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Congenital Esophageal Stenosis due to Tracheobronchial Remnants

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〈국문초록〉

기관지 잔유물에 의한 선천성 식도 협착증

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기관지 잔유물(tracheobronchial remnants)과 연골환(cartilaginous ring)형성은 선천성 식도 협착증의 드문 원인이 된다. 증상은 비특이적이며 과거에 선천성 식도협착증이라고 생각되는 많은 환자에서 결과적으로 열공 탈출(hiatus hernia)이나 식도염에 의한 염증성 협착증으로 밝혀져 왔었다.

본 질환의 방사선 소견의 특징은 협착 부위에서 돌출되는 선상의 식도벽내의 열(linear intramural clefts)로서 이러한 열은 병리학적으로 호흡상피가 내부로 배열된 벽내의 낭포 공간이다.

이러한 식도 협착은 기계적 조작에 의하여 확장되지 않고 염증성 점막 변화가 없는 것이 특징적 내시경 소견이다.

저자들은 1974년부터 최근까지 서울대학병원에서 기관지 잔유물에 의한 선천성식도 협착증 2예와 위궤양과 기관지 잔유물 1예를 경험하였다.

Tracheobronchial remanants and formation of cartilaginous ring are rare causes of congenital esophageal stenosis. Symptoms are nonspecific and many patients with presumed congenital esophageal stenosis subsequently have been demonstrated to have inflammatory strictures due to hiatus hernia or esophagitis.

Characteristic radiological findings of this disease are linear intramural clefts projecting from the area of stenosis and these clefts represent pathologically communicating intramural cystic spaces lined by respiratory epithelium.

Resistance to instrumental dilatation and the absence of inflammatory mucosal changes are characteristic endoscopic findings. Authors experienced 3 cases of tracheobronchial remnants associated with esophageal stenosis (2 cases) and esophageal duplication with tracheobronchial remnants in gastric fundus from 1974 until recently.

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I. Introduction

A cartilaginous ring associated with other tracheobronchial remnants is a rare cause of distal esophageal stenosis in infants and children.

The etiology is unknown. According to the most popular theory, its development is related to defective separation of the embroyonic respiratory tract from the primitive foregut, resulting in sequestration of tracheobronchial precursor cells in the wall of the esophagus[1-4].

We experienced 3 cases of tracheobronchial remnants associated with esophageal stenosis (2 cases) and esophageal duplication with gastric ulcer.

II. Materials

Case 1

A 7-year-old female patient was admitted to Seoul National University Hospital with the complaint of vomiting and dysphagia after ingestion of solid food since 1 year old.

Past medical history was unremarkable. Barium esophagogram showed abrupt narrowing at distal esophagus with marked proximal dilatation. Multiple linear recesses of barium extend intramurally from the region of stricture (Fig. 1).

Radiological diagnosis was esophageal stenosis due to tracheobronchial remnants. Resection of lower portion of esophagus and gastroesophagostomy were done.

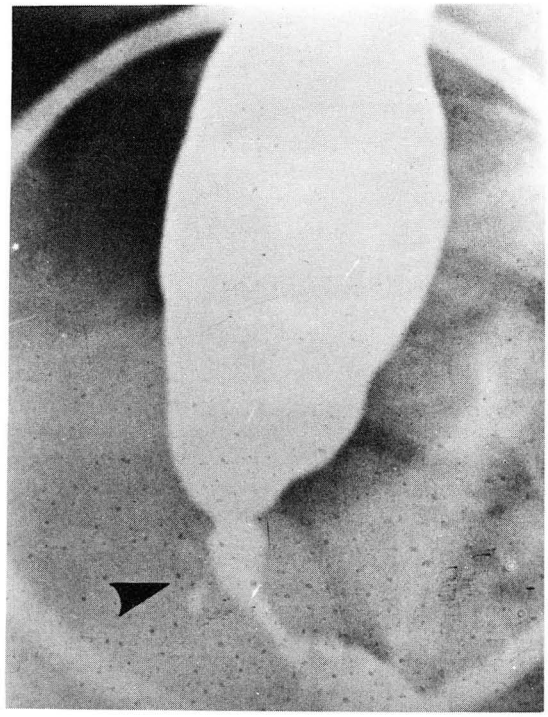


Fig. 1. Case 1
Esophagogram shows abrupt narrowing at distal esophagus with marked proximal dilatation. Multiple linear recesses of barium extend intramurally from the region of stricture (arrowhead).

