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*Case report*

# Esophageal intramural pseudodiverticulosis (diffuse type)

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A rare case of esophageal intramural pseudodiverticulosis (EIPD) in a 65-year-old woman with intermittent dysphagia is reported. An upper gastrointestinal series revealed multiple pseudodiverticula, which had tiny flask-shaped outpouchings with a narrow neck extending outward from the upper to the lower thoracic esophageal wall. In particular, the occurrence of the pseudodiverticula was coincident with a narrowed segment of inflamed esophagus. The length of the esophagus involved was approximately 15 cm. Endoscopic findings included mild stricture and chronic inflammation of the mucosa. Biopsy specimens showed active chronic esophagitis with bacterial and *Candida* superinfection, but no evidence of neoplasm. Cellular local immune reactions, as a consequence of chronic inflammation, and possibly abnormal motor activity in the narrowed esophagus, may explain the etiological agent or may be possible secondary factors that caused the EIPD.

**Key words:** intramural pseudodiverticulosis, chronic inflammation, esophageal gland, esophagus

## Introduction

Esophageal intramural pseudodiverticulosis (EIPD) is a very rare benign disease that was initially described by Mendel et al.[1] in 1960. The usual presenting symptoms are acute or chronic intermittent dysphagia, which typically is not severe and sometimes may be slowly progressive. Radiologically, EIPD is manifested by multiple, small, flask-shaped outpouchings in the wall of the esophagus that correspond histologically to dilated excretory ducts of the submucosal mucous glands.[2–6] To our knowledge, there have been few Japanese cases reported in the English-language literature,[7–9] but almost 100 cases worldwide have been described in published reports.[10] The etiology and pathogenesis of EIPD have remained obscure. Here, we report the characteristic findings of EIPD on barium swallow examination and endoscopy. The etiology and pathogenesis of EIPD is discussed in terms of bacterial and/or *Candida* superinfection and the possible role of motor disorder of the esophagus with active chronic inflammation.

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## Case report

A 65 year-old-woman was referred to our hospital in November 2000 because of intermittent dysphagia, mainly with solids. This pattern had begun 1½ years before the consultation, but was not progressive. Physical examination showed no abnormalities and laboratory findings were within the normal limits. There was no past medical history of hiatal hernia, achalasia, diabetes mellitus, or malignant disease.

Radiological examination revealed numerous fine barium-filled projections in the narrowed portion, an approximately 6-cm length of the upper thoracic esophagus with proximal dilatation (Fig. 1a,b). The condition of the entire esophagus is shown in Fig. 1c. Most of the projections were present from the wall of the upper to middle thoracic esophagus with occasional tiny sacculations below this level. The area of involvement was not segmental, but diffuse. The length of esophagus involved was approximately 15 cm. A closer-view of the X-ray shown in Fig. 1c showed small outpouchings with narrow necks projecting up to 2–5 mm in length from the esophageal lumen; the larger ones (maximal diameter, 4 mm) were flask-shaped with a flattened base (Fig. 2). Esophagoscopy showed an

