

Nutcracker esophagus, a rare presentation in the spectrum of esophageal motility disorders. Case report and literature review

Esófago en cascanueces, una infrecuente presentación del espectro de trastornos de la motilidad esofágica. Presentación de caso y revisión de literatura

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Summary
Esophageal motility disorders represent a spectrum of varied pathologies that present with distinct clinical manifestations and imaging findings. Nutcracker esophagus, named for its appearance on imaging studies, is a rare condition with a low incidence and unknown etiology. Its initial clinical manifestation is retrosternal pain of non-cardiac origin. Patients suffering from this disease are usually evaluated for conditions unrelated to the digestive system, until a correlation between food intake and swallowing inability becomes clear, which allows for a reorientation of the course within the diagnostic algorithm. There is no definitive treatment for all cases; this is a matter of individual testing and trial. We present a case of nutcracker esophagus, with classical findings of the disease, on radiological, endoscopic, and manometric studies.

Resumen
Las alteraciones de la motilidad esofágica representan un espectro de patologías variadas que cursan con distintas manifestaciones clínicas y hallazgos imagenológicos. El esófago en cascanueces, cuyo nombre se acuña debido a su aspecto en estudios de imagen, constituye una rara patología de baja incidencia y etiología desconocida, cuya manifestación clínica inicial es el dolor retroesternal de origen no cardiaco. Los pacientes que padecen esta enfermedad usualmente son estudiados buscando patologías no relacionadas con el sistema digestivo hasta que, eventualmente, se hace clara una correlación entre la ingesta de alimentos y la imposibilidad para deglutir, lo cual permite reorientar el curso en el algoritmo diagnóstico. No existe un tratamiento fijo y definitivo en todos los casos; este es un asunto de prueba y ensayo individual. Se presenta el caso de un esófago en cascanueces, con hallazgos clásicos de la enfermedad, tanto en imagen radiológica como endoscópica y manométrica.

Introduction
Nutcracker or corkscrew esophagus is a disorder characterized by increased peristaltic amplitude in the distal esophagus, accounting for 48% of esophageal motility disorders (1). The pathogenesis is unknown, although several theories have been proposed: an imbalance between excitatory and inhibitory neurons in the esophagus, inadequate autonomic innervation leading to a hypercholinergic state, thickening of the esophageal musculature, or hypertensive contractions of the organ due to chronic gastroesophageal reflux. Stress has also been suggested as a probable cause (2).

In a study conducted by Öztürk et al. on the epidemiology of the disease, they concluded that the average age of onset is 52 years, it is predominant in men, and the main symptom is retrosternal pain of non-cardiac origin (2).

Case presentation and radiological findings
A 66-year-old man with a history of controlled hypertension as his only significant medical history, who reports a one-month history of a sudden subjective sensation of being unable to swallow while eating, accompanied by retrosternal pain. He presented as an outpatient to the gastroenterology department for an esophagogastroduodenoscopy, which revealed a spirally tortuous esophagus, apparently due to tertiary-appearing peristaltic waves (altered motility), suggesting an esophageal motor disorder caused by a nutcracker esophagus (Figure 1); the stomach showed no abnormalities. Biopsies were taken from the esophagus and stomach, which showed no acute or chronic inflammatory infiltrates or neoplastic pathology.

Subsequently, to assess swallowing dynamics, a barium esophagram was performed, which was interpre-

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ted as follows: motility disorder with a corkscrew appearance due to a hypercontractile esophagus (Figure 2). The swallowing sequence in the esophageal portion, illustrated in the fluoroscopy video frames, more clearly demonstrates ascending hypercontractility originating from the lower esophageal sphincter (LES), which causes reflux up to the proximal third of the esophagus (Figure 3).

Medical treatment was initiated based on the typical findings observed during esophagogastroduodenoscopy and barium esophagram. Due to his underlying hypertension, the patient has been treated with amlodipine at a dose of 5 mg/day for more than 10 years; in addition, he uses sildenafil weekly as part of his sexual activity. Therefore, the treating gastroenterologist proposed adding 5 mg of this medication daily as part of his therapy.

After two months of treatment with sildenafil, the patient reported complete resolution of symptoms. At this time, an esophageal manometry was performed with normal results, showing a lower esophageal sphincter with normal tone and adequate relaxation, with a distal wave amplitude of 120 mm Hg. Esophageal body motor activity was reported as normal, with 100% normal peristaltic waves. Upper esophageal sphincter with adequate pharyngoesophageal coordination. Rapid-swallow provocation test with adequate swallowing inhibition and peristaltic reserve. Swallows in the sitting position were normal peristaltic (Figure 4).

