiary contractions in the distal third of the organ, which are compatible with an aging esophagus.

An esophageal endoscopy confirmed the presence of a diverticulum in cervical esophagus (Figure 2a) and extrinsic compression of the middle third of the posterior wall of the esophagus (Figure 2b).

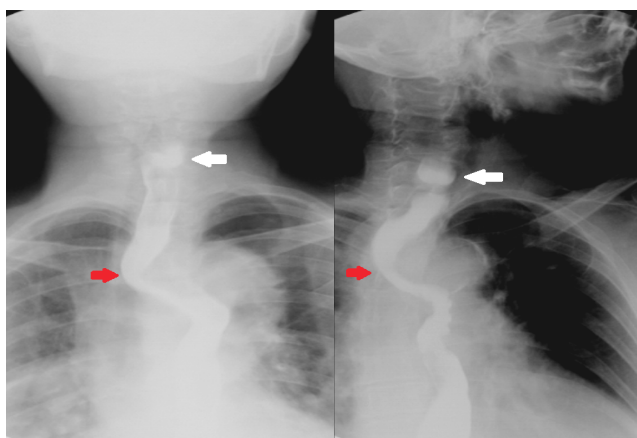


Figure 1. Esophageal x-ray.

Source: authors.

Note: the white arrows show a Zenker's diverticulum and the red arrows show extrinsic compression of the thoracic esophagus.



Figure 2a. Upper digestive endoscopy: Zenker's diverticulum.
Source: authors.

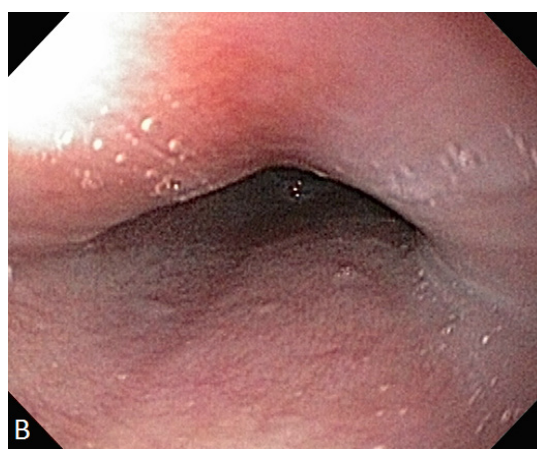


Figure 2b. Upper digestive endoscopy: Extrinsic Compression of the middle third of the esophagus.
Source: authors.

The images showed extrinsic thoracic compression, and a thoracic CT scan was ordered to rule out the possibility of mediastinal pathology. The scan showed changes in the morphology of the descending aorta. An aberrant right subclavian artery compromised the posterior esophageal wall, confirming the presence of dysphagia lusoria (Figure 3).

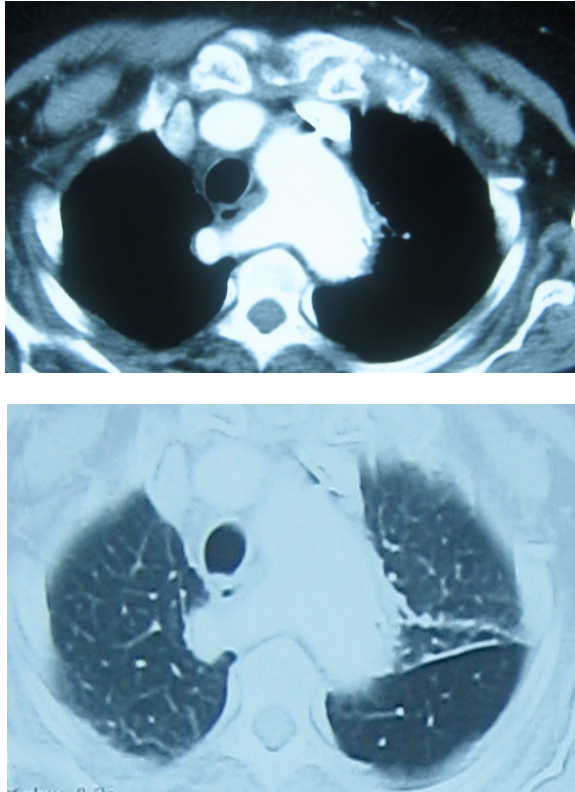


Figure 3. Thoracic CT scan.

Source: authors.

Note: the images show the presence of esophageal compression caused by an aberrant right subclavian artery.

Dysphagia lusoria

Dysphagia lusoria refers to dysphagia caused by extrinsic compression of the esophagus by mediastinal vascular structures. An aberrant right subclavian artery is the alteration associated with this condition which has been described most often in the literature. It is also the congenital malformation most often associated with the aortic arch. It has a prevalence of 1.8% (4). Other vascular malformations which cause extrinsic compression with dysphagia or respiratory symptoms include double aortic arches, persistent right aortic arches with patent ductus arteriosus, and aberrant innominate arteries (5-7).

In 1761, David Bayford, an English surgeon, described the case of an elderly woman who had died of severe malnutrition because of “obstructed deglutition”. He subsequently performed an autopsy on the woman and found that the cause of the obstruction had been an enlarged right subclavian artery that originated on the left side of the aortic arch and then crossed toward the right arm between the trachea and the esophagus causing esophageal compromise. He finally presented this case to a meeting of the medical society of London in 1787 (8).

