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Recurrent, Spontaneous Esophageal Ruptures Associated With Antiphospholipid Antibody Syndrome: Report of a Case

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A 52-year-old man was admitted to our hospital with a spontaneous esophageal rupture (Boerhaave syndrome) and was successfully treated. Eight years after the first incident, he was readmitted with a recurrent rupture. Recurrence of Boerhaave syndrome is extremely rare, with only 7 cases reported in the English literature. During treatment, the patient was also diagnosed with antiphospholipid syndrome (APS). Although APS is known to cause a variety of symptoms due to vascular thrombosis, recurrence of Boerhaave syndrome, coincident with APS, has never been reported. The pathogenesis of Boerhaave syndrome has not been clearly determined. This report serves to increase awareness of the risk of APS, which results in an increased risk of spontaneous rupture of the esophagus.

Key words: Boerhaave syndrome – Esophageal rupture – Antiphospholipid syndrome

Spontaneous rupture of the esophagus (Boerhaave syndrome) is a relatively rare entity among digestive tract emergencies.[1] Because Boerhaave syndrome has a high rate of mortality, few reports describe the late complications of the syndrome. Further, recurrence of the syndrome is extremely rare, with only 7 cases having been reported in the English literature.[2–7] In addition, the pathophysiologic mechanisms responsible for Boerhaave syndrome have not been clearly determined.

Antiphospholipid syndrome (APS) is a type of autoimmune syndrome and is known to cause a

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variety of complications, which are primarily caused by vascular disorders. In the present report we describe a case of recurrent Boerhaave syndrome associated with APS, and we discuss the pathogenesis of spontaneous esophageal ruptures.

Case Report

A 52-year-old man was admitted to our hospital with shock and severe chest pain. Before admission, the patient had experienced frequent vomiting and diarrhea for 3 days. Computed tomography (CT) indicated the presence of pneumothorax associated with pneumonia and pleural effusion of the right lung (Fig. 1). An esophagography indicated the leakage of contrast medium from the lower esophagus into the right pleural cavity (Fig. 2). As a result, the patient was diagnosed with a spontaneous rupture of the esophagus.

An emergent right thoracotomy was performed, and a 5-cm–long tear was observed in the mediastinal pleura; the tear was suspected of communicating with the esophagus. A thoracic drain was placed after a pleural lavage was performed, without closure of the ruptured esophagus because of the presence of severe inflammation. After the operation, the patient was successfully treated and discharged on day 56 of hospitalization. Thereafter, the patient's upper gastrointestinal tract was endoscopically examined annually. The presence of an esophageal hiatus hernia and reflux esophagitis, with severe scarring of the esophageal mucosa, was observed (Fig. 3); however, the patient was asymptomatic.

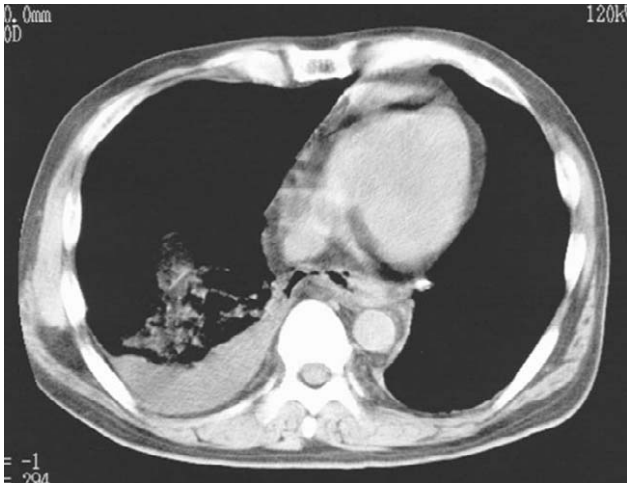

Fig. 1 CT scan showing the pneumothorax associated with pneumonia and pleural effusion of the right lung.

