Eosinophilic Esophagitis: Analysis of Food Impaction and Perforation in 251 Adolescent and Adult Patients

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Background & Aims: Eosinophilic esophagitis is a rapidly emerging, chronic inflammatory disorder. Prolonged inflammation evokes structural alterations and a fragile esophageal wall prone to perforation/rupture and food impaction. This report assesses the risk of spontaneously arising and procedure-induced complications and proposes practical recommendations.

Methods: The Swiss Esophageal Esophagitis Database documented 251 confirmed cases. A chart review identified which patients had required endoscopic bolus removal and/or experienced transmural esophageal perforation/rupture. In addition, a MEDLINE search for “eosinophilic esophagitis” with “esophageal perforation” or “esophageal rupture” was undertaken.

Results: During an 18-year period, 87 patients (34.7%) experienced 134 food impactions requiring flexible (124, 92.5%) or rigid (10, 7.5%) endoscopic bolus removal. Transmural perforation occurred in 20% (2/10) of rigid procedures, and 1 esophageal rupture (Boerhaave’s syndrome) was observed.

Conclusions: Bolus removal by rigid endoscopy is a high-risk procedure and should be avoided in eosinophilic esophagitis patients who require a gentler approach. Whether food impaction and esophageal wall remodeling can be prevented with anti-inflammatory medication is still undetermined. All Boerhaave’s syndrome cases should be evaluated for underlying eosinophilic esophagitis.

Eosinophilic esophagitis (EE) is a clinicopathologically defined disease, characterized by esophagus-related symptoms in combination with a dense esophageal eosinophilia, both of which are unresponsive to prolonged acid suppression with proton pump inhibitors (PPIs).1 EE is identified with considerable frequency2,3 and is a leading cause of dysphagia4 and food impaction.5 A huge body of evidence indicates that one sequela of long-standing uncontrolled eosinophilic inflammation is esophageal wall remodeling,6–9 which produces an extremely fragile and inelastic mucosa that tears easily. It is very likely that the altered esophageal wall structure presents a risk factor for both superficial lacerations as well as even deeper lesions, including transmural perforation or rupture. This risk is illustrated by articles documenting 5 EE patients who incurred procedure-induced esophageal perforations9–11 and 2 patients cited in the literature who experienced spontaneous esophageal ruptures associated with a retching event.11,12 This report presents another 3 EE patients with esophageal breaches. On the basis of the experience of our long-term cohort and on a literature review, we propose practical recommendations for gastroenterologists and ear, nose, and throat (ENT) surgeons. Our goal is to reduce the risk of further procedure-related esophageal perforations and to help protect physicians from medicolegal issues when treating patients with dysphagia.

Methods

The Swiss EE Study Group consecutively enrolled patients presenting with a confirmed (by using clinical, endoscopic, and histologic methods) diagnosis into a Swiss EE Database, SEED, which was nationally established in 1989. Inclusion criteria are (1) PPI-resistant esophageal-related symptoms, (2) EE-consistent endoscopic abnormalities, (3) infiltration of the esophageal mucosa with at least 24 eosinophils/high-power field (HPF), and (4) informed consent. We performed a chart review and included patients with persistent food impaction requiring bolus removal and those who experienced a “spontaneous” transmural esophageal rupture. In addition, a systematic review of the English language medical literature was performed with the key words “eosinophilic esophagitis” and “esophageal perforation” or “esophageal rupture.”

Results and Case Presentation

The SEED documents a total of 251 confirmed EE cases in adults and adolescents. During this 18-year period, 87 pa-
Patients (34.7%) experienced 1 or more (range, 1–5) persistent food impactions requiring endoscopic bolus removal. A total of 134 emergency endoscopies were undertaken to remove impacted food. Of these, 124 (92.5%) were performed with flexible gastrointestinal (GI) endoscopy under sedation and 10 (7.5%) by rigid ENT endoscopy under general anesthesia. The selection of the procedure depended exclusively on the local practices. All procedures proved successful in removing the bolus. However, in 2 of the 10 rigid procedures (20.0%), a transmural perforation occurred, whereas this severe complication did not arise in any of the patients treated with flexible endoscopy ($P < .0001$). The SEED data document 3 cases (1.2%) of transmural esophageal perforation or rupture. In addition to the 2 patients mentioned above where rigid endoscopy evoked perforation, a third patient suffered a “spontaneous” esophageal rupture occurring during severe vomiting, ie, Boerhaave’s syndrome. We present here further details of these 3 patients.

Case 1
A 17-year-old male patient had suffered from dysphagia since the age of 13 years, and at 15 years, EE was diagnosed and treated with topical corticosteroids. In July 2006, he experienced persistent food impaction necessitating emergency hospital admission. The on-call ENT surgeon removed the impacted bolus by using rigid esophagoscopy. Immediately afterwards, the patient had severe retrosternal and abdominal pain. A chest x-ray revealed a pneumoperitoneum and pneumomediastinum, and the suspected perforation was confirmed by a computed tomography scan. The patient recovered completely at the intensive care unit (ICU) under total parenteral nutrition and antibiotics.