The normal anatomy of the aortic arch is characterized by three great branches: the left common carotid artery, the left subclavian artery, and the brachiocephalic artery. The brachiocephalic artery passes upwards, outwards and behind the left brachiocephalic vein and in front of the trachea. Finally, it divides into the common carotid artery and the right subclavian arteries at the sternoclavicular joint. The dysphagia lusoria (in the case presented) was the result of abnormal development of the aortic arch into four branches whose origins were separated by the right common carotid artery and the right subclavian artery resulting in a trajectory behind the esophagus, which caused the extrinsic compression.

This alteration can be asymptomatic. In such cases, its diagnosis is likely to be incidental to a thoracic imaging study. During infancy, dysphagia lusoria may manifest itself as a combination of dysphagia and dyspnea while among adults its symptoms are most often dysphagia associated with solid foods (7).

The initial evaluation of patients should include barium X-rays of the esophagus (esophograms), thoracic CT scans, and complementary esophageal endoscopy (9). CT scans are indicated for patients with suspected mediastinal alterations such as those whose angiographs have demonstrated aberrant blood vessels. Modern multi-slice 3D CT scanning is now the diagnostic technique of choice both because of its high resolution and because it is not invasive. (10,11)

Zenker’s diverticulum

Zenker’s diverticulum, also known as pharyngoesophageal diverticulum, occurs most frequently among the elderly. It is the formation of a pouch in the upper esophageal sphincter, which obstructs the passage of food. Although it was first described by Ludlow, it takes its name from German physician Friedrich A. von Zenker who published an article about it in 1877 that included a series of 27 cases with complete descriptions of its clinical and pathological characteristics.

He defined it as a herniation of the mucosa of the posterior hypopharyngeal wall extending through the muscle layer (12).

Esophageal diverticula classifies into three types according to their locations. Zenker's diverticula are those pharyngeal herniations that occur within Killian's dehiscence, a weak area in between the thyropharyngeus part of the inferior constrictor of the pharynx and the cricopharyngeus muscle. Middle third esophageal diverticula are located down to 5 cm below the tracheal carina while epiphrenic diverticula occur in the lower 10 cm of the organ (13).

Failure of coordination related to either the superior or the inferior sphincter can produce increased intraluminal pressure, protrusion of the mucosa, and diverticula. Increased hypopharyngeal pressure can cause Zenker's diverticula, just as abnormal traction or pushing of the esophagus can produce diverticula in the middle and lower thirds of the esophagus.

The prevalence of esophageal diverticula in the whole population is estimated to be between 0.02% and 0.77% although the majority of diverticula do not present symptoms. At least 3% of all dysphagia patients are diagnosed with diverticula. The majority of these cases occur among elderly patients who visit the doctor because of severe halitosis, laryngeal symptoms and respiratory symptoms (13,14).

Although Zenker's diverticula are pulsion diverticula, the cause of high hypopharyngeal pressure and lack of coordination of the superior esophageal sphincter during swallowing cannot be identified in the majority of patients. Nevertheless, various theories postulate the cause as a constrictive myopathy of the sphincter combined with spasms of the sphincter. This type of diverticulum has been reported to be associated with various illnesses that, in one way or another, produce alterations of the normal movement of the esophagus and increase intraluminal pressure in the esophagus. These pathologies include gastric, esophageal and laryngeal neoplasias, esophageal rings in the upper third of the esophagus, Chagas disease, systemic diseases such as poliomyelitis, and anatomical defects such as hiatal hernias, reported in more than 30% of these patients (15-22).

Diagnosis base upon clinical evidence and confirmed with a barium esophogram that will show an accumulation of the contrast medium in the proximal third of the esophagus. In some cases, endoscopy of the upper digestive tract may be useful for detection of this abnormality (13). Treatment should usually be conservative and based on life style changes for patients with multiple comorbidities, small diverticula, or few symptoms. On the other hand, cricopharyngeus myotomies, either endoscopic or surgical, may be indicated to divide the

septum and allow the passage of food around the diverticulum and through the esophagus (23-25).

Discussion

No cases of dysphagia lusoria associated with Zenker's diverticulum have been previously reported in the literature of the world. In 1965 Schermuly reported on a case of Zenker's diverticulum associated with achalasia in Germany. Two decades later Schwartz and cols., postulated that achalasia and esophageal rings could cause pharyngeal protrusions such as Zenker's diverticula. They proposed that these pathologies should be ruled out before deciding upon any treatment of these cases. One of the author's of this article has reported on a case in which Zenker's diverticulum was associated with achalasia (26). Persistence of the motor disorder generated by achalasia is conducive toward increased intraluminal pressure in the esophagus followed by development Zenker's diverticulum, worsening dysphagia and increased risk of bronchial aspiration.

As the population of elderly people in Colombia grows, the number of patients with dysphagia due to various causes will increase. This will require well informed physicians who are prepared to perform the appropriate studies to establish correct and precise diagnoses and offer the best solutions available in our environment in an opportune manner.

Conflict of interest

None declared by the authors.

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