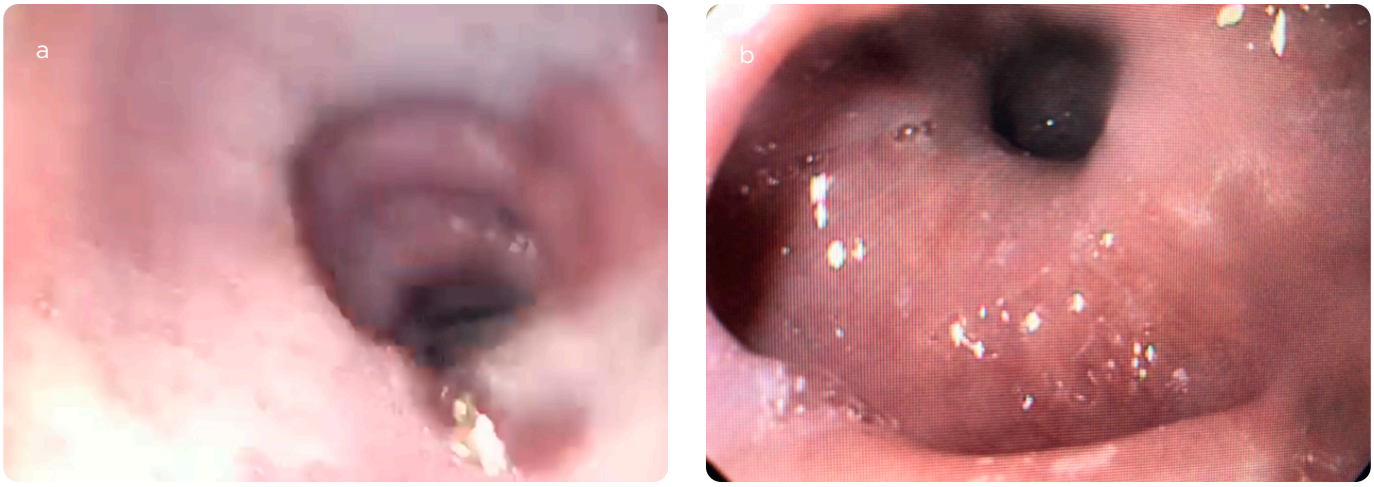


Figure 1. (a) and (b) The spiral appearance of the esophagus, clearly visible on esophagogastroduodenoscopy, from the proximal third (a) to its distal end (b).

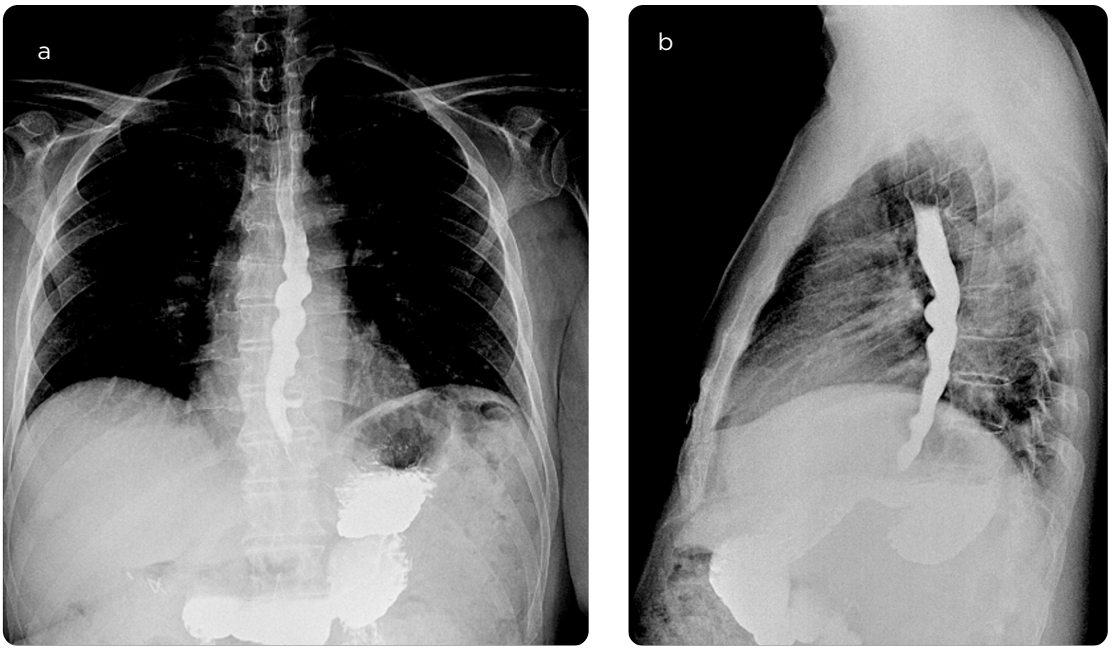


Figure 2. a, b, and c) Radiographic projections of the barium esophagram, which show the spiral appearance—referred to in the literature as a corkscrew or nutcracker—in both the frontal (a) and lateral (b) views. Note the thread-like, almost imperceptible appearance of the distal end of the esophagus, due to its hypercontractility. The gastric chamber appears normal, with no diaphragmatic hernia.