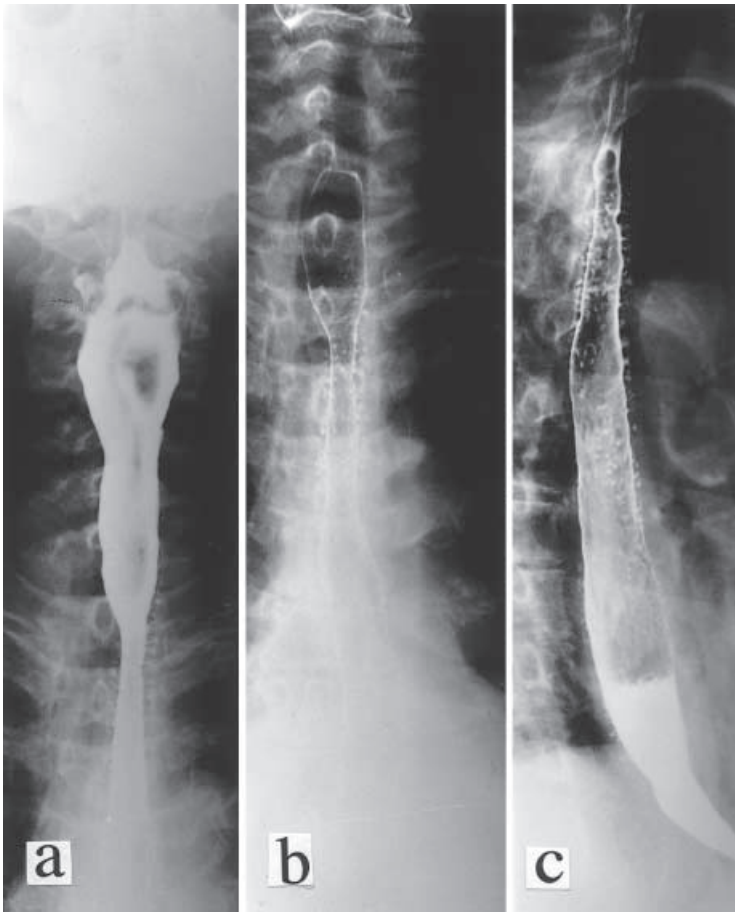


**Fig. 1a–c.** Radiography of the esophagus. **a** and **b** Filling and double-contrast esophagograms, showing dilated cervical esophagus with a distal 6-cm-long stricture. Protruding from this narrowed area are numerous small barium-filled projections. **c** Double-contrast esophagogram, demonstrating multiple diffuse tiny mucosal outpouchings, which are more apparent in the upper-to-middle thoracic esophagus

annular stricture of the esophagus 20 to 30 cm distal from the upper incisors. The endoscope (9-mm outside diameter; Evis XQ200; Olympus, Tokyo, Japan) passed through this narrowed region without difficulties. The esophageal mucosa exhibited chronic inflammation, showing redness, erosion, bleeding, friability, and exudate with numerous white plaques concentrated in the upper thoracic esophagus (Fig. 3). The multiple tiny orifices corresponding to pseudodiverticula were not clearly seen by endoscopic examination. Biopsy specimens of mucosa taken from the area of the stricture showed desquamative squamous epithelium with mild parakeratosis, without evidence of neoplasm (Fig. 4a). Bacterial colonies intermingled with pseudohyphae of *Candida* were better observed with periodic acid-Schiff staining (Fig. 4b). The prominent inflammatory infiltrate was composed of neutrophiles and lymphocytes, and these findings were compatible with active chronic inflammation. Based on the clinical, radiological and endoscopic evidence, the final diagnosis was EIPD, diffuse type. No direct treatments, such as antibacterial and antifungal therapy were required, because the patient reported symptomatic improvement with no further complaint of dysphagia after the endoscopic examination.

## Discussion

The esophagograms in our patient showed flask-shaped barium-filled projections, 2–5 mm in length along the esophageal wall and communicating with the esophageal lumen through narrow necks. These findings were frequent not only in the region of the strictures of the proximal esophagus, but throughout the entire esophagus. The histological findings in the biopsy specimens were of esophageal mucosa with chronic inflammation by bacterial and *Candida* superinfection.

According to a review of 97 patients with EIPD by Sabanathan et al.,[10] the distribution of pseudodiverticula was segmental in 57 patients (59%) and diffuse in 40 (41%). Radiological narrowing of the esophagus was present in 91% of the patients; 44% in the upper third, 23% in the middle third, and 33% in the distal third. Pathological findings from biopsy specimens are of limited value in detecting EIPD lesions, because the