Case 2

A 2-year-old female patient was admitted to Seoul National University Hospital with the complaint of intractable post-prandial vomiting since 6 month old. She was born with low birth weight of 1.1 kg and a first born baby of twin.

Her development was retarded and she has had recurrent aspiration pneumonia. She was known case of esophageal stenosis. Palliative gastrostomy was done at the age of 1 year and 3 month.

Barium esophagogram showed extreme narrowing at esophagogastric junction with marked proximal dilatation. Multiple linear recesses of barium extend intramurally from the region of strictures.

Radiologic diagnosis was esophageal stenosis due to tracheobronchial remnants (Fig. 2A).

Operation showed cartilaginous ring constricting the esophagus at 2cm above diaphragm. The ring was quite firm. Resection and end to end anastomosis were done. Postoperative diagnosis was esophageal stenosis due to tracheobronchial remnants.

Pathologic examination of resected specimen showed anomalous wall formation consisting of

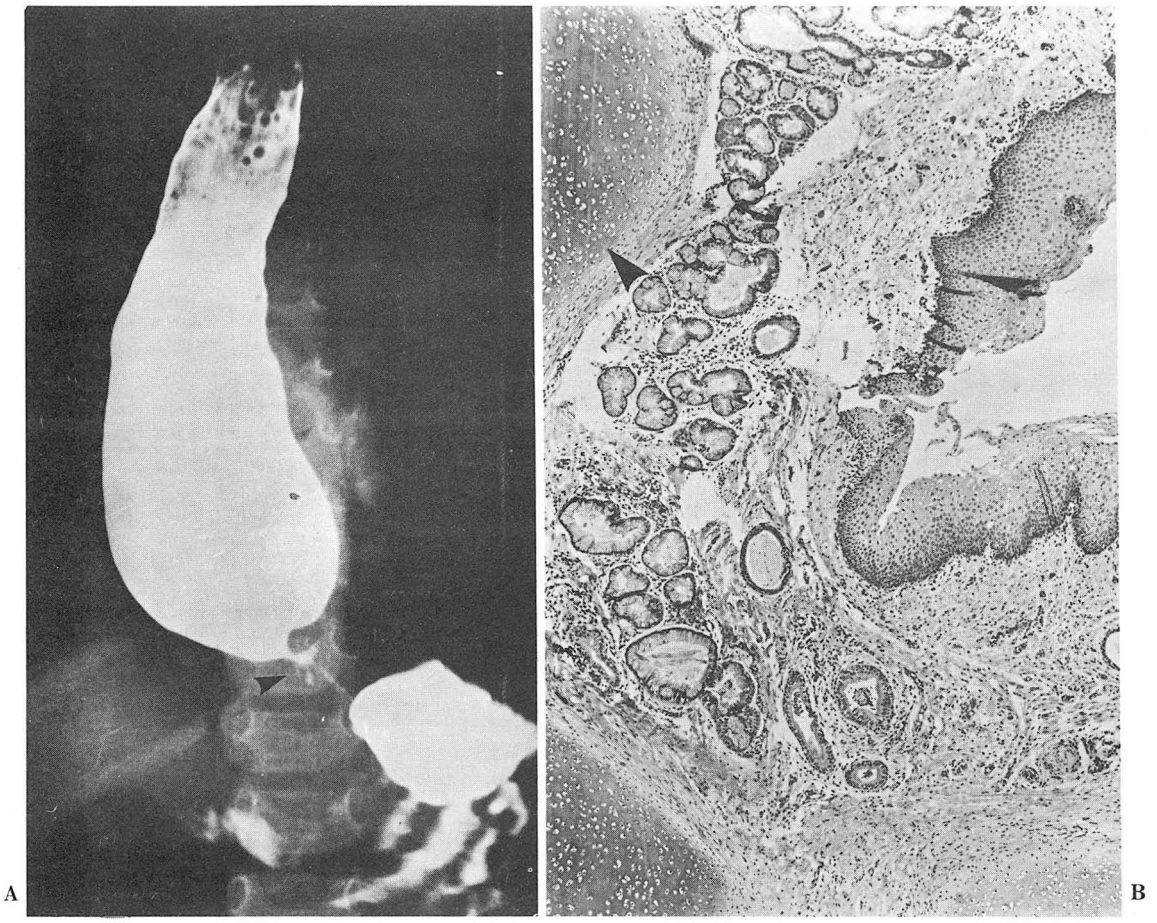


Fig. 2. Case 2

A. Esophagogram shows extreme narrowing at esophagogastric junction with marked proximal dilatation. Multiple linear recesses of barium extend intramurally from the region of strictures (arrowhead).

