

Sigmoid-shaped esophagus due to achalasia in an 80-year-old female: Case report

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Abstract

Esophageal achalasia is the most common of the primary esophageal motility disorders. It is characterized by the lower esophageal sphincter inability to relax. It significantly affects patient's quality of life and can be difficult to diagnose and treat. The authors present a case of an 80-year-old female patient who complained of dysphagia and weight loss. After further investigation she was diagnosed with end-stage achalasia. Considering her poor general condition, endoscopic pneumatic dilatation of the LES was performed with satisfactory results.

Keywords

esophageal achalasia; deglutition disorders; esophagus; contrast esophagogram

Abbreviations

LES: Lower Esophageal Sphincter; CT: Computed Tomography

Introduction

The word "achalasia" derives from the Greek word *khalasis*, meaning "not loosening or relaxing" [1]. It defines the lower esophageal sphincter (LES) inability to relax in response to swallowing. In most cases, this is also associated with absence of esophageal peristalsis [1,2]. It is a rare disease [4] and the exact etiology remains unknown, however neuronal degeneration, autoimmune and infectious processes seem to be determinant to its development [1,3].

The clinical presentation includes progressive dysphagia to both solids and liquids (the most common symptom), regurgitation, chest pain, heartburn, halitosis, odynophagia and weight loss [2,3].

In absence of therapy, progressive dilatation and increasing tortuosity of the esophagus may occur, resulting in end-stage disease [4].

Case Presentation

An 80-year-old Portuguese female, with past medical history of high blood pressure and dyslipidemia and with no surgical history, was referred to our unit with longstanding intermittent dysphagia and weight loss. She underwent an extensive workup, where an Esophagogastroduodenoscopy revealed a tortuous and dilated esophagus (Figure 1 & 2). The Contrast Esophagogram (Figure 3) showed esophageal dilatation and a narrowing at the gastroesophageal junction producing a “bird’s beak” image. Due to the poor collaboration of the patient, it was difficult to locate the LES upon the Manometry but it was possible to identify abnormal esophageal motility with low amplitude contractions and no profitable peristalses. A CT scan (Figure 4 & 5) was also performed revealing major distension of all esophageal lumen (8 cm of major diameter) and funnel-shaped cardia, suggesting achalasia and ruling out evidence of malignancy or extrinsic compression.

The patient initially refused any treatment by fear but, due to the progression of the disease, she began to suffer from severe dysphagia, which caused a need for more hospitalizations. Thus, she agreed with Endoscopic Pneumatic Dilatation of the LES.

Discussion & Conclusion

End stage achalasia occurs in less than 5% of all achalasia patients [4].

The diagnosis is achieved by performing a careful evaluation of symptoms combined with diagnostic testing. Patients typically present with severe dysphagia and nutritional failure. Radiological features include a tortuous (sigmoid) and massively dilated esophagus associated with a distal narrowing in a classic “bird’s beak” or “rat’s tail” at the level of the gastroesophageal junction [4].

Esophageal manometry is the gold standard study and demonstrates the incomplete relaxation of the LES after deglutition [1-3]. Esophagogastroduodenoscopy is useful for excluding secondary causes of achalasia (gastrointestinal reflux disease, eosinophilic esophagitis, radiation or medication-induced strictures, Chagas disease, malignancy or extrinsic compression) and may show a dilated esophagus [1]. Due to the patients age, it was important to exclude malignancy.

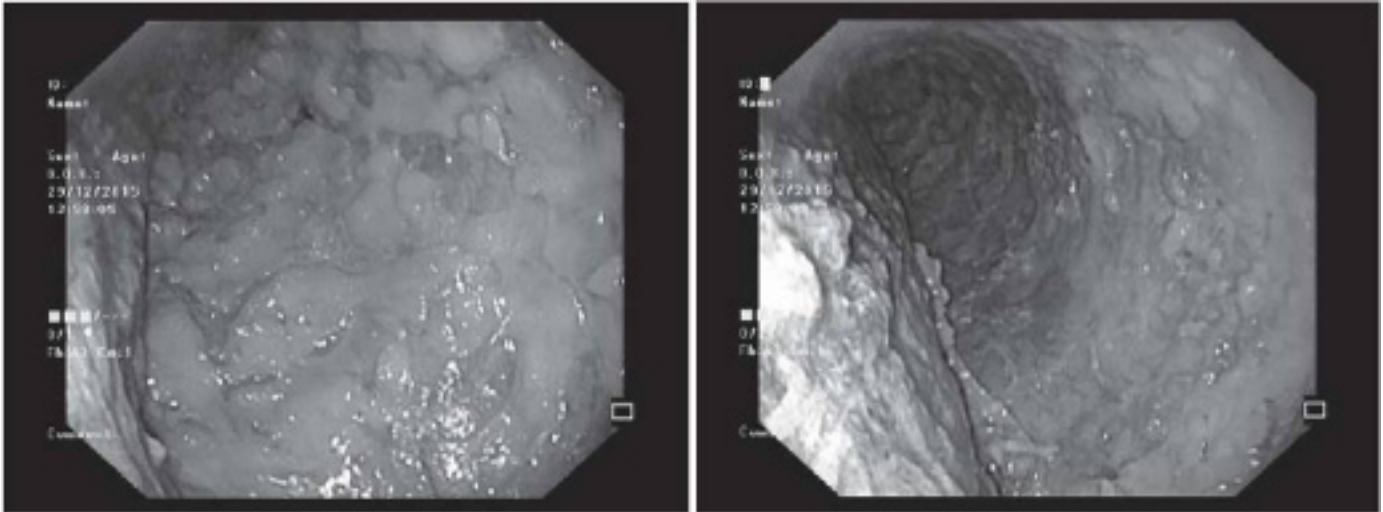
The management of patients with end-stage disease is challenging and no treatment is available to restore esophageal peristalsis and normalize LES relaxation [4]. In this case, the patient had little education and a low cultural background which led her to refuse any treatment for a long time by fear. After several approaches and due to progressive deterioration of her quality of life, she ultimately agreed with treatment.

The possible palliative treatments for this disease are: 1. Drugs (nitrates, calcium antagonists); 2. Endoscopic Pneumatic Dilatation of the LES; 3. Botulinum Toxin injections; 4. Surgery (Heller Myotomy and Fundoplication, Esophagectomy) [1-3].

We proposed Endoscopic Pneumatic Dilatation to this patient due to her poor general condition and because it has a greater efficacy in older patients [4]. Besides that, we had to take into consideration her non-acceptance for more invasive procedures.

The patient is currently tolerating liquids and soft diet with no vomiting episodes. She will be continuing with the follow up for identification of eventual relapse or pre-malignant lesions appearances.

Figures



Figures 1 & 2: Esophagogastroduodenoscopy – esophageal lumen filled with food debris.



Figures 3: Computed Tomography Scan - distension of all esophageal lumen (8 cm of major diameter) with food impaction.



Figures 4: Computed Tomography Scan - distension of all esophageal lumen with food impaction.

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