

A Rare Coincidence of Esophageal Intramural Pseudodiverticulosis with Esophageal Web

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We report a rare case of esophageal intramural pseudodiverticulosis (EIPD) associated with esophageal web in a 67-year-old man presenting with dysphagia. EIPD is characterized by multiple tiny flask-shaped outpouchings of the mucosa that extend into the muscular layer on esophagography. EIPD commonly presents with stricture and less commonly with esophageal web. Although etiologies of both EIPD and esophageal web are unclear, a chronic inflammatory condition has been proposed. Treatment of EIPD is usually directed at the associated conditions rather than at the pseudodiverticulosis itself. In our case, dysphagia was successfully relieved by endoscopic dilatation with incision methods for the esophageal web. (**Korean J Helicobacter Up Gastrointest Res 2015;15:196-199**)

Key Words: Esophageal intramural pseudodiverticulosis; Esophageal web; Dysphagia; Endoscopic incision

INTRODUCTION

To date, only three cases of esophageal intramural pseudodiverticulosis (EIPD) associated with esophageal web have been published world-wide.¹⁻³ EIPD is characterized by multiple tiny flask-shaped outpouchings of the mucosa that extend into the muscular layer and is diagnosed by characteristic findings on esophagography or endoscopy. We report a rare case of EIPD associated with esophageal web in a 67-year-old man presenting with dysphagia which was successfully treated by endoscopic incisional therapy.

CASE REPORT

A 67-year-old man presented with a 6-year history of dysphagia for solid foods and odynophagia. The patient adapted either by cutting the food into small pieces and eating along with consumption of liquid. The patient had a medical history of rheumatoid arthritis and benign prostatic hypertrophy. There was no history of alcohol abuse,

but there was a smoking history of 365 packs per year. Physical and laboratory evaluation was unremarkable. Laboratory evaluation at admission revealed hemoglobin 14.5 g/dL, hematocrit 42.8%, AFP 3.03 ng/mL, CEA 3.20 ng/mL, CA 19-9 27.10 U/mL, and glucose 95 mg/dL. Computed tomography of the neck and chest showed no abnormal finding.

Upper endoscopy revealed a circular web with luminal stenosis at the upper-third of the esophagus, 17 cm from

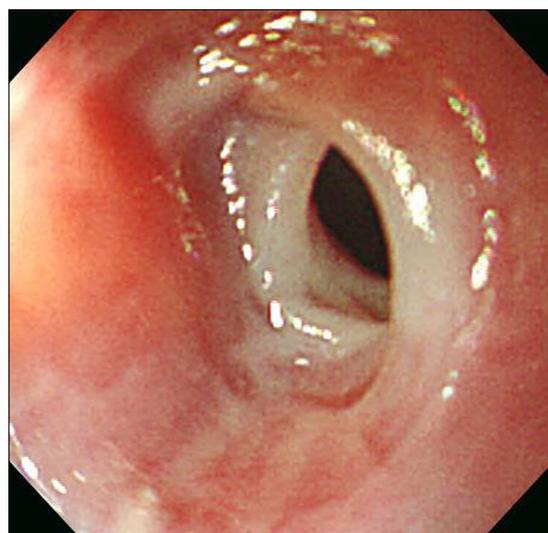


Fig. 1. Upper endoscopy showing circular esophageal web at upper esophagus (17 cm from incisor).

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the incisor (Fig. 1). The endoscope could not be advanced beyond the web. A barium esophagography showed a stenotic lesion at the upper thoracic esophagus and multiple 1~2 mm sized small outpouchings along the mid-esophageal wall (Fig. 2). We obtained the informed consent from the patient and approval of the ethics committee for endoscopic therapy for the stenotic lesion before the procedure. CRE balloon (Boston Scientific, Natick, MA, USA) dilatation was tried as an initial therapy. However, it was not tolerable because of pain and

coughing during the procedure. Endoscopic incisional therapy using insulation-tipped (IT) knife (MIW Endoscopie, Goldsbergstrasse, Germany) was planned as an alternative management (Fig. 3A). Once the web was incised without complications (Fig. 3B), the scope could be introduced through the stenotic lesion. Beyond the lesion, there were many small holes representing the diverticular orifice with diffuse post-inflammatory scarring surrounding mucosa at the mid esophagus (Fig. 3C). LA classification B reflux esophagitis was also noted at the gastroesophageal junction. No mass forming lesion suggesting malignancy was noted. Biopsy performed at some of diverticuli

