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Case Report Open Access

Circumferential Esophageal Dissection Treated Conservatively in a Young Patient with Eosinophilic Esophagitis

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Abstract

Eosinophilic esophagitis is an immune/antigen-mediated chronic esophagueal disease. The most frequent symptoms include dysphagia, food impaction, pain and gastroesophageal reflux. Endoscopic findings include esophageal rings, whitish exudates, longitudinal furrows, edema, esophageal narrowing and mucosal fragility, which may lead to complications such as esophageal perforation, mucosal tear or intramural dissection. Esophageal dissection is a rare entity mainly described in women receiving anticoagulant treatment or affected by coagulopathy; very few cases have been reported in patients with eosinophilic esophagitis, whether spontaneous or iatrogenic. We describe the case of a young 17-year-old male who suffered circumferential esophageal dissection as a consequence of a gastroscopy performed after presenting symptoms of food impaction and was subsequently diagnosed with eosinophilic esophagitis. As a result of the few cases reported thus far, there is not a unified standard of treatment for this complication; surgical, endoscopic and conservative treatments have been reported. In our case, the patient was successfully treated conservatively. In conclusion, this case report confirms the need to be extremely careful while performing endoscopy in EoE patients (including patients with suspected EoE) and the possibility to treat intramural circumferential esophageal dissection conservatively if perforation has been ruled out.

Keywords: Eosinophilic esophagitis; Intramural esophageal dissection; Circumferential esophageal dissection; Esophageal perforation

Abbreviations: EoE: Eosinophilic Esophagitis; GERD: Gastroesophageal Reflux Disease; PPI: Proton-Pump Inhibitors; CT: Computed Tomography

Introduction

Eosinophilic esophagitis (EoE) is an immune/antigen-mediated chronic disease characterized clinically by symptoms of esophageal dysfunction (dysphagia, food impaction, thoracic pain, symptoms of gastroesophageal reflux disease (GERD), abdominal pain) and histologically by eosinophil-predominant inflammation (15 eosinophilis per high-power field) [1]. EoE is restricted to the esophagus. The differential diagnosis of EoE includes other causes of esophageal eosinophilia such as: GERD, eosinophilic gastroenteritis, Crohn's disease, celiac disease, infection, achalasia and vasculitis. EoE is primarily present in young men and is frequently associated with a personal or a family history of atopy (asthma, rhinitis, atopic dermatitis or food allergies). An increase in its incidence and prevalence has been noted in the past years [2,3].

Since histology is basic for the diagnosis of EoE, biopsies obtained by means of upper endoscopy are still essential for the diagnosis. Typical endoscopic findings, although no pathognomonic include mucosal fragility, edema, fixed (corrugated esophagus) or transient rings (felinization), longitudinal furrows, whitish exudates and esophageal stenosis (which may require endoscopic dilation).

The following treatments have demonstrated effectiveness: elimination diets, topic corticoids (or systemic in severe cases) and endoscopic dilation. Proton-pump inhibitors (PPI) can relieve GERD symptoms that may coexist even in the absence of pathological reflux as defined by conventional criteria. Studies based on immunosuppressive drugs and monoclonal antibodies (anti-IL5, anti-IL13 and anti-eotaxin) are currently underway [4].

Esophageal stenosis, esophageal perforation (spontaneous, presenting as Boerhaave syndrome or iatrogenic), intramural esophageal tear or intramural dissections are among the possible complications of this disease.

Herein we present the case of a young who suffered circumferential esophageal dissection as a consequence of a gastroscopy performed after presenting symptoms of food impaction and was subsequently diagnosed with eosinophilic esophagitis.

