

Recurrent Spontaneous Esophageal Dissection

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ABSTRACT

Introduction: Spontaneous esophageal dissection is a rare disorder of the esophagus.

Case Description: We present what is believed to be the first reported case of recurrent esophageal dissection in a previously healthy 33-year-old man with chronic eosinophilic esophagitis. He had two episodes of spontaneous dissection of the midesophagus separated by a 5-month interval. Both episodes responded to treatment with endoscopic intervention. He has remained free of additional recurrences after definitive endoscopic therapy and oral steroid therapy. A complete description of the case, relevant radiologic imaging, and a review of the relevant literature are provided.

Discussion: Endoscopic therapy is an option for the management of recurrent esophageal dissection.

Key Words: Endoscopy, Esophageal dissection, Spontaneous, Recurrent, Case report.

INTRODUCTION

A 33-year-old man with a past medical history for esophageal ulcers and previous orthopedic surgeries presented with a 1-week history of substernal chest pain, dysphagia, fatigue, and subjective fevers. He was seen as an outpatient and given famotidine and sucralfate pending endoscopy. Because of his worsening symptoms, he presented to an outside hospital and was transferred to our facility after a computed tomography (CT) scan of his chest showed intramural esophageal dissection (**Figure 1**). He denied any relevant family medical history or the use of tobacco, alcohol, or other illicit drugs. The patient reported a similar episode approximately 15 years previously, underwent endoscopic evaluation, and was diagnosed as having esophageal ulcers. He was prescribed sucralfate during that presentation and failed to show for follow-up evaluation. He had 3 further episodes of dysphagia over the subsequent 6-year period and did well until his presentation to our facility.

Esophagogastroduodenoscopy (EGD) was performed. The distal esophagus was narrowed, and purulent material was noted to flow from the esophageal mucosa at approximately 35 cm (gastroesophageal junction, 40 cm) (**Figure 2**). Immediately after the procedure, he experienced prompt relief of his substernal chest pain and dysphagia. The aspirated sample of the drainage grew group C *Streptococcus* bacteria. The patient was maintained on bowel rest and placed on intravenous piperacillin/tazobactam (3.375 g every 6 hours) for 7 days. After an upper gastrointestinal study done before discharge noted no significant abnormalities, he was sent home with amoxicillin/clavulanate (250/125 mg every 8 hours) for 10 days. The patient was started on a soft diet on postprocedure day 5, with instructions to continue this for 6 weeks, and then to transition to a regular diet.

Approximately 5 months later, he represented with a 1-week history of sudden-onset substernal chest pain, odynophagia, and subjective fevers. One day before presentation, he underwent endoscopy, which showed findings of eosinophilic esophagitis (**Figure 3A**). Given the apparent extrinsic esophageal compression at 25 to 27 cm, the scope was unable to be advanced and the procedure was terminated. At presentation to the hospital, he was

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DOI: 10.4293/108680813X13693422518678

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Figure 1. CT chest scan demonstrating intramural dissection of the esophagus. Sections demonstrate intramural dissection of the esophagus. Lucency within the “true” esophageal lumen indicates a feeding tube.

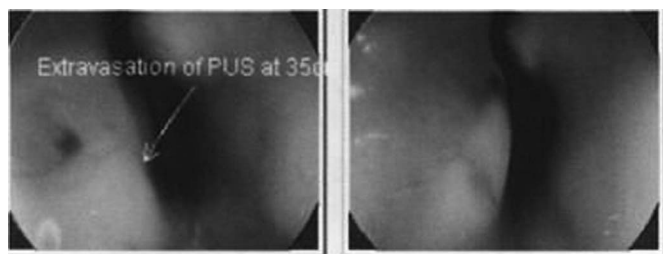


Figure 2. EGD performed during the patient's first hospitalization. Active extravasation of purulent fluid was noted on endoscopy (arrow).

afebrile, with low-grade tachycardia but otherwise normal vital signs. His laboratory test results revealed leukocytosis (white blood cell count 16.9 g/dL) but were otherwise unremarkable. An upright chest radiograph revealed no significant abnormalities. A subsequent CT chest scan demonstrated findings consistent with esophageal dissection versus intramural abscess. An upper gastrointestinal series of tests demonstrated intramural tracking of contrast through an area in the anterior proximal esophagus.

Shortly after endoscopy, and similar to his hospitalization several months earlier, the patient experienced dramatic relief of his pain and dysphagia, with expulsion of foul-smelling purulent mucus. Before discharge, a repeat endoscopy was performed, which demonstrated a markedly improved appearance of the esophagus. The lumen was noted to be patent, with significantly decreased macroscopic findings of eosinophilic esophagitis. A pinhole defect, thought to be the point of dissection, was noted and

closed with a single endoclip (**Figure 3B**). Repeat CT scan at approximately 1 and 6 months postprocedure demonstrated a normal appearance of the esophagus, with no evidence of recurrent dissection.

DISCUSSION

First described in 1957, spontaneous esophageal dissection was identified as a disruption in the submucosal space of the esophagus, allowing an intramural hematoma to form within it, compressing the lumen, and subsequently dissecting the submucosal plane from the underlying muscle.^{1,2} There are fewer than 50 reported cases. This disorder occurs most frequently in women in the sixth or seventh decade of life; however, it has been reported in younger male patients.³ Possible causes may include external trauma, foreign body entrapment, instrumentation, persistent retching, esophageal stricture, esophageal diverticula, and arteriovenous malformation.³ The clinical history, laboratory data, and imaging findings of this patient did not include any of the first 3 etiologies just listed.