Case 2
A 37-year-old nurse had suffered from dysphagia since she was 17. In July 2005, she experienced an episode of complete dysphagia. On referral to a local hospital, an ENT surgeon performed a rigid esophagoscopy, removing an impacted food bolus. Because of obvious postprocedural dissection (Figure 1), a gastrografin passage was performed, revealing a long esophageal dissection with leakage into the mediastinum (Figure 2). The patient recovered under antibiotic therapy and total parenteral nutrition. Two weeks after perforation, the esophageal gastrografin passage no longer showed the dissection. Three weeks later, an upper endoscopy was performed by a gastroenterologist, and EE was diagnosed.

Case 3
A 28-year-old woman with a 10-year history of dysphagia experienced an episode of gastroenteritis with severe vomiting and diarrhea. She was admitted to the hospital because of hematemesis, with acute and intensive thoracic and abdominal pain appearing after repeated retching. Before endoscopy, a computed tomography scan revealed a pneumomediastinum. An endoscopy showed a deep mucosal tear stretching 10 cm from the distal esophagus to the cardia. An attempt to close the tear with clips was not successful. The patient was operated on under the diagnosis of Boerhaave’s syndrome, and multiple mediastinal drainages were installed. Antibiotics were administered for 1 week, and the patient was parenterally nourished. After recovery, an endoscopy was performed, and EE was diagnosed.

Discussion and Conclusions
We report here on our 18-year experience with food impaction and esophageal perforation in 251 EE patients. In addition, we present 2 EE patients in whom the esophageal perforation was procedure-induced and 1 case in which rupture was associated with Boerhaave’s syndrome. The management of persistent food impaction with the potentially fatal complication of an esophageal breach has several important clinical and legal consequences.
Food impaction is a common complication in EE patients. More than one third of the SEED patients required emergency endoscopic bolus removal at some time during the course of their disease. Of note, from among the 87 patients experiencing food impaction, none was under sufficient anti-inflammatory therapy at the time of impaction. It is therefore tempting to speculate that this complication could be prevented by a consistent anti-eosinophilic therapy.

Endoscopic removal of an impacted bolus is the therapy of choice. Treatment for the 124 flexible removals was uneventful, whereas 20% (2/10) of those patients treated with rigid endoscopy experienced life-threatening complications. Despite the fact that perforations in EE might still occur even with flexible endoscopies, we strongly recommend that the impacted bolus be removed by using flexible endoscopy.

In all 3 patients experiencing a life-threatening transmural esophageal breach, the event occurred after a long-standing history of dysphagia, most probably indicative of a long history of EE (4, 19, and 10 years). This observation concurs with the literature, because most perforations have occurred with symptoms lasting many years. It is therefore presumable that esophageal perforation is a complication of long-standing EE that has led to esophageal remodeling. This assumption is further supported by the fact that to date, no young pediatric EE patients with perforations or ruptures have been reported. More data are needed to answer the question whether anti-inflammatory therapy, applied even in the absence of severe symptoms, could preserve esophageal structure and function.

Invasive diagnostic and therapeutic procedures are frequently required by EE patients. In general, these interventions are associated with a very small risk of esophageal perforation. For instance, in a series of 110,469 esophagogastroduodenoscopies (EGDs) performed in Germany under outpatient conditions, a perforation rate of 0.0009% was found, and in a Canadian university teaching hospital, a perforation rate of 6 per 10,000 procedures was documented. The total number of EGDs performed in EE patients is not known, but considering the limited number of published EE cases, it might be rather small. However, the fact that one transmural perforation has already occurred during a purely diagnostic endoscopy indicates that the risk for mucosal damage and even perforation is increased in EE patients. As a consequence, endoscopies in EE patients must be performed with extreme care, and dilations should be performed less aggressively than in patients with non-EE stenoses. Furthermore, it is mandatory to inform patients about the increased risk of an esophageal injury before performing an intervention in a patient with EE.

To date, 2 cases of EE patients experiencing spontaneous esophageal rupture during an episode of retching have been published. We add another typical Boerhaave’s syndrome patient. This retching complication should raise the suspicion of an underlying EE. We recommend therefore that after recovery from Boerhaave’s syndrome, an upper endoscopy should be performed for EE clarification.

All 7 patients with procedure-induced perforations were treated conservatively and recovered completely without any further complications. In contrast, all 3 patients with spontaneous esophageal ruptures required surgery. The number of EE patients with esophageal breaches is definitely too small to draw general therapeutic guidelines. However, we conclude that when procedure-induced perforations are recognized early, a conservative therapy can often successfully be implemented, thereby avoiding surgical measures. Spontaneous ruptures likely require more aggressive medical attention.

In summary, persistent food impaction is a common complication in patients with untreated EE, and the impacted bolus has to be removed by flexible and not by rigid endoscopy. Because the risk of procedure-induced injuries of the esophagus is likely increased in patients with EE, endoscopists should perform procedures extremely gently and less aggressively than in non-EE patients and incorporate this information in their medicolegal information practice. Furthermore, this is the third report describing an association between EE and Boerhaave’s syndrome, and patients with Boerhaave’s syndrome should therefore systematically be evaluated for underlying EE.

References