A Patient with Vigorous Achalasia and Rapid Enlargement of an Epiphrenic Esophageal Diverticulum

Yasuhiro Tamura¹, Yasushi Funaki¹, Kazuori Adachi¹, Hisatsugu Noda¹, Shinya Izawa¹, Akihito Iida¹, Naotaka Ogawara¹, Masahiko Miyaji³, Makoto Sasaki² and Kunio Kasugai¹

Abstract

A 47-year-old man was found to have a 3-cm epiphrenic esophageal diverticulum on an upper gastrointestinal (UGI) barium study. He developed the symptoms of heartburn approximately 12 months later. UGI endoscopy indicated non-erosive gastroesophageal reflux disease (NERD) and an epiphrenic esophageal diverticulum. A proton pump inhibitor (PPI) did not relieve the symptoms. An UGI barium study at that time showed that the epiphrenic esophageal diverticulum had enlarged to 7 cm, and esophageal manometry showed findings of achalasia and diffuse esophageal spasm (DES), thus vigorous achalasia was diagnosed. Resection of the epiphrenic esophageal diverticulum, myotomy, and fundoplication (the Heller-Dor procedure) were successfully performed and no postoperative symptoms were encountered.

Key words: diffuse esophageal spasm, giant epiphrenic esophageal diverticulum, vigorous achalasia


Introduction

Vigorous achalasia, first reported by Sanderson et al. in 1967, is an esophageal motility disorder characterized by both non-peristaltic simultaneous contractions, as seen in diffuse esophageal spasm (DES), and the incomplete relaxation of the lower esophageal sphincter (LES), as seen in achalasia (1). A diagnosis of epiphrenic esophageal diverticula based on an X-ray barium contrast examination is extremely rare (2). We herein report the case of a patient with a rapidly enlarging epiphrenic esophageal diverticulum which was found to be associated with vigorous achalasia.

Case Report

A 47-year-old man without previously systemic diseases, such as diabetes mellitus, scleroderma or other neurodegenerative disease which would affect the esophagus motility, was found to have an epiphrenic esophageal diverticulum measuring 3.0×2.0 cm on an upper gastrointestinal (GI) barium study during a routine medical examination (Fig. 1A); however, it was asymptomatic and left untreated. Twelve months later, the patient began experiencing heartburn after eating, and his symptoms of heartburn and dysphagia worsened. After an evaluation by a local physician, upper GI endoscopy showed non-erosive esophagitis and an epiphrenic esophageal diverticulum, and treatment with a proton pump inhibitor (PPI) was thus prescribed. However, the symptoms of reflux did not improve with oral PPI, thus the patient was referred and admitted to our hospital for further evaluation and treatment. An upper GI barium study at that time showed a 7.0×7.0 cm diverticulum extending into the right thoracic cavity from the lower esophagus (Fig. 1B) and the classic “bird-beak” appearance in the lower esophagus (Fig. 1C). Upper GI endoscopy showed a diverticulum with a wide orifice in the right esophageal wall approximately 35 cm from the incisors (Fig. 2). Chest computed tomography (CT) showed food residue and an air-fluid level near the right diaphragm, but no thickening of the esophageal wall (Fig. 3). Esophageal manometry showed the median pressure of the lower esophageal sphincter (LES) increased to 43.2 mmHg, and the incomplete relaxation of LES with high contraction waves of 180 mmHg or more
was observed at water deglutition. The primary peristaltic wave in the middle and lower part of the esophagus indicated high-amplitude (80-270 mmHg) non-peristaltic simultaneous contractions lasting ≥10 seconds (Fig. 4). According to the results of esophageal imaging and manometry, an epiphrenic esophageal diverticulum associated with vigorous achalasia belonging to Type III of achalasia according to the Chicago classification was diagnosed (3). Because the patient had a giant diverticulum measuring ≥5 cm in diameter and a deteriorated gastroesophageal reflux symptom over a short period of time, resection of the diverticulum, myotomy, and Dor fundoplication to prevent reflux were performed by our hospital’s Department of Surgery. The oral intake was resumed on postoperative day 5, the preoperative reflux symptoms were resolved, and the patient was discharged on day 13. The postoperative histopathology showed a true diverticulum with mild muscle layer atrophy in the diverticulum wall (Fig. 5).

