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Esophageal tubular Duplication Complicated with Intraluminal Hematoma : A Case Report

Esophageal tubular duplication is a rare congenital anomaly. We experienced a patient with esophageal tubular duplication who presented with a swallowing difficulty which was aggravated after a gastrofiberscopic examination. Preoperative diagnosis was intramural hematoma of the esophagus due to trauma caused by endoscopy. Surgical specimen revealed that hematoma was located within a duplicated lumen of the esophagus. The radiologic and endoscopic findings are discussed in correlation with its pathology.

Key Words: *Esophageal Diseases; Abnormalities; Wounds and Injuries; X-ray Tomography, Computed; Endoscopy, Digestive System; Pathology, Surgical*

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INTRODUCTION

Duplications of the alimentary tract are rare congenital malformations that may occur at any level from the mouth to the anus. By definition, it is located in or adjacent to the wall of the gastrointestinal tract, possessing smooth muscle in the wall, and is lined by alimentary tract mucosa. The lining mucosa is not necessarily that of adjacent segment of the gastrointestinal tract (1).

Esophagus is the second most common site of alimentary tract duplications. Pathologically, cystic and tubular types have been recognized. Most of them are cystic, and tubular type is extremely rare. There has been no well-documented report regarding esophageal tubular duplication on CT and endoscopic examination with pathologic specimen. We report the endoscopic and radiological findings in a patient with esophageal tubular duplication.

CASE REPORT

A 68-year-old man presented with swallowing difficulty which was aggravated since the first endoscopic examination at a private clinic. He was a heavy drinker. On the second endoscopic examination at our hospital, multiple variable sized mucosal defects were found at the mid-esophagus 24 to 28 cm from the incisor teeth and the defects were communicating with another esophageal lumen (Fig. 1A). The lumen of the esophagus showed gradual narrowing. Another small mucosal defect was found 38 cm from the incisor teeth. Chest radiograph demonstrated widening of the mediastinum and thickening of the retrotracheal stripe. Initial contrast-enhanced CT scan demonstrated double lumens along the nearly entire esophagus. The larger lumen was filled with hematoma that was intermediate attenuation (30 HU). The smaller lumen was encompassed by the larger one. A thin-enhancing rim was seen around the outer margin of the larger lumen (Fig. 1B). We made a diagnosis of intramural hematoma of the esophagus caused by traumatic endoscopic examination.

Twenty days after the initial visit, a follow-up third endoscopic examination was performed. The esophageal luminal narrowing was slightly relieved, but the mucosal defects have not been changed.

On the follow-up CT scans three months later, the intermediate density of hematoma disappeared, but air-filled double esophageal lumens still persisted (Fig. 2A). Esophagogram showed double-barrel shaped esophagus communicating between two lumens (Fig. 2B).

About ten months after initial presentation, transthoracic esophagectomy with esophagogastrostomy was performed because the patient complained of persistent

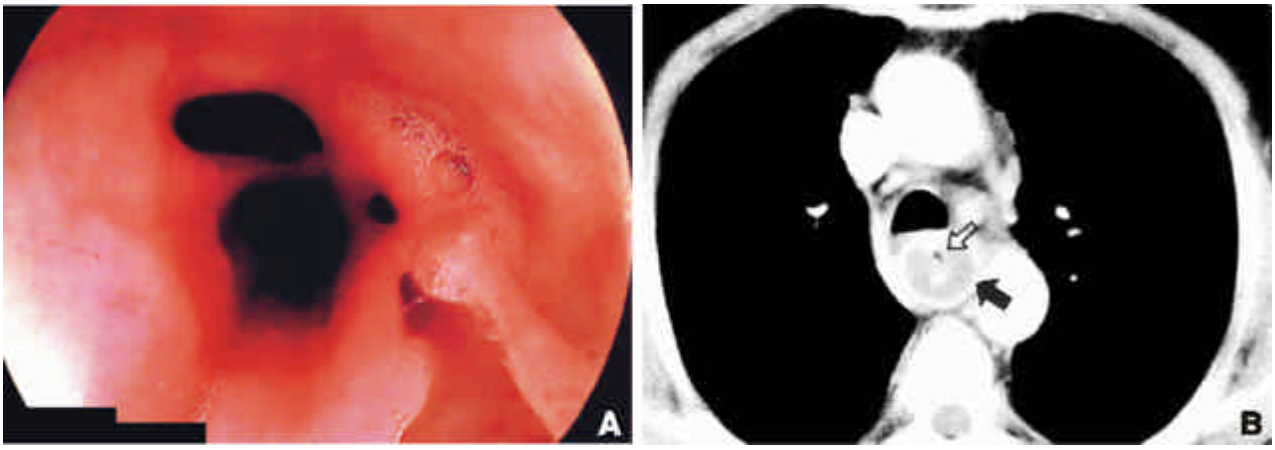

Fig. 1. A 68-year-old man with aggravating swallowing difficulty after endoscopy. Initial studies. **A:** Endoscopic examination shows multiple and irregular mucosal defects communicating with another lumen. **B:** Initial CT scan at the level of aorticopulmonary window shows two lumens of the esophagus. Air-filled smaller inner lumen (open arrow) is surrounded by hematoma-filled larger outer lumen (closed arrow). A thin enhancing rim of the esophagus suggests mucosal lining.