Case Report

17-year-old male, with no relevant previous diagnoses in his medical history, was admitted at the Emergency room of our hospital reporting he perceived the presence of a foreign body just below the cervical gland level after eating sunflower seeds 2 days earlier. Fibroscopy was unrevealing. A subsequent gastroscopy observed superficial erosion at the level of the upper esophageal sphincter, no foreign bodies, and a corrugated esophagus with a slight reduction of his caliber that did not hinder the passage of the endoscope, all of it compatible with a diagnosis of eosinophilic esophagitis (Figure 1). The patient had poor gastroscopy tolerance by means of frequent retching, and as distal esophageal biopsies were taken, blood coming from proximal regions was observed. Extensive mucosal tears were visualized in the medium and proximal esophagus during withdrawal. At the end of the procedure, the patient presented cervical and supraclavicular

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subcutaneous emphysema. A thoracic-abdominal CT was performed, resulting in the finding of pneumomediastinum and the presence of air dissecting the esophageal wall completely, without evidence of perforation (Figures 2 and 3). The laboratory test showed neutrophilic leukocytosis and elevated C-reactive protein.

In view of the findings, the patient was hospitalized at the Gastroesophageal Surgery Unit of our hospital and conservative treatment with antibiotics, intravenous fluid therapy and total parenteral nutrition was implemented for 9 days. A barium esophagogram ruled out contrast leaks. The patient responded to treatment positively and oral tolerance was achieved by the sixth day. Before the patient left the hospital EoE treatment was initiated (1 mg of viscous Budesonide every 12 hours and PPIs) and maintained until control endoscopy 5 months later. The patient was questioned again and admitted having previous swallowing difficulties, including 3-4 episodes of food impaction since the age of seven that spontaneously resolved. He did not recall having thoracic pain, GERD symptoms or abdominal pain.

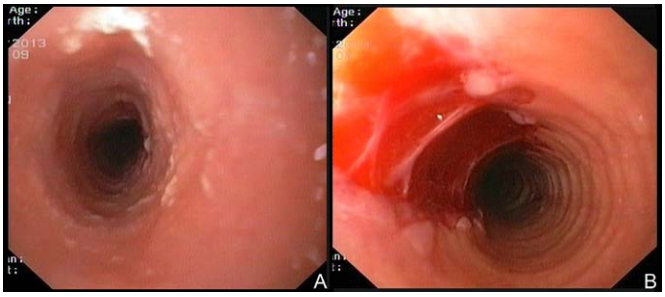

Figure 1: A) Corrugated esophagus with dispersed white exudates. **B)** Mucosal tear in middle esophagus.

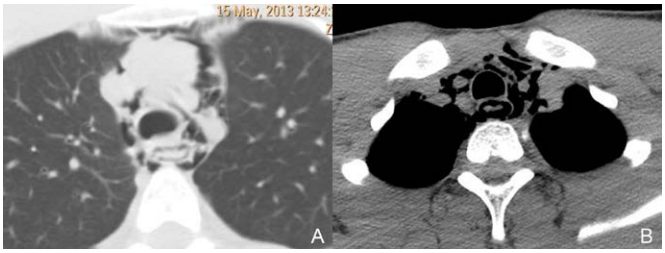

Figure 2: A) y **B)** Air dissecting the structures of mediastinum, neck and circumferentially in the esophageal wall, which is unbroken.

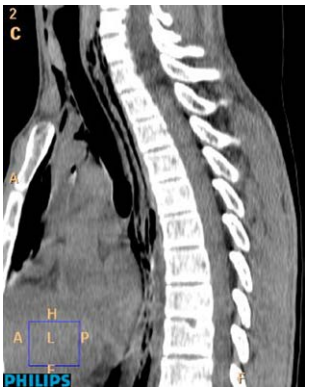

Figure 3: Sagittal reconstruction of CT, showing the presence of air dissecting the esophageal wall completely.

Allergologic tests showed subclinic sensitization to acarus, with no food sensitization, and so the patient was kept on a normal diet.

In subsequent checkups the patient remained asymptomatic. pH-metry showed no evidence of pathologic acidic reflux. No pathological findings were observed in a control CT performed two months after the patient was admitted at the hospital (Figure 5). Five months after discharge a control gastroscopy was performed, under sedation and using an ultrathin endoscope to minimize risks, given the previous complication. The esophagus showed mucosal edema, a diffuse squamous exudate and longitudinal furrows but no rings (Figure 4). An eosinophilic infiltrate was still visible in biopsies from the gastroesophageal junction, distal and proximal esophagus.

