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CASE REPORT | ESOPHAGUS

H-type Tracheoesophageal Fistula: A Rare Cause of Cough and Dysphagia in Adults

Jeffrey Bank, MD[1], Rebecca Voaklander, MD[1], and Michael Sossenheimer, MD, PhD[1]

[1]*Division of Gastroenterology and Hepatology, University of Utah School of Medicine, Salt Lake City, UT*

ABSTRACT

Tracheoesophageal fistula without associated esophageal atresia (H-type) is a rare congenital anomaly, accounting for about 4% of esophageal malformations. However, it can occasionally be seen in adults with chronic cough and respiratory infections. We present a 38-year-old woman with a new diagnosis of H-type tracheoesophageal fistula.

INTRODUCTION

In the adult population, congenital tracheoesophageal fistulas (TEFs) are rare. Most congenital TEFs are associated with esophageal atresia and identified in childhood. TEF without associated esophageal atresia (H-type) is a rare congenital anomaly, accounting for about 4% of esophageal malformations.[1] It is defined as a connection between the posterior tracheal and anterior esophageal walls and is typically diagnosed before age 3. It is an important, albeit unusual, cause of chronic cough and respiratory infections in adults. We present a 38-year-old woman with a new diagnosis of H-type TEF.

CASE REPORT

A 38-year-old woman with multiple previous hospitalizations for hypoxia (presumed because of asthma and aspiration in the setting of achalasia) was admitted with hemolysis, elevated liver enzymes, and low platelet count syndrome complicated by hypoxia. Her previous pulmonary function tests showed severe airway obstruction and echocardiogram showed normal left and right ventricular systolic function. Her medical history was notable for a congenital lung abnormality, severe pneumonia in infancy, upper lobe resection, and yearly hospitalizations for hypoxic respiratory failure. She was diagnosed with achalasia in 2006 at age 25. Workup included an esophagogastroduodenoscopy (EGD) in 2006 that was notable for narrowing at the gastroesophageal junction and an irregular Z line; esophageal biopsies were normal. Esophageal manometry in 2006 showed absence of peristalsis and hypotensive contractions per review of limited outside records. A double contrast esophagram in 2012 (without pressure injection technique) demonstrated proximal esophageal dilation, distal esophageal narrowing without passage of a 13 mm barium tablet, and poor peristalsis, all suggestive of achalasia. It also showed significant tracheal aspiration, worse in the recumbent position.

After delivery, she was transferred to the pulmonary service for management of her hypoxia. Subjectively, she reported coughing when supine and with meals. A TEF without inflammation was noted on thoracic computed tomography and confirmed by EGD and bronchoscopy. EGD showed a 3 mm proximal esophageal fistula (Figure 1). Bronchoscopy demonstrated a normal oropharynx and vocal cords, erythematous larynx, inflamed arytenoids, a 4 mm proximal TEF without mucosal abnormalities, and right upper lobe absence (Figure 2). A swallow study showed aspiration on the posterior side of the trachea presumed to be due to the TEF. A multidisciplinary literature review and case discussion by pulmonary, gastroenterology, thoracic surgery, and ENT specialists determined that surgical repair rather than endoscopic clip or stent placement was the optimal treatment for her. The patient underwent esophagoplasty, muscle flap reconstruction TEF repair, and laparoscopic Heller myotomy with Dor fundoplication. The patient's tracheal repair healed well without evidence of fistula recurrence or further dysphagia.

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Correspondence: Michael Sossenheimer, MD, PhD (michael.sossenheimer@hsc.utah.edu).

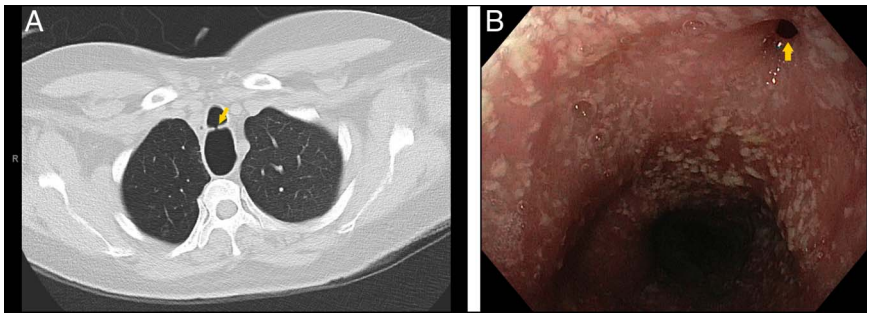

Figure 1. (A) Thoracic computed tomography in axial view showing tracheoesophageal fistulas without inflammation and (B) esophagogastroduodenoscopy with a 3-mm proximal esophageal fistula located 17 cm from the incisors.

DISCUSSION

H-type TEF is rare and can mimic chronic lung disease due to aspiration, recurrent infection, and bronchiectasis. This diagnosis must be considered in adults with recurrent respiratory infections, coughing with oral intake, and rarely, hemoptysis. Complaints of dysphagia or an achalasia diagnosis offer clues to the presence of a H-type TEF because the fistula disrupts the esophageal myenteric plexus development, causing impaired peristalsis and lower esophageal sphincter dysfunction. A thoracic computed tomography, bronchoscopy, esophagoscopy, and barium esophagram are crucial to confirm the diagnosis of a TEF. Although over-the-scope clip (OTSC) closure of fibrotic TEFs has been technically successful in case studies, an international study of 16 cases reported a 62% technical success rate.[2–11] Overall, the technical success rate using OTSC devices is unclear.[12] In addition, OTSCs may obliterate surgical tissue planes. Surgery is the optimal treatment for H-type TEF and reported to be 94%–100% successful.[13–15] However, there is a high rate (22%) of recurrent laryngeal nerve injury after H-type TEF repair and gastroesophageal reflux (56%).[14,15] A simultaneous Heller myotomy and Dor fundoplication should be considered at the time of surgical repair as H-type TEF is associated with impaired esophageal peristalsis and lower esophageal sphincter dysfunction.

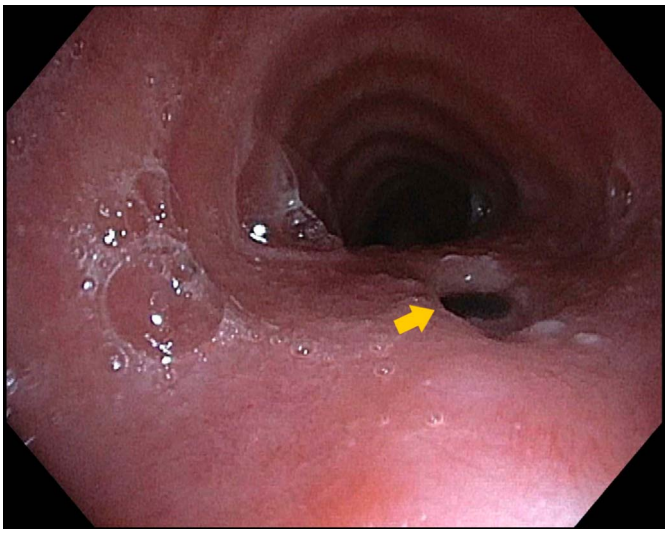

Figure 2. Bronchoscopy showed 4-mm proximal tracheoesophageal fistulas without mucosal abnormalities 5 cm from the vocal cords.

DISCLOSURES

Author contributions: J. Bank wrote the manuscript. R. Voaklander edited the manuscript. M. Sossenheimer edited and revised the manuscript for intellectual content and is the article guarantor.

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