

## Pneumomediastinum as a complication of esophageal intramural pseudodiverticulosis

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### Abstract

Dysphagia is a common complaint of patients seen at the outpatient clinic as well as at the emergency room. We report esophageal intramural pseudodiverticulosis (EIPD) as a cause of dysphagia that is less known by physicians and it is rarely described in the literature. EIPD is characterized by multiple, segmental or diffuse, flask-like outpouchings in the esophageal wall corresponding to dilated and inflamed excretory ducts of the submucosal esophageal glands. The underlying etiology remains unclear. Esophageal strictures, esophageal candidiasis and gastroesophageal reflux disease are often associated. The diagnosis can be made by upper gastrointestinal endoscopy, but barium esophagography is the modality of choice. Complications of EIPD are rare and include broncho-esophageal and esophagomediastinal fistula, pleural and pericardial effusion, abscesses, gastrointestinal bleeding from a web-like stenosis or esophageal perforation with pneumomediastinum. The treatment for EIPD should be directed towards treating underlying associated conditions and relieving symptoms rather than the pseudodiverticulosis itself. (*Acta gastroenterol. belg.*, 2018, 81, 433-435).

**Key words** : Dysphagia, Esophageal intramural pseudodiverticulosis, Esophagogram, Perforation, Pneumomediastinum.

**Abbreviations** : EIPD, esophageal intramural pseudodiverticulosis, EGD, esophagogastroduodenoscopy, CT, computed tomography.

### Introduction

Dysphagia is a common complaint of patients seen at the outpatient clinic as well as at the emergency room. The differential diagnosis is broad and includes common causes such as gastroesophageal reflux disease, strictures, webs, tumors, foreign bodies, functional disorders etc. that are well known by physicians. However, more rare causes should always be kept in mind.

In this article, we report a rare cause of dysphagia due to esophageal intramural pseudodiverticulosis (EIPD) complicated by spontaneous esophageal perforation resulting in a pneumomediastinum.

### Case report

A 64-year-old man was seen at the outpatient clinic complaining about progressive dysphagia since the last week. He suffered from a pronounced intent to vomit, an excessive salivation and a vague chest pain. Physical examination was unremarkable.

Prior medical history revealed intermittent dysphagia the past two years with an episode of acute dysphagia one year ago due to a food impaction that was endoscopically removed. The esophagogastroduodenoscopy (EGD) at that time showed a benign esophageal stricture in the distal esophagus. Biopsy specimens that were taken from the stricture at that time showed active esophagitis and the presence of mycosis. The patient had been successfully treated with proton pump inhibitors and antimycotics for respectively four and two weeks and there was no recurrence of severe dysphagia until today.

Because of a recurrent episode of severe dysphagia today, a new EGD was performed as next step in the evaluation of this case. Endoscopic evaluation showed a stenosis of the distal esophagus that could be passed by the scope. Because of the vague chest pain already present before the procedure, we performed a standard chest X-ray that revealed free air as a sign of a pneumomediastinum (Fig. 1A). We consequently performed a computed tomography (CT) scan of the chest which confirmed the presence of a pneumomediastinum with dominant localization around the esophagus from the level of the gastroesophageal junction to the cervical base (Fig. 1B). Small intramural air bubbles in the proximal part of the esophagus were seen on these CT images suggestive for intramural esophageal pseudodiverticulosis. The next diagnostic step was to perform an X-ray esophagogram with Telebrix gastro® (joxitalamaat, meglumine) which could not reveal a leakage of contrast but revealed pathognomonic signs of EIPD. It also showed a linear to circular loss of filling probably due to the stenosis in the distal esophagus that was visualized on the EGD (Fig. 2A, B, C). We admitted the patient in the hospital for transient fasting, proton pump inhibitors and treatment with intravenous antibiotics and total parenteral nutrition. Radiographic control after one week no longer showed the presence of free air as a sign of a pneumomediastinum. Oral feeding was restarted without problems and the