It is important to differentiate submucosal esophageal dissection from other disorders that may present in similar fashion, such as Mallory-Weiss syndrome, esophageal perforation, or dissecting aneurysm (all of which require surgical intervention).^{3,4} The triad of symptoms for this disorder includes retrosternal pain (often initiated by the swallowing of food), mild hematemesis, and odynophagia.⁵ Clinical examination may also reveal a low-grade fever and the absence of surgical emphysema in the neck.⁴

When intramural dissection is suspected, the best initial diagnostic test is a barium swallow, which is less invasive than endoscopy and may identify whether the esophagus is intact.² The dissection of the intramural hematoma into the submucosa will give the radiographic appearance of a “double-barreled” lumen (diagnostic for this condition).^{1,5} Endoscopy may reveal narrowing or compression of the lumen, making the procedure difficult to perform. Prognosis of this condition is greatly improved by restricting oral intake and initiating intravenous fluid hydration and administering antacids.^{2,3} Surgical or endoscopic treatment should be reserved for patients with defects that do not resolve or for complications such as transmural perforation with mediastinitis or mucosal flap formation.⁶

Endoscopic therapy offers a unique approach to treatment and has been previously described. In patients in whom endoscopic mucosal resection is performed, iatrogenic

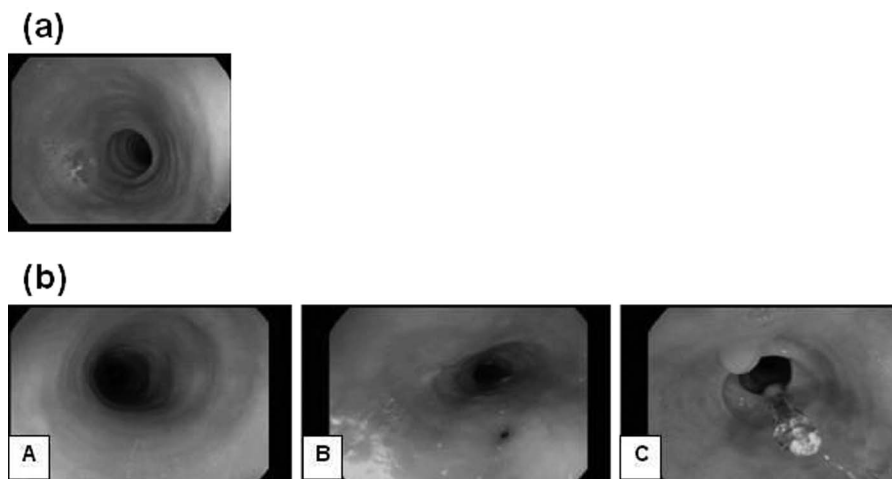


Figure 3. A, EGD 1 day before hospital admission. Note the tracheal pattern of the esophagus, which is suggestive of eosinophilic esophagitis. B, EGD before hospital discharge. Note the significantly decreased trachealization of the esophagus (a). A pinhole defect was noted in the cervical esophagus; pre (b) and post endoclip placement (c).

perforations have occurred and been successfully repaired using endoclips. Although the etiology of the esophageal defect in our patient was different, an endoclip was used successfully.

One case reported a patient with false esophageal lumen. In this instance, endoscopic incision of the long septum was performed using a needle-knife papillotome without complications.⁷ Another case described a patient with a circumferential esophageal dissection who underwent endoscopic incision of the septum and transection of the true internal esophageal wall. A resultant stricture required balloon dilatation/stent placement.⁶ Gluck et al⁸ reported two patients with intramural dissection, where therapeutic balloon dilatation was performed for the strictured esophagus after spontaneous healing had occurred.

On this patient's initial presentation, chest CT scan revealed the characteristic circumferential esophageal dissection. The initial EGD revealed a narrowed distal esophagus, with purulent material identified from the esophageal mucosa at approximately 35 cm. We do not believe that this purulence was simply an esophageal abscess because there was no history of preceding trauma, foreign body ingestion, or explosive vomiting. It is more likely that an infection resulted from luminal bacterial transmural migration or microperforation and hence presented as a submucosal abscess during endoscopy. Once the devitalized mucosa overlying the lower third of the thoracic esophagus was penetrated by the endoscope, the purulence was released and the patient had dramatic improvement of his presenting chest pain,

defervescence, and improvement in his general sense of well-being.

Although spontaneous dissection has been well-documented, the recurrence in this patient is what makes his presentation quite interesting and unique. As has been documented in prior cases,^{9–11} the pathogenesis of this esophageal dissection was eosinophilic esophagitis, documented by initial endoscopic biopsy. It responded only partially to inhaled corticosteroids, which likely predisposed the patient to the additional episode of dissection in the upper thoracic esophagus.

Our patient responded well with the aforementioned conservative management and endoscopic therapy, and repeat CT imaging at 1 and 6 months demonstrated a normal esophageal appearance. The clinical presentation, imaging, and endoscopic findings were consistent with previous reports of spontaneous esophageal dissection. Other reports of esophageal dissection exist describing the spontaneous nature of the disease, diagnostic studies, and treatment modalities, all similar to this patient's case.^{12–14} We believe, however, that this is the only report of "recurrent" spontaneous esophageal dissection found in the English literature that was treated successfully by endoscopic and endoclip therapy.

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