Intramural Esophageal Dissection as an Esophageal Emergency

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ABSTRACT

Esophageal laceration with intramural dissection as an esophageal emergency is rare. At presentation, it is difficult to differentiate this disorder from esophageal perforation. We present the case of a 50-year-old Taiwanese man who was admitted to our institution with symptoms of progressive chest pain and odynophagia for two weeks. Esophagoscopy revealed a deep longitudinal laceration with hemorrhage in the esophagus. Computed tomography of the chest revealed intramural esophageal dissection. An esophagogram showed the absence of esophageal perforation. After supportive treatment with total parenteral nutrition for two weeks, the symptoms resolved, and follow-up esophagoscopy revealed a healed mucosal laceration and disappearance of intramural dissection of the esophagus.

Keywords: esophagus, dissection, barium, CT

INTRODUCTION

Intramural esophageal dissection and intramural hematoma of the esophagus are rare and are usually referred to as submucosal dissection or intramural rupture, which may be considered intermediate forms of esophageal injury and are managed conservatively [1, 2]. On the contrary, esophageal transmural perforation is often a surgical emergency, and can lead to sepsis, multiple organ failure or even death [3]. Symptoms of intramural esophageal dissection include retrosternal chest pain, dysphagia or odynophagia, and hematemesis; however, nausea, vomiting and backache may also be present [1, 4, 5]. A prompt diagnosis is extremely important. In this report, we present a case of chest pain due to intramural esophageal dissection in a 50-year-old man who has liver cirrhosis with a bleeding tendency, and we focus on the etiology and radiologic features of this rare entity.

CASE REPORT

A 50-year-old Taiwanese man presented to the emergency department with the initial symptom of a stinging sensation in the chest during swallowing for two weeks, which progressed to chest pain and odynophagia. His medical history included liver cirrhosis, Child-Pugh class B and hepatitis B carrier status. One episode of tarry stool and tea-colored urine occurred two days prior to presentation. Icteric sclera was noted during physical examination. The laboratory results showed a bleeding tendency with a low platelet count (50×10^9 per liter) and a prolonged prothrombin time (prothrombin time 18.2 seconds and control 10.6 seconds). Urgent upper gastrointestinal endoscopy showed esophageal mucosal laceration with hemorrhage and an entry of false lumen of intramural esophageal dissection (Fig. 1a). An axial image from a contrast-enhanced chest computed tomography (CT) scan showed the false lumen of esophageal dissection located anterior to the true lumen of the esophagus (Fig. 2a). A sagittal maximum intensity projection image from the contrast-enhanced chest CT scan clearly depicted a characteristic double-barreled esophagus and thin mucosal flap, findings indicative of intramural esophageal dissection (Fig. 2b). An esophagogram showed the absence of esophageal transmural perforation (Fig. 3). The symptoms relieved after conservative treatment with total parenteral nutrition for two weeks. Three weeks later,
healed mucosal laceration and disappearance of intramural dissection of the esophagus were observed by esophagoscopy (Fig. 1b).

DISCUSSION

Intramural esophageal dissection is a relatively rare clinical entity with a concept similar to vascular dissection, which is characterized by a mucosal tear and formation of true and false lumens in the esophagus, with or without a re-entry point. First described by Williams in 1957, this condition has been referred to by several terms, including esophageal apoplexy, intramural esophageal rupture, submucosal hematoma, and intramucosal esophageal dissection [6]. Due to the descriptive nature, Phan and Heitmiller [7] advocated usage of the term intramural esophageal dissection. The clinical trial of intramural esophageal dissection

Figure 1

1a 1b

Figure 1. a. Endoscopic image demonstrated two lumens including the true esophageal lumen (double arrow) and the false lumen (arrow), separated by a mucosal bridge in proximal portion of the esophagus. b. Follow up endoscopic study after two weeks revealed healed mucosal laceration and intramural dissection of the esophagus.

Figure 2

2a 2b

Figure 2. a. Axial image from contrast-enhanced chest computed tomography scan showed the false lumen (arrow) of esophageal dissection located anterior to the true lumen (double white arrow) of the esophagus. b. Sagittal maximum intensity projection image from contrast-enhanced chest computed tomography scan clearly depicted a characteristic double-barreled esophagus with false lumen (white arrow) and mucosal flap (double white arrow), a finding indicative of intramural esophageal dissection.
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dissection includes retrosternal pain (83%), hematemesis (71%), and odynophagia and dysphagia (32%), although other well-documented symptoms such as nausea, vomiting, and back pain have been reported [8]. The pattern of pain is gradual in onset, exacerbated by swallowing, and may become intense, localized to the retrosternal or epigastric region, and infrequently in the back.