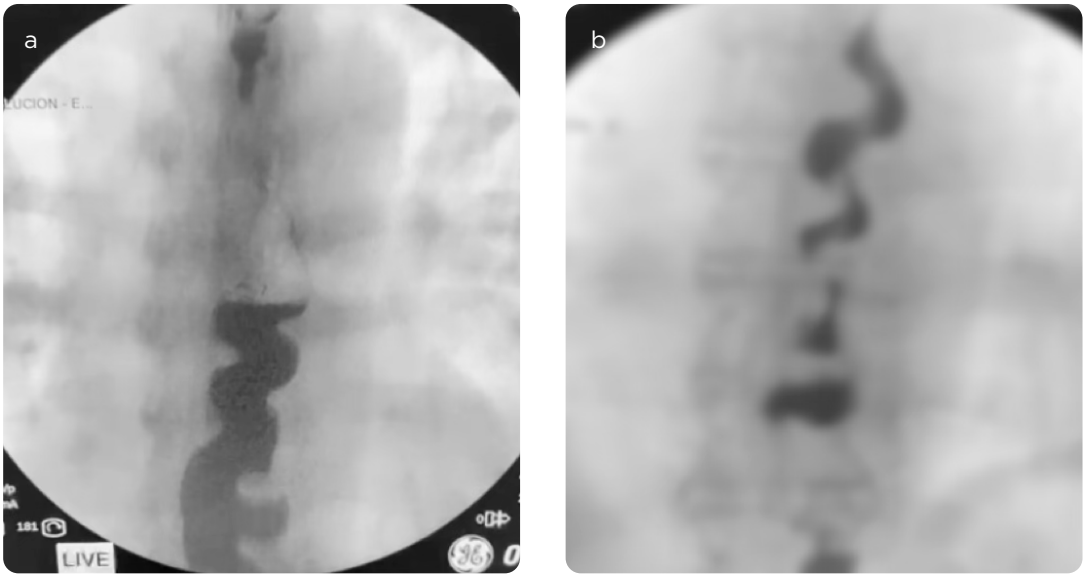


Figure 3. a and b) Fluoroscopy of the barium esophagram illustrates how, during swallowing, the ascending hypertonicity of the distal end causes a momentary stagnation of the food bolus in the middle and distal thirds of the esophagus (a). Subsequently, there is a partial relaxation of the lower esophageal sphincter (LES), which allows barium to enter the gastric chamber, leaving a considerable residue of it in the form of reflux, which ascends again to the proximal third of the esophagus (b).

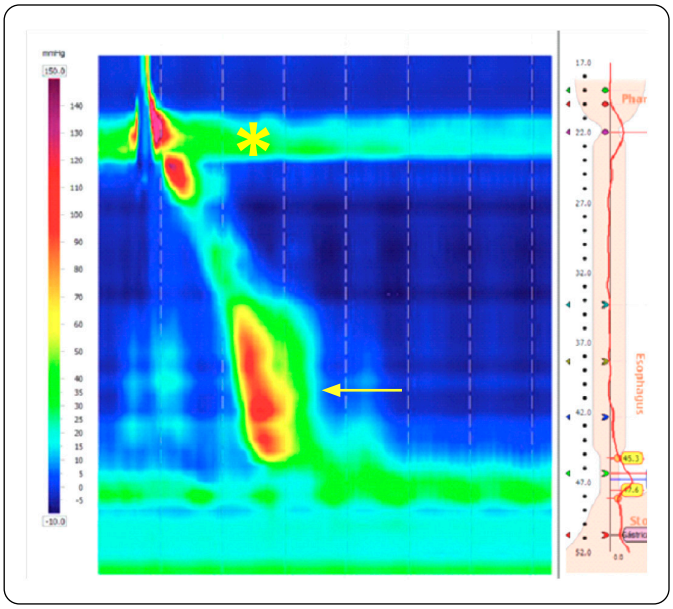


Figure 4. Esophageal manometry following initiation of sildenafil therapy. In the upper left quadrant, in the red range, an adequate contractile pattern is evident at the junction of the pharynx and the proximal third of the esophagus (asterisk), which is subsequently accompanied by a point of contractile deceleration (arrow) at the expected site in the distal esophagus, with an adequate decrease in velocity relative to the wave.