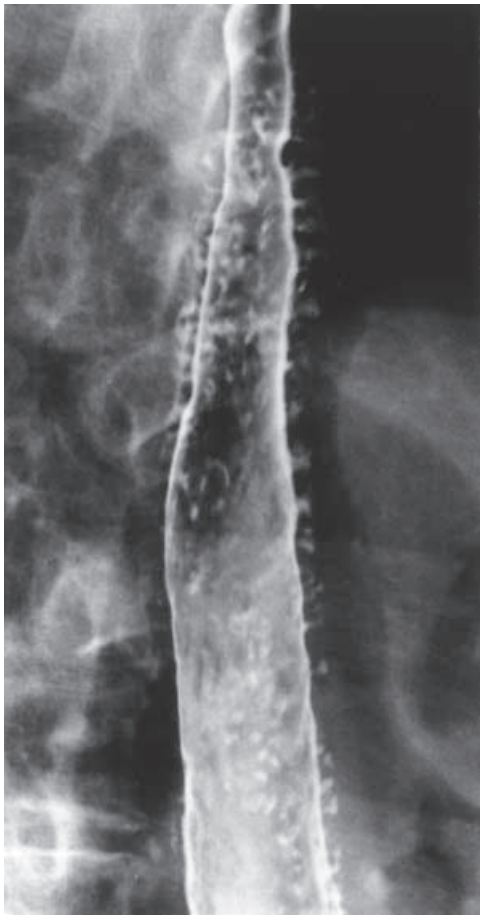

**Fig. 2.** Closer view of the outpouchings in shown Fig. 1c. Characteristic flask-shaped outpouchings of various sizes, some with narrow necks indicate numerous intramural pseudodiverticuloses

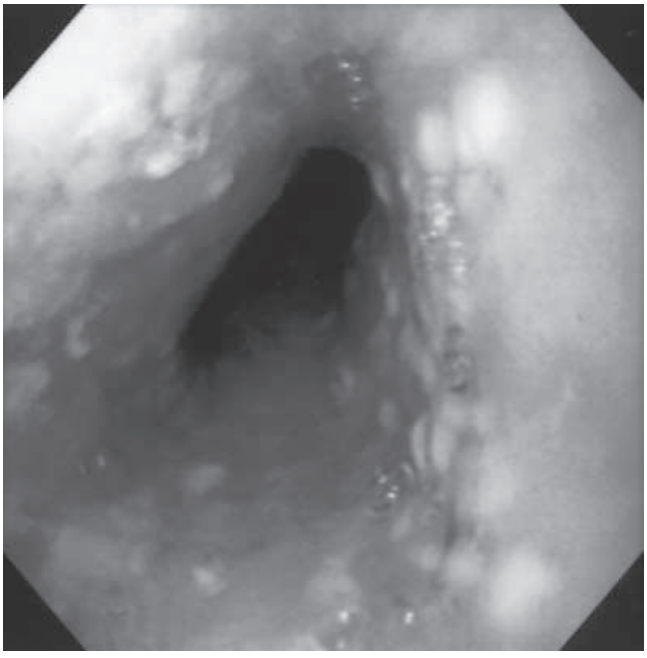

**Fig. 3.** Endoscopy of the proximal esophageal stricture showed mild annular stenosis with chronic inflammation. Redness, bleeding, and numerous white plaques are observed on the involved esophageal mucosa

pseudodiverticular formations are intramural, and are usually not included in the submitted specimens. In several reports of autopsy series[2–6,9] or surgical series[8,11] pathological examination of full-thickness sections from the esophageal wall showed that the flask-shaped outpouchings indicated cystically dilated excretory ducts of esophageal submucosal glands. In some specimens, these ducts were markedly dilated, producing grossly visible intramural cysts. It is likely that the fine barium-filled projections radiologically documented in our patient were pseudodiverticula, rather than true diverticula. Thus, this case could be reasonably diagnosed as a diffuse type of EIPD.

The precise etiology and pathogenesis of EIPD are not known. Most investigators consider that a multifactorial process is involved with the main problem being obstruction of the duct by desquamated epithelium, mucus, inflammatory materials, and/or submucosal fibrosis.[2,3,9,12] This condition results in an inflammatory duct-dilatation of the esophageal gland, revealed as flask-shaped outpouchings on esophagograms. Thus, the chronic inflammatory cellular reactions within the esophageal mucosa may be regarded as a primary etiological agent or secondary factors in EIPD. These are supported by the evidence that most of the reported psuedodiverticula could be located at or below the level of narrowing, and not above it. However, it has remained unclear that EIPD is rare, while chronic esophagitis with inflammatory cellular reactions is a relatively common condition. A great deal will certainly be learned about the etiology and pathogenesis of EIPD when full-thickness histological specimens become available from patients.

