

## A RARE CAUSE OF DISTAL ESOPHAGEAL STENOSIS DUE TO HETEROTOPIC TRACHEOBRONCHIAL REMNANT

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Gazi Medical Journal 7 : 141-143, 1996

**SUMMARY :** *In this report, we present a nineteen-month-old boy, who referred to our clinic for vomiting and failure to thrive. At the operation, there was a hard tissue resembling cartilage at the distal esophagus. The patient underwent esophagocardiomyotomy, extramucosal excision of the structure and Gavrilu procedure.*

**Key Words :** *Esophageal Stenosis, Tracheobronchial Remnant.*

### INTRODUCTION

Various causes of congenital esophageal stenosis have been reported, one of which is the tracheobronchial rest. Most of the patients presented with isolated lesions, and Deiraniya reported the first ones associated with esophageal atresia and tracheoesophageal fistula (1). We report a nineteen-month-old boy who became symptomatic at the age of six months. He was operated on with the prediagnosis of achalasia, and a tracheobronchial remnant was encountered during the operation. Our aim is to discuss this case in the light of the relevant literature.

### CASE REPORT

A nineteen-month-old boy was admitted to our clinic with vomiting after each feeding and failure to thrive. His complaints began at the age of six months when semisolid and solid food were introduced into his diet. He had been treated several times for repeated pulmonary infections. Esophagogram taken during the search of the recurrent pneumonia showed stenosis at the distal esophagus

with proximal dilatation, and the patient was sent to our clinic with the prediagnosis of achalasia (Fig. 1).

On physical examination, no pathological findings were observed other than his percentile line of 10-25 % in weight. Esophageal pH monitoring re-

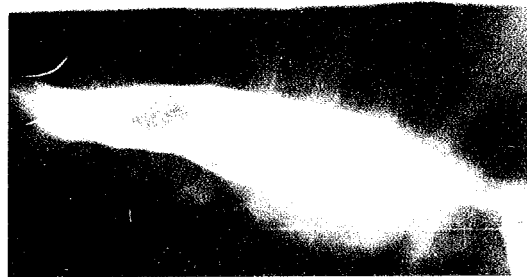


Fig - 1 : Preoperative esophagogram.

vealed no evidences of gastroesophageal reflux.

Laparotomy was performed with the prediagnosis of achalasia. A hard tissue on the anterior portion of the distal esophagus which was resembling cartilage, was palpated. Extramucosal excision of this crescent-like 1.5 x 0.5 cm lesion was done with esophagocardiomyotomy, and the gastric fundus was sutured to the edges of the myotomy line separately (Gavrilu procedure).

Esophageal mucosa was perforated iatrogenically during the excision of the structure and repaired with interrupted sutures.

Histological examination of the resected specimen showed that it consisted of cartilaginous tissue and mucus glands (Fig. 2).



Fig - 2 : Histological appearance of the resected specimen.

Some postoperative leakage occurred from the repaired mucosa and was treated conservatively. On the 38th postoperative day, relaparotomy was performed with the prediagnosis of adhesive intestinal obstruction and the bands were released. The area of the former operation and the esophagogastric junction was normal.

In the follow-up period, esophageal pH monitoring showed no gastroesophageal reflux, and the passage was normal in contrast radiograms.

## DISCUSSION

Frey and Duschl (1936) reported the first case of esophageal stenosis due to heterotopic tracheobronchial tissue in a nineteen-year-old girl who died with the diagnosis of achalasia. Approximately 150

further cases have been reported throughout the world until today (1, 6). Congenital esophageal stenosis (CES) is a rare anomaly. According to Nihoul and Fekete, CES is an intrinsic stenosis of the esophageal wall; generally not symptomatic at birth. They classify the etiology as : 1- tracheobronchial rest (TBR) (cartilage, respiratory mucus glands and/or ciliated epithelium), 2- amembraneous diaphragm, 3- segmentary hypertrophy of the muscularis and diffuse fibrosis of the submucosa (fibromuscular stenosis) (3). The incidence of CES with esophageal atresia (EA) and the more distal location of CES caused by TBR can be explained on an embriological basis as the sequestration of tracheobronchial anlage before the separation of the respiratory tract from the esophagus and the caudal movement of the esophagus by a differential growth (2, 3, 5, 6). Cartilaginous tissue, mucus glands and respiratory epithelium can be found histologically in TBR in different combinations (1, 2, 3, 5). Cartilaginous tissue and mucus glands were encountered on histological examination of the specimen excised in our case. Symptoms usually start in early infancy, patients presenting with progressive dysphagia and vomiting after solid food is introduced into the diet (2, 4, 6). Recurrent respiratory infections, probably as a result of aspiration pneumonia were reported in a number of cases. The diagnosis of CES associated with EA begins with a high index of suspicion. It is important to verify patency of the distal esophagus at the time of the primary anastomosis by passage of a tube to the stomach intraoperatively. Early symptoms also include aspiration manifesting with recurrent pneumonia or apnea. Failure to thrive can also be an important manifestation of a TBR due to nutritional insufficiency.

Although symptoms usually start in early infancy, delay in diagnosis is common. Radiographic contrast studies typically show a narrow stricture at lower esophagus. The esophagus above the stenotic segment is dilated and often has poor peristaltic activity. An evidence of GER cannot always be encountered in contrast studies. Esophageal pH monitoring was normal and GER was not observed in our case. A stenosis lined by normal esophageal mucosa at the distal part of the esophagus can be seen at esophagoscopy. The complaints of our patient had started at the age of six months, and after repeated pulmonary infections, he was operated on with the prediagnosis of achalasia. In the treatment of TBR, dilation and bouginage offer only a transient

benefit. Primary resection of the stenotic segment in annular stenosis and end-to-end anastomosis or colonic transposition is recommended as definitive treatments (6). Extramucosal excision of the remnant is a choice of treatment in crescent-shaped lesions. As the lesion was on the anterior portion of the esophagus and there was no need for an esophageal resection, we preferred this type of operation. Diagnosis can only be confirmed by histological examination of the resected specimen. The disease should not be overlooked and surgeons must always consider the probability of a TBR in similar cases.

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