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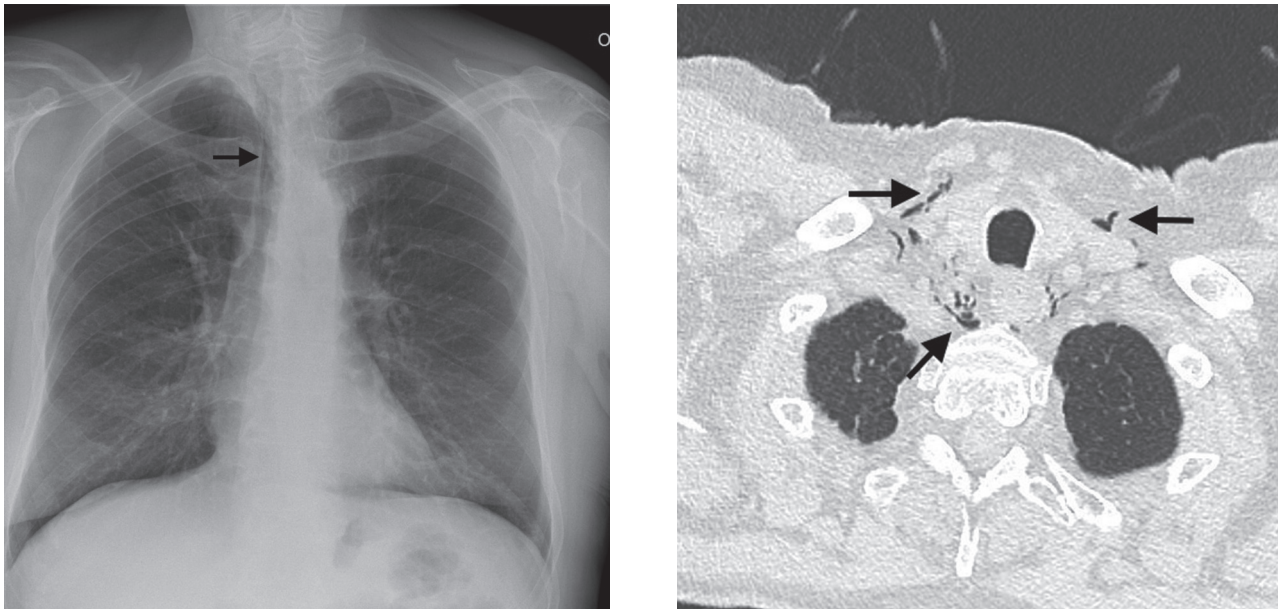


Fig. 1. — A. Chest X-ray after the esofagogastroduodenoscopy shows a radiolucent line at the right side in the upper mediastinum suspect for a pneumomediastinum due to a perforation of the esophageal wall. B. Computed tomography image, axial image, lung frame, shows free air in the mediastinum and intramural air bubbles in the esophageal wall. Black arrows on both images show free air as a sign of pneumomediastinum.

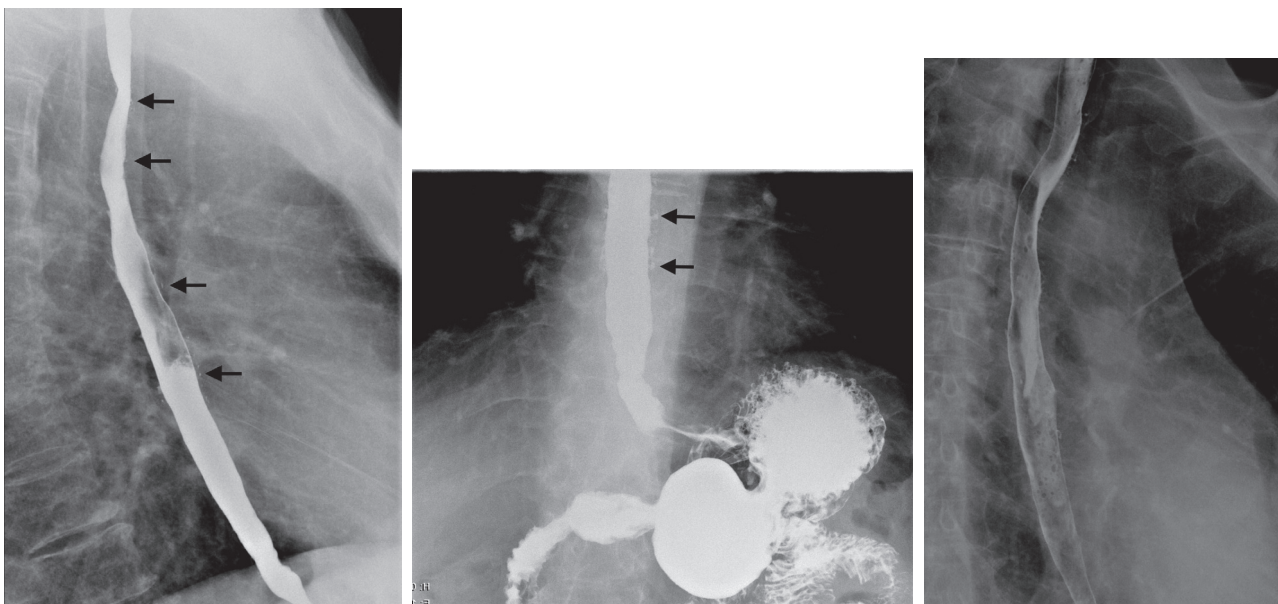


Fig. 2. — Esophagogram. The images show esophageal intramural pseudodiverticulosis and an esophageal stricture. A, B : Filling phase. C : Double contrast. Black arrows on the images show the typical signs of esophageal intramural pseudodiverticulosis.

patient was discharged out of the hospital. Follow-up consultation was scheduled and a balloon dilatation of the stenosis will be considered.

