Intramural esophageal dissection is a rare disorder that should be considered in patients presenting with chest pain, dysphagia, and hematemesis. Although most commonly occurring in elderly women with impaired coagulation, esophageal dissection has also been observed in other demographics including in those with eosinophilic esophagitis. In our report, we present the case of a 19-year-old man who was found to have an intramural esophageal dissection in the setting of undiagnosed eosinophilic esophagitis. There have been multiple, proposed management strategies; however, we implemented a nonoperative approach and obtained successful results. Intramural esophageal dissection is an important diagnosis for thoracic surgeons to be aware of as these patients often present as surgical emergencies, but often do not require an acute surgical intervention.

Keywords: esophageal dissection, eosinophilic esophagitis, Mallory-Weiss tear

Introduction

Intramural esophageal dissection (also known as esophageal apoplexy, intramural hematoma of the esophagus, and intramural esophageal perforation) is an uncommon disorder characterized by a laceration between the mucosal and submucosal layers of the esophageal wall, without perforation. Patients diagnosed with intramural esophageal dissection are typically women in their seventh or eighth decade with known coagulopathy or who are taking anticoagulants. Presenting symptoms most commonly include chest pain, odynophagia and dysphagia, hematemesis; however, nausea, vomiting, and back pain have also been reported. The diagnosis is typically discovered by computed tomography (CT), upper esophagoscopy, or contrast esophagography. Conservative management of nil per os (NPO) and intravenous hydration is standard of care. Progression to perforation is exceedingly rare.

In our report, we present a case of an upper gastrointestinal bleed with intramural esophageal dissection in a 19-year-old man in the setting of undiagnosed eosinophilic esophagitis.

Case Report

A 19-year-old man with no prior medical problems presented to the emergency department with complaints of hematemesis and melena. Fourteen hours prior to presentation, the patient reported nausea and an episode of retching following dinner. The patient subsequently felt...
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fine and resumed his normal routine. Seven hours later, the patient reported several episodes of dark, black, tarry bowel movements. Another seven hours later, he reported an episode of hematemesis of a significant amount of bright red blood. On detailed review of history, the patient admitted to dysphagia over the last three years, but denied reflux, abdominal pain, or melena. The patient described a history of seasonal allergies, but denied symptoms consistent with asthma or atopic dermatitis. The remainder of his past medical history was unremarkable.

On physical examination, the patient was pale and diaphoretic with bloody vomitus on his shins. His vitals were stable. There was no subcutaneous crepitus appreciated in the neck or along the anterior chest wall. His abdominal examination revealed decreased bowel sounds, but no tenderness or distension was appreciated. A rectal exam was performed and was notable for black stool caking the perineum. A complete blood count (CBC) and coagulation profile were obtained, which were unremarkable.

Shortly following completion of the initial examination, the patient’s blood pressure dropped to 60 mmHg systolic and heart rate increased to 120 beats per second. Intravenous access was obtained and aggressive resuscitation with normal saline was begun. The patient’s pressure responded well and promptly returned to 110 mmHg systolic. A nasogastric tube was inserted. Lavage demonstrated a significant amount of bright red blood. The patient was immediately admitted to the intensive care unit and transfused with two units of packed red blood cells.

A chest X-ray was obtained and demonstrated pneumomediastinum with no evidence of pleural effusions (Fig. 1A). Urgent esophagogastroduodenoscopy (EGD) was then performed and revealed the presence of two

Fig. 1  Findings consistent with Mallory-Weiss tear and esophageal dissection: (A) Chest radiograph demonstrating pneumomediastinum (black arrows). (B) EGD revealing two mucosal tears superior to the gastroesophageal junction. The first tear (one arrow) was actively bleeding and repaired using three endoscopic clips. The second tear (two arrows) extended to the underlying muscularis propria and required no intervention. (C) Left Image: Non-contrast chest CT using lung windows demonstrating intraluminal air, “double barrel sign”, (black arrow) consistent with an intramural esophageal dissection. Right Image: Increased magnification of the mediastinum better demonstrating the esophageal intramural air and pneumomediastinum (white arrow).
Mallory-Weiss tears just superior to the gastroesophageal junction, one of which had a visible bleeding vessel (Fig. 1B). Three endoscopic clips were placed, and epinephrine was injected around the Mallory-Weiss tear with the visible vessel. During the procedure, the patient began to retch, and the non-bleeding tear visibly increased in size, but did not appear to extend beyond the submucosa. Additionally, the possibility of a corrugated esophagus (consistent with eosinophilic esophagitis) was noted; however, requisite full insufflation was not implemented to avoid injury worsening. Following EGD, a non-contrast CT of the chest revealed a “double barrel” sign consistent with intramural esophageal dissection (Fig. 1C). There was no evidence of frank perforation or pleural effusion. An esophagram was later performed using gastrografin followed by barium which confirmed the absence of an esophageal leak, thus ruling out the diagnosis of esophageal perforation. Of note, the presence of mediastinal emphysema was likely a result of decreased esophageal wall thickness which subsequently permitted diffusion of air into the mediastinum and pleural spaces.