B. Cross-section of the stenotic area of esophagus shows cartilaginous ring (arrowhead) located outside the muscle layer and prominent mucous glands.

prominent mucous glands and cartilage islands (Fig. 2B). Post-operative esophagography showed no unusual findings. The patient's post-operative course was uneventful.

Case 3

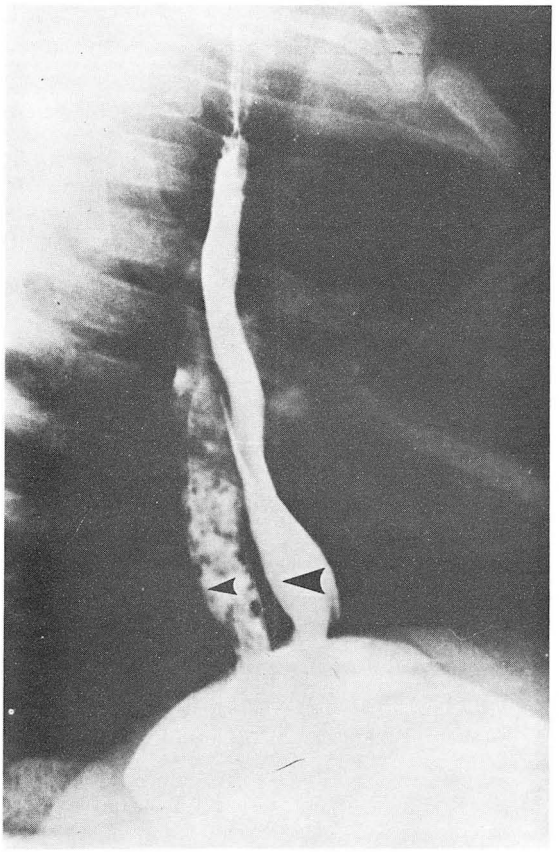
A 2-year-old female was admitted to Seoul National University Hospital with the complaint of vomiting since born. She underwent an operation due to intestinal obstruction caused by jejunal web. After surgery she had persistent postprandial vomiting and melena.

Esophagography showed duplication of esophagus, ulcer at esophagogastric junction, narrowing of distal esophagus and gastroesophageal reflux of minor degree suggestive of esophagitis (Fig. 3A).

U.G.I. showed small and ovoid barium collection at fundus along the lesser curvature(Fig. 3B).

Operation showed duplication of esophagus, deep penetrating gastric ulcer along lesser curvature and Meckel's diverticulum at ileum.

Esophagoesophagostomy and fundoplication were done. Pathologic examination of tissues