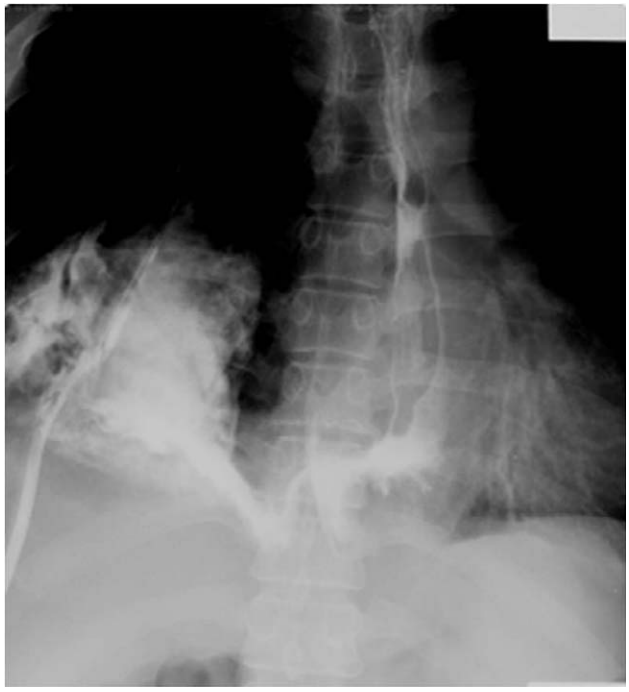

Fig. 2 Esophagography image indicating the leakage of contrast medium from the lower esophagus into the right pleural cavity.

Eight years after the first rupture, the patient was readmitted to our hospital with hematemesis, following severe vomiting. A CT scan showed the presence of a mediastinal abscess associated with pneumomediastinum and subcutaneous emphysema (Fig. 4). An esophagographic examination indicated the communication of the esophagus with the abscess. Thus, we diagnosed the patient with a recurrent, spontaneous rupture of the esophagus. The patient was not treated surgically because the leakage was limited to the abscess and the general condition of the patient was fair. On day 18 of hospitalization, central vein catheter thrombosis was detected, and the serum anticardiolipin antibody and lupus anticoagulant levels were elevated. The patient was diagnosed with APS. However, he did not present with systemic lupus erythematosus (SLE), which is a well-known complication of APS, according to the criteria of the American College of Rheumatology. With anticoagulation therapy, the thrombosis disappeared and the esophageal rupture was closed conservatively. The patient was discharged on day 107 of hospitalization.

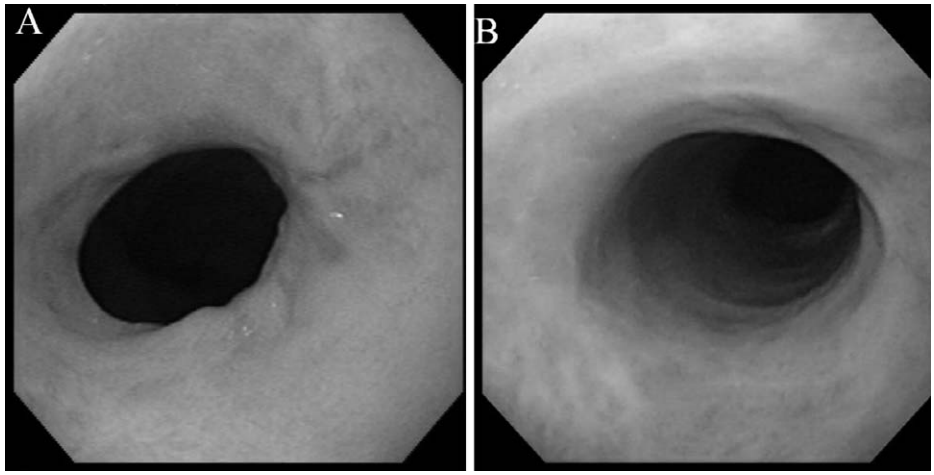

Fig. 3 Endoscopic images obtained 1 year (A) and 4 years (B) after the first perforation, indicating a hiatus hernia and reflux esophagitis, with severe scarring.