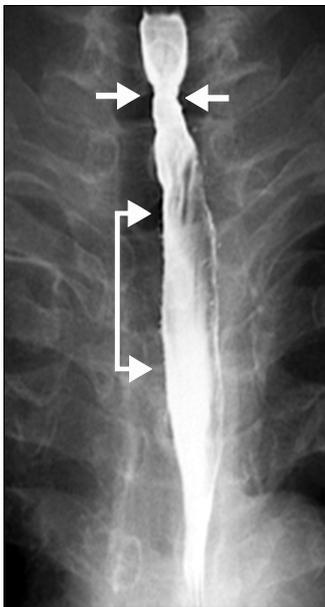


Fig. 2. Barium esophagography shows esophageal web and multiple esophageal pseudodiverticuli. Arrowheads indicate cervical esophageal web. Multiple outpouchings are found on the upper esophagus between arrows.

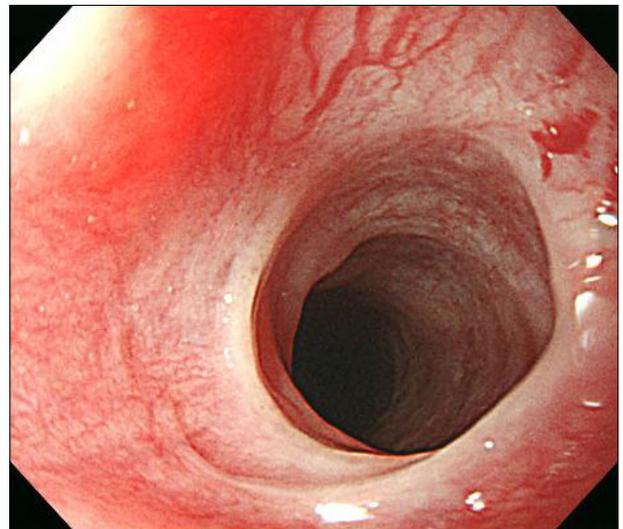


Fig. 4. Follow-up endoscopic finding after 6 months from initial treatment shows residual web, but much improved luminal narrowing.

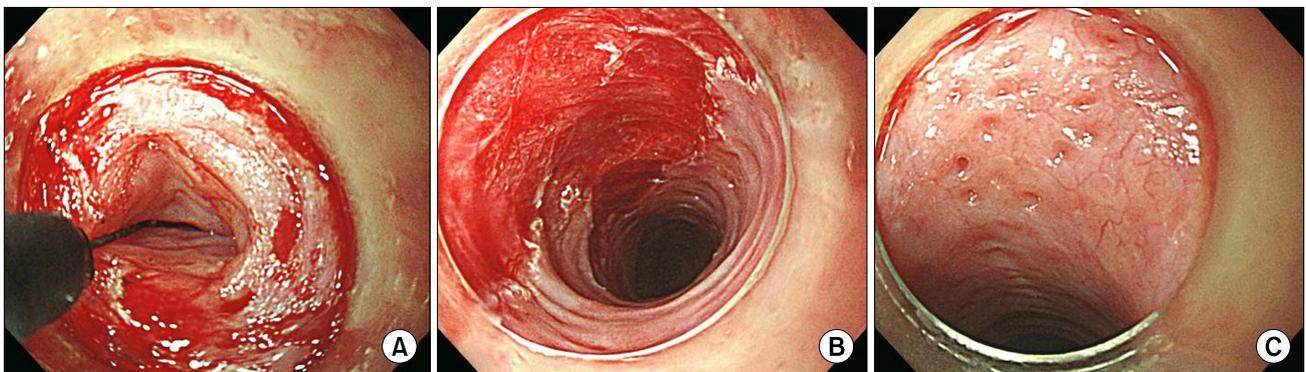


Fig. 3. (A) Endoscopic incision of esophageal web with insulation-tipped knife. (B) Esophageal web was incised without complication. (C) Endoscopy revealed multiple and small dimples representing the orifices of pseudodiverticuli beyond the web (18~22 cm from incisor).

proved to be non-specific acanthotic squamous mucosa. The procedure was well tolerated by the patient without any complication and dysphagia was relieved 2 days after the procedure. On the basis of symptoms and endoscopic and esophagographic images, EIPD with esophageal web was diagnosed. Proton pump inhibitor (lansoprazole 30 mg once a day) and sucralfate was applied for a month. Follow-up upper endoscopy performed 6 months later showed a small residual web, but much improved finding of luminal narrowing (Fig. 4) and complete healing reflux esophagitis (Fig. 4), but no interval change in number of diverticular orifice. At a 2-year follow-up, the patient was asymptomatic. Treatment with endoscopic incisional therapy was successful.