Discussion

Circumferential dissection is a highly infrequent subtype of esophageal intramural dissection, which is a rare entity first described in 1968 by Marks and Keet [5]. Most common etiologies include iatrogenic (gastroscopy, transesophageal echocardiography, variceal sclerosis) and non-iatrogenic causes (impaction or swallowing of foreign bodies). Dissection can also happen spontaneously, generally as a consequence of a hematoma in the context of coagulopathy, but also in healthy individuals after an episode of retching or vomiting leading to a Mallory-Weiss or Boerhaave syndrome [6].

Common symptoms include thoracic pain, dysphagia, odynophagia and hematemesis. Nausea, vomiting and back pain have been described as well.

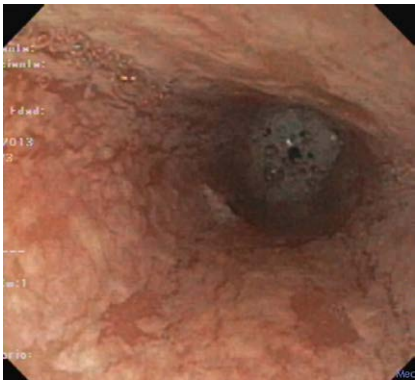

Figure 4: Esophageal re-epithelialization with no stenosis but with exudates.

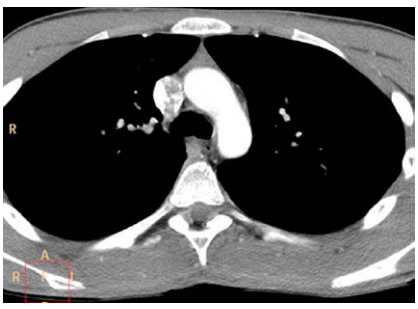

Figure 5: Resolution of dissection and pneumomediastinum.

The diagnosis is achieved by means of imaging techniques such as CT and esophagogram. CT may show a double lumen (a true and false lumen), while in a esophagogram the presence of contrast between the mucosal and submucosal esophageal layers may be seen. More invasive techniques such as upper endoscopy and echoendoscopy should be avoided whenever possible because of the increased risk of esophageal perforation.

In our case, the patient, who lacked a relevant medical record, showed a lineal mucosal tear during a gastroscopy that secondarily led to a circumferential dissection as observed in CT. The endoscopic findings and posterior biopsy results confirmed the diagnosis of eosinophilic esophagitis. The higher fragility of the esophageal wall in EoE patients has been previously associated with a higher risk of iatrogenic and spontaneous perforation [7-12]. However, we could find only 4 reported cases in which the patient, as ours, suffered esophageal dissection [4,7,8,13]. Even though uncommon, it is thought that chronic eosinophilic inflammation produces mucosal friability and a more fragile esophageal wall allowing for a propagation of a superficial damage deep into the wall and thus leading to dissection.

The treatment of esophageal dissection is typically conservative, involving intravenous fluid-therapy and total parenteral nutrition, hence avoiding the risks of a surgical procedure, which is only considered in severe cases. Even though several endoscopic techniques, including the use of auto-expansible metallic prosthesis, have been reported [11], they are not recommended because of the high risk of perforation.

Our patient displayed an extensive circumferential dissection of the esophageal wall as shown in the CT, making it less susceptible for a conservative approach [4,6]. In fact, the other 2 cases in which patients with EoE had circumferential esophageal dissection reported to this date had complications requiring surgery [7,8]. However, the CT and esophagogram findings in our patient ruled out perforation. Moreover, the presence of subcutaneous emphysema and pneumomediastinum could be explained by the fact that the slimming of the esophageal wall allowed air diffusion to those areas. The patient responded positively to the conservative treatment and the absence of complications (stenosis, lumen duplication…) was confirmed in a follow up control endoscopy carried out 5 months later.

Conclusion

- This case report confirms the need to be extremely careful while performing endoscopy in EoE patients (including patients with suspected EoE) with fibrostenotic features (stricture, narrow calibre, multiringed esophagus) because the risk of potential complications (esophageal tear, dissection or perforation) might be higher.
- Intramural esophageal dissection presenting in EoE patients, including circumferential dissection, is amenable to conservative treatment if perforation is ruled out.

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