Discussion

The etiology of epiphrenic esophageal diverticula involves an increased lower esophageal pressure and congenital weakness of the esophageal wall (4, 5). Pulsion pseudodiverticula due to a lower esophageal hypermotility and increased LES pressure, together with esophageal hiatal hernias and esophageal dysmotility, such as esophageal spasm and achalasia, are known to occur (6, 7). Preoperative manometry and 24-h esophageal/gastric pH monitoring have also been recommended (8). In the present patient, esophageal motility disorder was not diagnosed based on an upper GI barium study or upper GI endoscopy; rather, esophageal...
manometry was required for the differential diagnosis.

Vigorous achalasia, first described by Sanderson et al. in 1967, is an esophageal motility disorder characterized by both the incomplete relaxation of the LES, as seen in achalasia, and non-peristaltic esophageal contractions, as seen in DES (1). To the best of our knowledge, a coexisting giant true epiphrenic diverticulum, as seen in our patient, has not been previously reported. Compared to the clinical symptoms in classical achalasia, some reports have showed that vigorous achalasia may also present as dysphagia, vomiting, heartburn, and chest pain, along with a higher frequency of chest pain (1, 9); however, other reports have found no difference between classical and vigorous achalasia (10, 11). Esophageal barium imaging in vigorous achalasia shows a dilated esophagus and abnormal spasm. Sanderson et al. (1) reported that approximately 46% of patients with moderately or severely diffuse dilatation of the esophagus have vigorous achalasia. The present patient mainly complained of heartburn, with no other typical clinical symptoms of dysphagia or chest pain consistent with achalasia. Esophageal imaging showed a bird-beak appearance, which is a specific finding of achalasia, with no additional findings consistent with achalasia or DES. However, esophageal manometry while swallowing water showed the incomplete relaxation of the LES, an increased LES pressure, and high-amplitude non-peristaltic simultaneous contractions, which are characteristic of both achalasia and DES. Furthermore, our patient had no other systemic disease associated with esophageal dysmotility, thus vigorous achalasia was diagnosed.

Pharmacotherapy, such as isosorbide dinitrate (ISDN) (4) or nifedipine (12), may be effective for vigorous and classical achalasia, but pneumatic balloon dilation of the cardi (13) and other surgical treatments (1, 14) may be more effective in some cases. However, the present patient had a giant epiphrenic esophageal diverticulum, which is an indication for surgery. Resection of the diverticulum, myotomy, and Dor fundoplication to prevent reflux were thus performed via thoracotomy. These modalities improved the patient’s symptoms and no postoperative symptoms of obstruction or NERD were observed.

The present patient had esophageal dysmotility due to DES and an increased LES pressure due to achalasia, with worsening symptoms due to the rapid enlargement of a true

**Manometric Recording**

The manometric recording of the esophageal body and lower esophageal sphincter (LES) on admission showed a high LES pressure and simultaneous prolonged, high-amplitude contractions in response to swallowing.

**Histopathology Findings**

The histopathology findings demonstrated a true diverticulum with muscle layer in the diverticulum wall (arrows) (Hematoxylin and Eosin staining; original magnification, 100×).
diverticulum in the lower esophagus. The transition from DES to vigorous achalasia and classical achalasia (9, 14), as well as that from vigorous achalasia to classical achalasia (9, 15), have been previously reported. Moreover, cases with no distinct differences in the clinical symptoms or test findings between vigorous and classical achalasia have been reported (11). Further studies in a larger number of patients are thus needed to clarify the pathology of these esophageal motility disorders.

In conclusion, we herein presented a rare case of a true giant epiphrenic esophageal diverticulum associated with vigorous achalasia diagnosed by preoperative manometry.

The authors state that they have no Conflict of Interest (COI).

References