### Discussion

Esophageal intramural pseudodiverticulosis (EIPD) is a rare condition first described by Mendl et al in 1960 (1). It is characterized by multiple, flask-like outpouchings in the esophageal wall with segmental or diffuse involvement. Histologically, the outpouchings correspond to dilated and inflamed excretory ducts of the submucosal esophageal glands and therefore, is it not true diverticulosis (2). The presence of this esophageal

inflammation in autopsies of patients with EIPD suggests that the disease may be the result of chronic esophagitis. This can lead to extrinsic compression of the ducts by periductal inflammation and fibrosis (3). Some authors report that obstruction and resultant dilation of the ducts is caused by inflammatory material, mucus and desquamated epithelium (4). Otherwise, the association between EIPD and certain motoric disorders of the esophageal wall such as diabetes and achalasia suggests a disturbance of neurologic function as a possible cause. This could explain the manometric abnormalities such as longer tubular peristalsis waves or aperistalsis common seen with EIPD (2,4).

EIPD occurs predominantly in the sixth and seventh decade and is slightly more prevalent in males. It seems to be associated with conditions like diabetes mellitus, candidiasis, reflux esophagitis and chronic alcohol abuse (4,5). However, the explanation for these associations remains unclear.

The predominant symptom of EIPD is dysphagia that can be constant, intermittent or progressive. It can also cause odynophagia, chest discomfort or can be asymptomatic. A few case reports mention weight loss due to anorexia or, rarely, hematemesis or melena (4,6). Since dysphagia is the most common symptom, and due to its evolution and widespread utilization, an EGD is often the first diagnostic procedure performed in these patients. During the examination, a direct visualization of the small orifices of the pseudodiverticula can be diagnostic; however, according to literature, it can only detect 25% of the cases (5). According to Hahne et al. the endoscopic detection of EIPD depends on a sufficient air insufflation during EGD. Endoscopy may also determine if disease is segmental or diffuse and it allows the treatment of strictures. When perforation is suspected, endoscopy should be strictly avoided in order not to aggravate by insufflating more air through the perforation (7). For establishing the diagnosis of EIPD, a barium esophagogram is the modality of choice (4). While a single-contrast technique provides maximal esophageal dilatation and is useful for the depiction of a stricture, a double-contrast examination allows accurate imaging of the esophageal mucosa and wall (8). Barium swallow study may also help to detect many associated conditions eg hiatal hernia, gastrointestinal reflux, motility disturbances or cervical webs (4). Manometry studies can show various patterns of esophageal motility, including normal, localized or diffuse aperistalsis, hypermotility or other non-specific findings (5,6).

The treatment for EIPD should be directed towards treating underlying associated conditions and relieving symptoms rather than the pseudodiverticulosis itself. Hence the association of EIPD with gastro-esophageal reflux disease and esophagitis, proton pump inhibitors are reasonable and commonly administered. Although the role of candidiasis in the etiopathogenesis remains unclear, antifungal therapy is usually prescribed in cases with documented candidiasis (6). In cases of dysphagia, a drug therapy with nifedipine or metoclopramide can be tried, especially if hypercontractile motility disturbances are found on manometry reports (4). Esophageal strictures are frequently associated ; in this case, mechanical

endoscopic dilatation should be performed and can bring quick and lasting relief. In our case, as in the case reported by Termote et al (7), the pneumomediastinum was probably caused by spontaneous rupture of intramural pseudodiverticula and probably exaggerated by air insufflation during EGD. Other complications of EIPD are rare and include broncho-esophageal and esophagomediastinal fistula, esophageal perforation, pleural and pericardial effusion, abscesses or gastrointestinal bleeding from a web-like stenosis (4,6,9). When complications are suspected on a clinical basis or based on other examinations (eg mediastinal free air on a chest X-ray) one should not use barium but a non-ionic iodinated water soluble contrast medium to avoid barium leaking into the mediastinum (7). In patients with EIPD, an increased risk for the development of squamous cell carcinoma of the esophagus has been reported. Therefore, every esophageal stricture should be evaluated individually for signs of malignancy. Periodic surveillance of patients with EIPD may be worthwhile though further investigation is necessary to confirm this association (10). Yet, there is still no consensus about timing and frequency of endoscopic follow-up in these patients.

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