Case Report

Esophageal Intramural Pseudodiverticulosis Complicated With Stricture

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We report a rare case of diffuse esophageal intramural pseudodiverticulosis in a 35-year-old man complaining of severe dysphagia and vomiting for several months. The advanced morphological change in the esophagus caused irregular track formation, mimicking an ulcerative lesion on esophagogram. Endoscopic examination revealed an esophageal stricture with intact mucosa. Endoscopic ultrasonography and chest computed tomography showed multiple hyperechoic lesions of unknown nature and multiple air collection sites in the esophageal wall, respectively, making diagnosis difficult. The patient finally received a subtotal esophagectomy because of severe symptoms. The lesion was pathologically proven to be intramural pseudodiverticulosis with marked submucosal fibrosis. Our experience suggests that awareness of this rare pathology and the related image changes will be helpful for early diagnosis and treatment in the future.

Key Words: computed tomography, endoscopic ultrasound, esophagogram, esophagus, pseudodiverticulosis

Esophageal intramural pseudodiverticulosis (EIPD) is defined as multiple small pinhead-shaped outpouchings, occurring in the submucosa of the esophageal wall. It is caused by the dilation of the excretory ducts of the esophageal mucus glands. EIPD was first described in 1960.1 Since then, around 220 cases have been reported in the literature,2 with most being diagnosed using image studies. EIPD occurs in all age groups, predominantly in the 6th and the 7th decades, with a slight predilection for males.3,4

The prevalence rate is unknown. An estimation of about 0.09–0.15% detection rate in those cases undergoing esophageal radiological examination have been reported.3,5,6 The most common symptom is dysphagia with a clinically progressive course.3 Many patients are symptom-free and are only incidentally discovered.

Herein, we present a rare case of EIPD with esophageal stricture. The advanced morphological change presented in this case caused diagnostic confusion.
Case Report

A 35-year-old male was admitted because of severe dysphagia and vomiting for several months. He was noted to have suffered occasional dysphagia in his adolescence. The condition worsened in spite of medications (data not shown) provided by many other hospitals. There was no history of hypertension or diabetic mellitus, but his history was remarkable, including Fournier’s gangrene in October 2001, duodenal ulcer in March 2002, and multiple excisions of sebaceous cysts in the past.

The patient was well developed, with a height of 170 cm, but malnourished with a weight of 42 kg. Multiple variable-sized sebaceous cysts were present on his face, body, and extremities. Otherwise, he appeared normal. Laboratory analysis including blood, urine, stool, and biochemistry were all within normal range. A barium esophagogram revealed irregular tracks and some pinhead-sized outpouchings on the esophagus (Figure 1). These observations were initially interpreted as esophageal ulcerative lesions of an undetermined nature. Computed tomography (CT) showed thickening of the esophageal wall. Air collection in the wall was also present (Figure 2). An endoscopic examination showed mid-esophageal stricture with smooth mucosa surface (Figure 3). Endoscopic ultrasonography (EUS) performed during endoscopic examination disclosed multiple intramural submucosal hyperechoic shadows, indicating probable abscess or malignancy (Figure 4). These differing results confused clinicians’ judgment. In the belief that the patient had an esophageal stricture, we performed a total esophagectomy and reconstruction to treat severe intractable dysphagia and vomiting.

The resected esophagus revealed diffuse wall thickening and fibrosis with marked narrowing of the middle portion. The mucosal surface was smooth. Multiple tiny cystic lesions were found in the submucosal area of the esophageal wall.

![Figure 1. Esophagogram reveals pinhead-sized lesions (black arrow) with irregular tracking (white arrow) over the esophagus mimicking an esophageal ulcerative lesion.](image1)

![Figure 2. Intravenous enhanced chest computed tomography scan shows intramural air (arrow) adjacent to the esophageal lumen, thickening of the esophageal wall.](image2)

![Figure 3. Endoscopic examination reveals esophageal structure. The lining mucosa is intact. A submucosal lesion cannot be ruled out.](image3)
Cysts were lined with metaplastic squamous epithelium (Figure 5) and their openings were in continuity with the esophageal lumen. Some cysts were connected and formed irregular channels. Candidal and bacterial colonies were identified in some cysts. Chronic inflammation and marked submucosal fibrosis were also present.

**Discussion**

EIPD is a rare disease and can be segmentally (59%) or diffusely (41%) distributed. The underlying etiology is still unknown, but EIPD is usually considered to be acquired because it occurs in elderly patients in most cases. However, congenital factors cannot be ruled out in cases found in childhood. In the current case, congenital factors may play an important role, given the fact that his symptoms began in adolescence.

EIPD is often associated with other conditions such as gastroesophageal reflux (24%), hiatal hernia (7%), diabetes mellitus (16%) and chronic alcoholism (16%). Other associated conditions include Mallory-Weiss syndrome, esophageal web, squamous cell carcinoma, and achalasia. EIPD associated with multiple cutaneous sebaceous cysts has not been previously described, and their relationship is not clearly understood. This observation may be a significant interesting association because both lesions share a similar pathogenesis during the course of cyst development.

Medeiros et al found submucosal chronic inflammation surrounding esophageal mucous glands in more than 60% of autopsy cases of EIPD. This finding reflects that early pathologic changes of EIPD might be more common than expected. Infection is also present in the cystic development of EIPD, with *Candida* being the most common pathogen in the lesions.

Diagnosis is often difficult due to a lack of awareness. Image studies are often helpful but may also cause problems when formulating a diagnosis. Generally, the traditional barium esophagogram is a more sensitive clinical method than endoscopic examination. Endoscopic examinations usually fail to disclose tiny cystic openings and often leads to misdiagnosis as a normal esophagus in early cases. The classical features of EIPD on barium esophagogram are multiple flask-shaped outpouchings with narrow necks contiguous with the esophageal lumen. Great variation may be present because of the change of localization and the severity of the disease. An irregular track mimicking an ulcerative lesion or malignancy on the barium esophagogram may be present in advanced cases, as shown here. A combination of clinical history, endoscopic examination, and other image studies are indicated to avoid confusion and misdiagnosis.
EUS and CT should be performed when endoscopic examination fails to disclose any lesion in the esophagus in such a case with abnormal esophagogram. Using EUS, Yoshimoto et al noted the thickening of the second and third sonographic layers and multiple acoustic shadows due to gas collection within the esophageal wall.\textsuperscript{12} Pearlberg et al demonstrated typical CT features of EIPD characterized by marked esophageal wall thickening, irregularity of the esophageal lumen, and intramural gas collections.\textsuperscript{13} Awareness of these changes on both EUS and CT might be important in the diagnostic procedure.

The major complication of EIPD is stenosis.\textsuperscript{14} Other complications include esophagopulmonary fistula, perforation, and upper gastrointestinal bleeding. Hahne et al pointed out that conservative management may lead to satisfactory control of the symptoms in early diagnostic cases.\textsuperscript{10} Balloon dilatation may help some cases of early esophageal stricture,\textsuperscript{15} but esophagectomy should be considered for those cases with advanced esophageal stricture. Early diagnosis and treatment with adequate antibiotics is recommended for the prevention of severe complications.

References