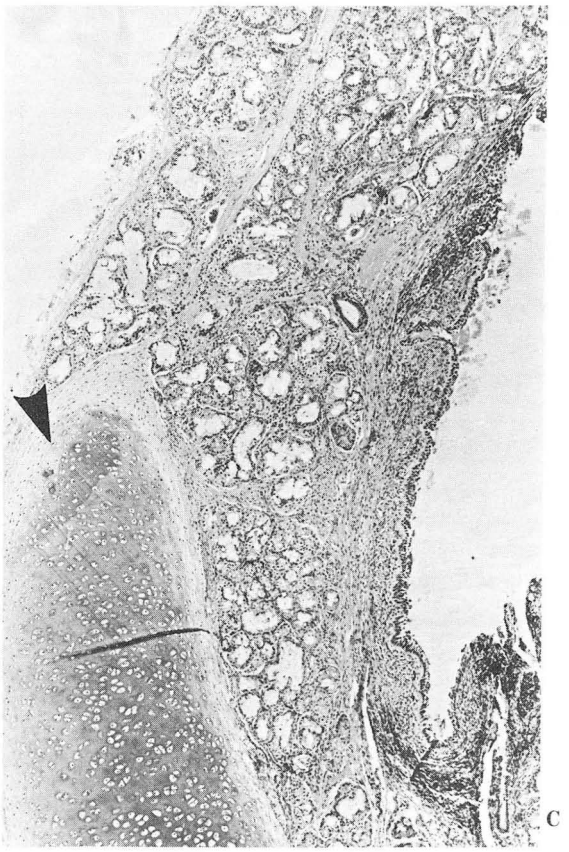


Fig. 3. Case 3

A. Lateral esophagogram shows duplication of esophagus. (large arrow: esophagus proper small arrow: duplicated esophagus posteriorly)

B. U.G.I. shows small and ovoid barium collection at fundus along the lesser curvature (arrowhead).

C. Cross-section of specimen from stomach shows bronchial lining, prominent submucous glands and cartilaginous ring (arrowhead) located outside the muscle layer.

from gastric ulcer showed tracheobronchial remnants with chronic inflammation (Fig. 3C). The patient's post-operative course was uneventful.

III. Discussion

The occurrence of esophageal stenosis associated with remnants of the tracheobronchial tree has been reported by many authors[1-4]. Stenosis due to tracheobronchial remnants in the esophageal wall is quite rare as a cause of symptomatic narrowing of the esophagus[1-8].

The etiology of the anomaly is unknown. According to the most popular theory, its development is related to defective separation of the embryonic respiratory tract from the primitive foregut, resulting in sequestration of tracheobronchial precursor cells in the wall of the esophagus[1-4].

The clinical features are common to other causes of symptomatic esophageal narrowing in infants and children and include recurrent vomiting, particularly after the addition of solid foods to the diet. Dysphagia has been the predominant symptom cited in the few reports of older children. Most cases have occurred in infancy or childhood. In the cases reported to date, boys and girls have been equally affected[4].

The X-ray findings are often nonspecific, showing a localized constriction in the distal esophagus with proximal dilatation. Hiatal hernia is not an associate finding. The multiple small projections of barium extending perpendicularly from the stenosis in our cases are distinctive[4,7,10].

From the standpoint of diagnosis it is important to be sure that one is not dealing with stenosis as a result of hiatus hernia. Resistance to instrumental dilatation and the absence of inflammatory mucosal changes are characteristic endoscopic findings[1,2,4,7]. Cinefluoroscopic evaluation of the esophagus is very helpful in establishing the diagnosis of esophageal stenosis due to congenital tracheobronchial remnants, since the stenosis does not dilate with swallowing.

The relationship of the clefts to intramural esophageal diverticula is not entirely clear[4,11,12]. Intramural esophageal diverticula are most commonly thought to be acquired lesioins, occurring in patients with a history of dysphagia and representing mucosal herniations into the submucosa which are triggered by abnormal motor activity[4,11]. The small luminal outpouchings seen in intramural esophageal diverticulosis are radiographically indistinguishable from congenital clefts lined by respiratory epithelium. In the latter condition, however, the association of the clefts with localized distal esophageal stenosis in a child or in an adult with symptoms dating back to childhood has been distinctive in the cases reported thus far and should suggest the correct diagnosis.

In terms of treatment, primary resection with reanastomosis is recommended for congenital stenosis not amenable to dilatation[2]. The treatment is operative, either by segmental resection of the esophagus, or by extramucosal resection of the abnormal tissue which might appear preferable when feasible[1].

IV. Summary

Two cases of congenital esophageal stenosis caused by tracheobronchial remnants and one case of esophageal duplication with tracheobronchial remnants are reported.

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