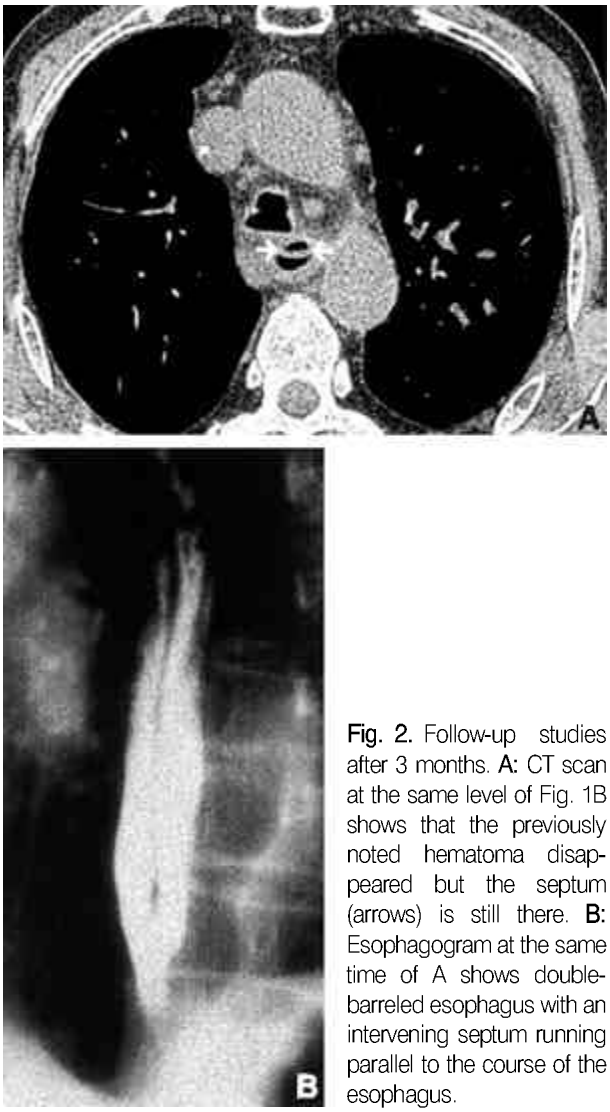

Fig. 2. Follow-up studies after 3 months. **A:** CT scan at the same level of Fig. 1B shows that the previously noted hematoma disappeared but the septum (arrows) is still there. **B:** Esophagogram at the same time of A shows double-barreled esophagus with an intervening septum running parallel to the course of the esophagus.

dysphagia and recurrent symptoms of esophagitis. Pathologic specimen revealed that the entire esophagus had irregular longitudinal septum-like structures with multiple defects (Fig. 3A). Cross-cut 1:1 specimen revealed double lumens lined with stratified squamous epithelium, submucosa, and muscularis propria (Fig. 3B). Epithelial layers of each lumen were contiguous through the multiple mucosal defects. Pathologic diagnosis was esophageal tubular duplication. There were no ectopic gastric mucosa. The intermediate density area found in the initial CT scan and luminal narrowing of esophagus were thought to be caused by an intraluminal hematoma in the duplicated lumen. The hematoma might have been induced by an initial traumatic endoscopic examination.

DISCUSSION

The term, 'alimentary tract duplication', was first used in 1941 by Ladd, who made a comprehensive compilation of many descriptive terms, such as enterogenous cyst, ileum duplex, giant diverticula, or unusual Meckel's diverticulum (2). Esophageal tubular duplication is extremely rare. In an analysis of 281 duplications in 4 previous series, esophagus is the second most common site of alimentary tract duplication, following the terminal ileum, and consisting of 20% of all lesions (1). All cases of esophageal duplications in that series were noncommunicating and cystic. In another review, 2 of 20 esophageal duplications were communicating tubular type (2). Only several cases of tubular type of esophageal duplication, including the above-mentioned 2 cases, have been reported in English literature (3, 4). But none of them described specific radiographic findings. To our best