In EIPD, although the pseudodiverticula are frequently associated with esophageal narrowing together with infection, Sabanatham et al.,[10] in their review also described esophageal motility disorders; they noted that evidence of disordered motility was present in 29 (30%) of the total of 97 patients. These motility disorders included irregular tonic simultaneous contractions, tertiary contractions, aperistalsis, impaired peristalsis, lack of distensibility, exaggerated normal peristalsis and a nonspecific motility defect. Motility was described as normal in only 2 patients. Thus, the etiology of EIPD may involve abnormal motor activity of the esophagus to some extent. In fact, EIPD associated with achalasia or esophageal web has been reported increasing;[13–15] these abnormalities may possibly raise the esophageal

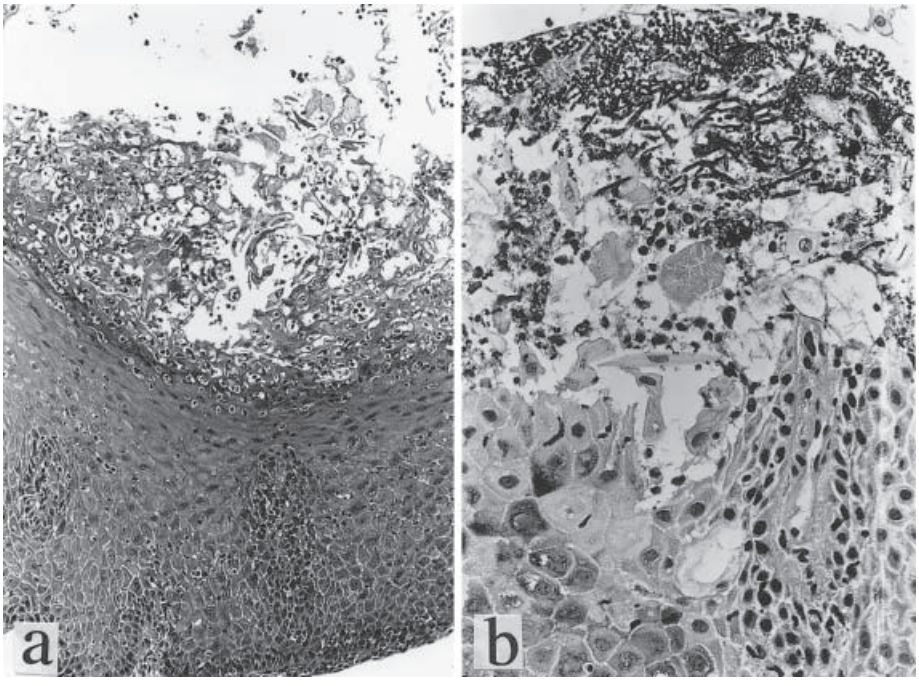


**Fig. 4a,b.** Microscopic findings of biopsy specimens of the esophageal mucosa. **a** Desquamative squamous epithelium and chronic inflammatory cells, showing active chronic esophagitis. H&E, ×40. **b** Bacterial colonies mixed with pseudohyphae of *Candida* (*upper part of the picture*) are noted. Periodic acid-Schiff, ×100

intraluminal pressure in the inflamed esophagus, and thus may play a role in the pathogenesis of EIPD. Contraction of the esophageal muscularis may also obliterate the narrow necks of the excretory ducts of submucosal esophageal glands.

The course of EIPD is benign with a good response to medical treatment for inflammation and/or to endoscopic dilatation of the esophagus,[10] but there have been a few instances in which the cysts perforated, thereby forming fistulae that extended into the mediastinum.[11,16] In our patient, there was no need for medical treatment, because the main clinical sign, dysphagia resolved spontaneously during the course of the examination. In particular, endoscopic maneuvers may contribute to the dilatation of the involved esophagus.

In summary, we reported a rare case of typical EIPD (diffuse type), which showed numerous, tiny, flask-shaped outpouchings with very narrow necks in the esophageal wall. Chronic inflammatory cellular infiltration triggered by bacterial and/or *Candida* superinfection, and possibly enhanced intraluminal pressure in the narrowed esophagus may represent the primary etiological agent or secondary factors in EIPD.

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