Discussion

Recurrent ruptures of the esophagus are extremely rare, with only 8 cases, including the present case, being reported in the English literature.[2–7] One case involved a 17-year-old female patient, and the remainder of the cases involved male patients ranging in age from 45 to 66 years. In one of these cases, the rupture occurred at the level of the midesophagus, whereas the other 7 patients experienced perforations of the lower esophagus. The interval between the ruptures ranged from 6 months to 30 years.

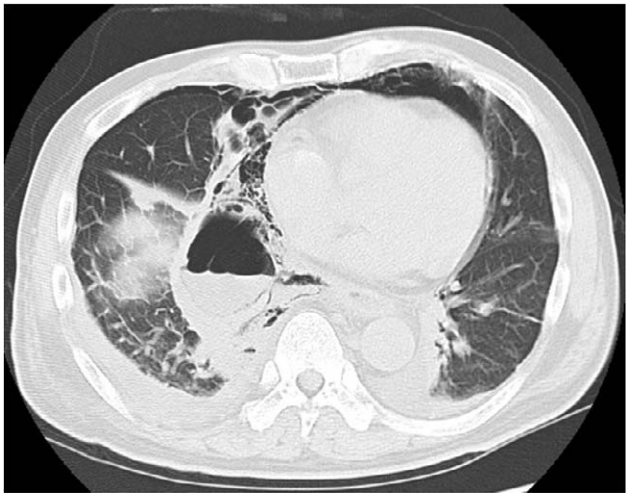

Fig. 4 CT scan showing a mediastinal abscess associated with pneumomediastinum and subcutaneous emphysema.

Boerhaave syndrome is believed to develop acutely as a result of intraesophageal pressure, such as that caused by vomiting. However, a comparison of the incidence of severe vomiting, or other conditions that cause intramural esophageal pressure, suggests that the incidence of Boerhaave syndrome is extremely low. Therefore, other conditions must also exist to cause the spontaneous rupture of the esophagus. However, weakness of the esophageal wall has been suggested to contribute to Boerhaave syndrome.[1,8,9] Korn *et al*[8] studied the weakness of the esophageal wall anatomically and experimentally with fresh human cadaver tissue and concluded that the connective tissue at the junction between the clasp and oblique fibers appears to constitute a weak point in the lower esophagus. Kuwano *et al*[9] described 2 cases of Boerhaave syndrome where the patient lacked a muscularis mucosa, and the authors suggested that the absence of this structure in the esophageal wall may have contributed to the spontaneous rupture of the esophagus. Esophagitis, due to reflux, alcohol, eosinophilia, and mixed connective tissue syndrome all the causes of esophagitis mentioned here have also been reported to be factors contributing to Boerhaave syndrome.[10,11]

Our patient was also diagnosed with APS—an autoimmune syndrome characterized by the synthesis of antiphospholipid antibodies, with vascular thrombosis.[12] Because the clinical complications of APS are mainly caused by vascular disorders, a wide variety of symptoms are known, and a previous report has described a case of acute

necrosis and esophageal perforation related to APS.[13] APS is also known to be highly coincident with SLE. The present patient was not diagnosed with SLE, but the levels of some of the serum markers for SLE, such as lupus anticoagulant, were noted to be elevated. SLE is one of the well-known conditions that cause esophagitis, and, as mentioned above, esophagitis that decreases the motility and strength of the esophageal wall may have an important role in the spontaneous rupture of the esophagus.[14] Thus, although we could not indicate the presence of a relationship between Boerhaave syndrome and APS, with or without SLE, this case indicates that the fragility of the esophageal wall, caused by APS, may increase the risk of spontaneous esophageal rupture.

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