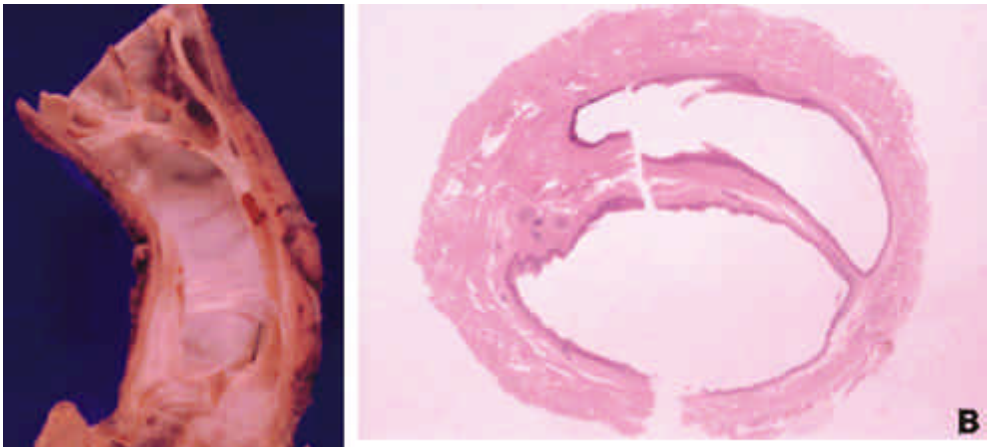

Fig. 3. Pathologic specimen of esophagectomy 10 months after initial presentation. **A:** Pathologic specimen of the esophagus after longitudinal incision of the wall and the septum shows two separate lumens and a shaggy septum along the entire course of the esophagus with multiple septal defects. **B:** Crosscut 1:1 specimen reveals double lumens with their own stratified squamous epithelium, submucosa, and muscularis mucosa. Section above this specimen shows a direct continuation of the epithelial layers between two lumens (H&E, ×1).

knowledge, this is the first to describe radiological and endoscopic findings with a pathologic correlation.

Although the etiology of duplication of the esophagus has not been well established, several theories have been proposed: 1) abortive twinning theory; 2) persistent embryologic diverticula theory; 3) bronchopulmonary foregut malformation theory; 4) split notochord theory; 5) aberrant luminal recanalization theory; 6) intrauterine vascular accident theory (1). Although no single theory can satisfactorily explain the various clinical and pathologic manifestations, most investigators considered that aberrant luminal recanalization theory would be the most likely. During embryologic development, the esophagus is partially solid. The final lumen is established by the formation of vacuoles that coalesce. Several vacuoles may fail to become connected with the esophageal lumen and remain as a duplication of esophagus.

Clinically, esophageal duplications tend to be diagnosed more frequently in adult life rather than in childhood, because these patients are asymptomatic unless complications occur. When symptomatic, they presented with recurrent dysphagia, occasional chest pain, and signs and symptoms of acute mediastinitis due to rupture of the duplication (4). Mass effect on adjacent structures in the mediastinum is usually seen in cystic type, but there has been no reported case showing mass effect in tubular duplications. Most of the reported esophageal tubular duplications communicate with their native lumen, often through multiple openings (3). But most duplication cysts of esophagus are of the noncommunicating type (1, 5).

Plain radiographic findings of esophageal tubular duplication are nonspecific. Mediastinal widening on frontal projection and thickening of retrotracheal stripe on lateral projection are demonstrated suggesting esophageal abnormality. Esophagogram of communicating esophageal tubular duplication shows a specific finding of double-barreled esophagus with a longitudinal septum between two lumens. Contrast-enhanced CT scan may show additional tubular structure along the esophagus with a thin and slightly enhancing rim encircling the wall of the esophagus. These findings may mimic dissecting intramural hematoma of the esophagus in Boerhaave syndrome (6). In our case, initial diagnosis was dissecting intramural hematoma. Using CT scan alone, it is difficult to differentiate between tubular duplication and dissecting intramural hematoma, particularly in patients who have a history of esophageal trauma.

Although radiological and endoscopic examinations may suggest the diagnosis, a definitive preoperative diagnosis is difficult. A definite diagnosis can be made by pathologic examination after surgical excision. On the pathologic examination, both lumens of the esophagus should be lined by stratified squamous epithelium and covered by muscularis propria for sure diagnosis of duplications. Forceps biopsy is usually not helpful. The definitive treatment for esophageal duplication is surgical excision.

REFERENCES

1. Macpherson RI. *Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. RadioGraphics 1993; 13: 1063-80.*
2. Ildstad ST, Tollerud DJ, Weiss RG, Ryan DP, McGowan MA, Martin LW. *Duplications of the alimentary tract: clinical characteristics, preferred treatment, and associated malformations.*

Ann Surg 1988; 208: 184-9.

3. Granelli P, Angelini GP, Tambussi AM, Nicolini A, Celli L. *Tubular duplication of the oesophagus in a child: clinical, radiologic and endoscopic features. Endoscopy 1983; 15: 263-5.*
4. Mikaelian DO, O'keefe JJ, Simonian S. *Duplication of the esophagus. Ann Otol Rhinol Laryngol 1981; 90: 392-5.*
5. Fernbach SK. *Lesions involving multiple areas of the gastrointestinal tract. In: Gore RM, Levine MS, Laufer I, eds. Textbook of gastrointestinal radiology. Philadelphia: Saunders, 1994; 1485-8.*
6. Lee KS, Kim YI, Kim PN. *Dissecting intramural hematoma of the esophagus in Boerhaave syndrome: CT findings. Am J Roentgenol 1991; 157: 197-8.*