Discussion

The esophagus is a hollow, deformable muscular tube that runs in a relatively straight path toward the abdomen and measures approximately 24 cm in length in adults. Histologically, the esophageal wall consists of the following layers, starting from the lumen and moving outward: mucosa, submucosa, outer muscle layer, and adventitia (3). Auerbach's plexus, also known as the myenteric plexus, is composed of a group of ganglia that run the entire length of the gastrointestinal tract and innervate multiple layers of smooth muscle. These nerves, located between two layers of the external muscularis, combined with others originating from Meissner's plexus in the submucosa, constitute the enteric nervous system. The myenteric plexus is primarily responsible for motility and peristalsis (2).

The four main patterns of esophageal (manometric) abnormalities include: 1) inadequate relaxation of the LES; 2) uncoordinated contraction; 3) hypocontractility; and 4) hypercontractility. Of these, the corkscrew esophagus belongs to the fourth group, along with isolated LES hypertension (4).

The cause of most esophageal motility disorders is not well understood. In many cases, symptoms may not respond to therapies that correct the abnormalities seen on manometric testing. For example, patients presenting with retrosternal pain and a high-amplitude peristaltic wave on manometry do not report pain relief despite treatment with calcium channel blockers, which cause a significant decrease in the amplitude of the contraction wave (4). Consequently, it is not entirely clear whether certain esophageal motility disorders are actually disorders; in some cases, the observed manometric abnormality may correspond to a simple epiphenomenon (an effect or association, but not the cause). Finally, there may be no non-manometric means to validate the diagnosis of disorders that have been defined solely on the basis of manometric abnormalities, such as nutcracker esophagus (4).

In the body of the esophagus, abnormal motility manifests as uncoordinated contraction, hypercontractility, or hypocontractility. In patients with conditions causing both inadequate relaxation of the lower esophageal sphincter (LES) and impaired esophageal body function (achalasia), symptoms are primarily caused by LES dysfunction. This finding has important physiological and therapeutic implications, as these symptomatic individuals have a disorder with clinical manifestations, and this aspect is of the utmost importance (4).

The term "nutcracker esophagus" was coined by Castell et al. to describe a condition in which patients present with retrosternal pain and dysphagia, exhibiting peristaltic waves in the distal esophagus with amplitudes exceeding the expected value by more than 2 standard deviations (SD) (5). In 1979, Benjamin et al. reported the average amplitude of the distal esophageal peristaltic wave in 40 healthy individuals as 81 mm Hg. The authors suggest a cutoff point of 120 mm Hg for the

nutcracker esophagus as a manometric diagnostic criterion for peristaltic waves of increased amplitude (6); however, a subsequent study of 95 healthy adults found the average amplitude of the distal esophageal peristaltic wave to be 99 mm Hg; consequently, several later reports have adopted 180 mm Hg as the cutoff point for diagnostic criteria, which is currently accepted (7). This cutoff value should be obtained from the average of 10 swallowing attempts, taken at two different sites, 3 cm and 8 cm proximal to the EEI (3).

Radiographic evaluation of patients with dysphagia is useful for assessing both the structural and functional anatomy of the esophagus. Excluding cases of mild esophagitis, the barium esophagram is capable of detecting up to 90–95% of structural abnormalities that may cause dysphagia, including esophageal carcinoma, areas of stenosis, and severe forms of reflux esophagitis. The sensitivity for detecting a nutcracker esophagus varies depending on the study, ranging from 56% to 89% (8).

In the case presented, a condition of unknown origin is identified, and several options are proposed for its medical management, including: nitrates, phosphodiesterase-5 inhibitors (sildenafil), calcium channel blockers (amlodipine, diltiazem), peppermint oil, and low doses of proton pump inhibitors (omeprazole) (9).

The effectiveness of these medical management options varies greatly from person to person, as some patients report partial relief of symptoms while others show no improvement.

Myotomy as a surgical management option is a more aggressive and apparently effective approach, as documented by Kahrilas in his report, in which 100% of patients reported improvement in symptoms; however, the number of patients who underwent surgery and were followed up at 2 years was only four (10). This approach was ruled out for the time being in the patient described here.

The low prevalence of this disease and the characteristic and typical imaging features make this a relevant case, and this literature review is one of the few available in Spanish. It is important to become familiar with the classic corkscrew-like image, which allows for a suggested diagnosis, leading to timely therapeutic intervention in the patient to alleviate symptoms.

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