DISCUSSION

EIPD can occur in any age group, and shows an age peak between 50 and 70 years, and is more prevalent in men.^{4,5} Herter et al.⁴ summarized 197 cases and found that more than 90% of cases are involved with esophageal stenosis. The distal third of the esophagus predominates (42%) and most of these stenosis are typically long and concentric.⁴ Because EIPD is generally associated with the presence of esophageal strictures, these patients invariably complain of dysphagia. Indeed, dysphagia is the predominant symptom of this disorder.^{4,5} The severity of dysphagia does not correlate with the number of pseudodiverticula or the extent of involved esophagus. In our case, the patient presented with dysphagia and odynophagia for 6 years, and the symptom may have been associated with esophageal web.

EIPD has been described in association with diabetes mellitus, esophageal candidiasis, reflux esophagitis, chronic alcohol abuse, Mallory-Weiss syndrome, esophageal web, and esophageal cancer.⁴⁻⁸ Although several etiologies have been proposed for this condition, it is not yet well known. Obstruction of the exit site of the esophageal mucus glands by inflammation has been suggested as main cause.^{5,9,10} The obstruction of the ductal orifices by periductal inflammation or fibrosis gives rise to an increase in the viscosity of mucus, exfoliated epithelial cells, and fibrosis of mucosal and submucosal connective tissue.

As a consequence, the ductal portion of the glands is considered to become dilated, forming pseudodiverticular pouches.^{10,11}

Although congenital causes are believed to be the most common cause, esophageal web has been associated with several other conditions, including thyroid disease, Zenker's diverticular, esophageal duplication cyst, and especially also several inflammatory states.¹² We suppose that the chronic inflammatory condition is one of the common etiologies for rare coincidence of EIPD with esophageal web. In the present case, diffuse post-inflammatory mucosal scarring was also present in the endoscopic finding.

Some authors have hypothesized that when EIPD manifests in childhood, a congenital origin is most likely which should be considered as a separate clinical entity.^{13,14} However another case report has been documented about a child with a previously documented normal esophagus who acquired EIPD after the development of gastroesophageal reflux disease.⁹

Esophagography is critical in the diagnosis of EIPD. The classical features are multiple flask-shaped outpouchings with narrow neck continuous with the esophageal lumen. The present case correlates well with these findings, and led us to confirm the diagnosis of EIPD.

On endoscopic examination, the orifices of pouches are observed as small holes on the mucosal surface. But an endoscopic examination usually fails to find the small openings and often leads to misdiagnosis as a normal esophagus in early cases. In one report, these findings were seen in only about 20% of patients.^{4,11}

EIPD is characterized by distinct excretory ducts with dilatation of the submucosal esophageal mucus glands on histology. Although endoscopic biopsy revealed acute or chronic inflammation in many cases, the diagnoses are often indecisive because intramural pseudodiverticula was not included in the biopsy specimen.¹¹ Diagnosis was usually made based on symptom assessment and endoscopic and radiologic findings, not by endoscopic forceps biopsy.^{7,8} In our case, the biopsy specimens from the pseudodiverticula contained only acanthotic squamous mucosa.

Treatment of EIPD is not necessary in the absence of symptom. Treatment is usually directed at the associated

condition rather than at the pseudodiverticulosis itself.^{5,11} When there is an accompanying esophageal stricture, mechanical endoscopic dilatation leads to clinical response.⁹ Dilatation of the stricture usually gives immediate and long-lasting relief of dysphagia.^{3,11} Recent reports showed that esophageal stenosis was combined at initial presentation in 52~88% of cases, and dysphagia recurred after initial mechanical dilatation therapy such as bougienage in 44~57% of cases.^{7,8} In addition, presence of reflux esophagitis was a significant prognostic factor for symptom recurrence.⁸ Serious complication associated with mechanical dilatation such as perforation was not reported in these studies.^{7,8}

We performed an endoscopic incision of esophageal web using an IT knife. The procedure was well tolerated by the patient without any complications during the 2-year follow-up period.

The clinical course of esophageal EIPD is generally good, although several serious complications such as perforation, bleeding, fatal mediastinitis, and esophagobronchial fistula have been reported.^{4,6,10,15-17} Non-surgical endoscopic therapy such as stent insertion in case complicated by fistula was also reported.¹⁸

In our case, EIPD was combined with esophageal web, and we could not diagnose EIPD until we perform incisional therapy for esophageal web. Probably, these two diseases coincide by the same etiology, such as heavy smoking and chronic irritation.¹² This case suggests that esophageal web should be considered as a concomitant disorder with EIPD, and endoscopic therapy could be effective for the relief of symptom.

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