The etiology of intramural esophageal dissection may be primary mucosal injury with secondary intramural dissection. Alternatively, it may be primary esophageal intramural dissection with secondary rupture into the esophageal lumen. This disease entity has mainly been found in elderly females with coagulopathy or under anticoagulation therapy [9]. However, intramural esophageal dissection has been found in a wide range of patients, including healthy young adults [10, 11]. Intramural esophageal dissection is associated with mucosal trauma during eating, external trauma, instrumentation, persistent retching, esophageal stricture, esophageal diverticula, eosinophilic esophagitis, pharyngeal abscess, and arteriovenous malformation [10, 12-15]. Shay and Berendson [16] postulated that submucosal bleeding with intramural dissection initially occurs, then the hematoma remains contained or may secondarily rupture through the mucosa or muscularis. A propensity for intramural esophageal dissection to occur in patients with abnormal hemostasis supports this theory. In our patient, the bleeding tendency due to his underlying liver cirrhosis may have played a key role in the pathogenesis.

Differentiation of intramural esophageal dissection from other disorders, such as Boerhaave syndrome, Mallory-Weiss syndrome, and esophageal perforation, is difficult without image study. These disorders may be similar in terms of clinical presentation and may require surgical intervention. Boerhaave syndrome is characterized

Figure 3

Figure 3. Esophagogram with oblique and anteroposterior views of the thoracic esophagus. a. The oblique view demonstrated a triangular outpouching lesion (arrow) in anteriolateral wall of midesophagus, suggest inlet of intramural dissection of esophagus, without evidence of contrast extravasation. b. The anteroposterior view showed a double-barreled esophagus with a false lumen (white arrow) and a true lumen (black arrow). This appearance is caused by the barium that collects in the false lumen between the dissected esophageal mucosa and surrounding esophageal muscle.
by severe chest pain followed by subcutaneous emphysema, pneumomediastinum and sepsis, resulting from transmural rupture of the distal esophagus due to a sudden and massive increase in intraesophageal pressure. A Mallory–Weiss tear is defined as laceration of the venous plexus at the gastroesophageal junction, which manifests as hematemesis as a result of a sudden increase in pressure during vomiting together with regurgitation of corrosive gastric contents. Esophageal perforation can range in severity from air leakage only to contained perforation with self-drainage, free mediastinal leakage, or free leakage with extension into the pleural space [2].

Chest radiographs do not usually disclose relevant abnormality, but occasionally, a large hematoma due to intramural esophageal dissection can be seen on plain films as mediastinal widening, whereas hemothorax, pneumothorax, or pneumomediastinum can be demonstrated, which suggest esophageal perforation. Owing to the invasive and less readily available nature of upper gastrointestinal endoscopy, we advocate the use of esophagography or CT if active bleeding is absent, so as to minimize the potential risks of enlarging a small laceration or producing frank perforation. Esophagography provides an effective way in which to identify the type of esophageal perforation, such as air leakage only, without contrast extravasation, contained perforation, or free, noncontained perforation. The former can be observed and resolves spontaneously, and the latter can be managed nonoperatively. However, the most serious cases should be aggressively managed, preferably by primary repair [17]. Considering the fact that in the vast proportion of cases of intramural esophageal dissection, there is a low prevalence of perforation, patients can usually be managed nonoperatively, with complete resolution within weeks [18].

CT is non-invasive, readily available, and useful for differentiating esophageal emergencies from other thoracic disorders. As the majority of these patients are referred for chest pain or vascular abnormalities evaluation, an intravenous (IV) contrast medium is often used. If esophageal perforation is considered, oral contrast (Omnipaque 240mgI/ml, GE Healthcare) can be administered, typically 50 ml of iodinated contrast medium [4]. CT has a more sensitive ability to determine the extent of pneumomediastinum than esophagography. Intramural esophageal dissection tends to be found posterior to the true lumen of the esophagus, with a characteristic double-barreled esophageus appearance, and multiplanar reformatted images are ideal and often provide better information regarding the extent of the disease [1]. In magnetic resonance imaging, intramural esophageal dissection typical characteristics include double rim enhancement over the mucosa and the muscular layers of the esophagus or a lesion of intermediate signal with scattered areas of high signal intensity on T1- and T2-weighted images along the esophagus if a hematoma is present [19]. Magnetic resonance imaging is not recommended as a routine study due to the significantly lower ready availability in most facilities.

Intramural esophageal dissection is a rare entity that should be considered in patients presenting with retrosternal pain, hematemesis, odynophagia and dysphagia. It is important for clinicians and radiologists to understand the clinical, pathological, and radiological presentations of intramural esophageal dissection and the diagnostic tools that can be utilized in order to obtain a prompt correct diagnosis.

REFERENCES

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