The patient remained afebrile without evidence of disease progression for the remainder of his six day hospital stay. He was started on oral nutrition on admission day five and discharged the next day on a PPI and inhaled corticosteroids. On follow-up EGD, the presence of a corrugated esophagus was confirmed (Fig. 2A). Biopsies were taken at the distal esophagus which revealed greater than 15 eosinophils per high power field (Fig. 2B), thus confirming the diagnosis of eosinophilic esophagitis. The patient remained on inhaled corticosteroids and a proton pump inhibitor. He has been seen regularly in follow-up and is tolerating a normal diet without symptoms of nausea, vomiting, dysphagia, chest pain, melena, hematemesis, or retching. He remains asymptomatic at six months from the time of presentation.

**Discussion**

Eosinophilic esophagitis is an isolated, eosinophilic inflammation of the superficial layers of the esophagus. Common symptoms include dysphagia, food impaction, heartburn and history of atopia. The disease is most commonly diagnosed in young Caucasian men, and the prevalence within the general population is 0.4% and is found in 6.5% of those undergoing upper endoscopy for abnormal gastrointestinal symptomatology. Typical findings on endoscopy include mucosal fragility/edema, rings, strictures, whitish pinpoint exudates and a small-caliber esophagus. Patients are also commonly diagnosed with asthma, allergy, and atopic dermatitis. Management options for eosinophilic esophagitis involve diet restriction, medical management (systemic or inhaled corticosteroids, leukotriene receptor antagonists, proton pump inhibitors, and monoclonal antibodies), and endoscopic dilation when necessary. Most patients respond well to treatment, and do not develop life threatening complications. Although uncommon, it is hypothesized that chronic eosinophilic infiltration may result in a friable esophageal mucosa and fragile wall which allows superficial lacerations to secondarily propagate into deeper esophageal layers resulting in dissection.

First described by Marks and Keet in 1968, there have...
been approximately 50 reported cases of intramural esophageal dissection. The diagnosis is part of the spectrum of esophageal injuries which includes a Mallory-Weiss tear and Boerhaave’s syndrome. Patients are typically women in their seventh or eighth decade with known coagulopathy or who are on anticoagulant therapy. Our patient, however, was an otherwise healthy young man with no identifiable bleeding risk factors. Patients presenting with acute esophageal dissection most commonly describe retrosternal pain (83%), hematemesis (71%), odynophagia and dysphagia (32%). This clinical triad, however, is only observed in 35% of patients. Although our patient required a blood transfusion for hemodynamic instability, blood loss is uncommon and less than 10% of patients with the diagnosis require a transfusion. Other well-documented symptoms include nausea, vomiting, and back pain.

The diagnosis of intramural esophageal dissection can be made using multiple modalities including contrast esophagography, upper GI endoscopy, endoscopic ultrasound or CT scan. In the absence of substantial active bleeding, we advocate the use of an esophagram or CT over more invasive techniques (such as endoscopy or ultrasound) as to minimize potential risks of process extension or perforation. Characteristic findings on barium esophagram and CT scan are a “double-barreled” esophagus and thickening of the esophageal wall with variable degrees of obliteration of the lumen, respectively. In the case of frank bleeding, such as in the report described, the use of endoscopy should be implemented as it also allows for diagnosis with simultaneous intervention. On upper GI endoscopy, typical characteristics include a bulging, purplish lesion with a smooth, normal overlying mucosa.

Nonoperative management of intramural esophageal dissection is well supported as perforation is exceedingly rare. A strict regimen of nil per os with intravenous fluids and nutrition is recommended. Symptoms resolve rapidly, and most patients can advance to oral intake within a few days. The intramural hematoma typically resolves within 1 to 3 weeks, and complete healing of the mucosa and full recovery of esophageal function should be expected. In the rare case of persistent hematoma, further intervention may be needed. The creation of a septum between the true and false lumen using a needle-type diathermy knife has been documented in this circumstance with excellent results. In the scenario of a circumferential dissection, a variant, which is less amenable to conservative treatment alone, the placement of an expandable metal stent has been described.

Upon further review of the literature, we identified two recent reports linking eosinophilic esophagitis to esophageal dissections. In both reports, the dissection was circumferential and associated with complications: perforation in one report and abscess formation in the other. To our knowledge, this is the first documented case of a non-circumferential esophageal dissection observed in a patient with eosinophilic esophagitis. Although unclear, it is most likely that friable mucosa and an unstable esophageal wall may place patients with eosinophilic esophagitis at increased risk for esophageal dissection during times of acute esophageal stress.

Conclusion

Intramural esophageal dissection is a rare disorder that should be considered in patients presenting with chest pain, dysphagia, and hematemesis. Although most commonly occurring in elderly women with impaired coagulation, esophageal dissection has been observed in young males with co-morbid eosinophilic esophagitis. It is likely that the friable mucosa and fragile esophageal wall predispose patients with eosinophilic esophagitis to esophageal dissection. Thoracic surgeons are often involved in the acute management of these patients and should appreciate that full recovery with conservative measures is the norm. In rare instances, additional interventions have been utilized with favorable long-term outcomes being observed.

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References