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Clinical Images

A Quarterly Column

Early Presentation of H-Type Tracheoesophageal Fistula

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INTRODUCTION

H-type or isolated tracheoesophageal fistula occurs along the spectrum of esophageal malformations. Five types of esophageal malformation are described in the literature.[1] The most common type is esophageal atresia with a distal tracheoesophageal fistula (75%). H-type tracheoesophageal fistulas occur without associated esophageal atresia and represent approximately 5% of all cases.

HISTORY

An 8-day-old, large-for-gestational-age male infant born to a mother with poorly controlled diabetes mellitus was admitted to the neonatal intensive care unit for possible congenital heart disease, congenital umbilical hernia, and hypoglycemia at birth. The umbilical hernia was reduced shortly after birth without incidence. The infant developed episodic cyanotic and coughing spells with associated respiratory distress during oral feeds. An echocardiogram revealed a right-sided aortic arch with concerns for a vascular ring. The patient was referred to Radiology for further investigation.

RADIOGRAPHIC APPEARANCE AND TREATMENT

Initial esophagram demonstrated significant swallowed oral contrast material within the tracheobronchial tree (Figure 1). No oropharyngeal aspiration was identified, and a tracheoesophageal fistula was strongly suspected. A pull-back tube esophagram demonstrated a small fistulous tract between the esophagus and trachea at the level of the thoracic inlet compatible with an H-type tracheoesophageal fistula (Figure 2).

Primary surgical repair of the tracheoesophageal fistula was performed. Intraoperative direct laryngotracheal bronchoscopy demonstrated a fistula that was identified in the upper trachea. The fistula tract was cannulated with a Fogarty catheter (Figure 3). Through a small supraclavicular incision, the sternocleidomastoid muscle was retracted laterally, the strap muscles were retracted medially, and the tracheoesophageal groove was identified. The fistula was identified and encircled with 2 vessel loops (Figure 4). The exposed anterior wall of the fistula was opened and the catheter exposed for illustrative purposes and removed. The fistula was then suture ligated on the esophageal and tracheal sides.

DISCUSSION

H-type tracheoesophageal fistulas represent approximately 5% of cases of esophageal malformation. Tracheoesophageal fistula is thought to represent esophageal dysembryogenesis during the 4th/5th week of embryogenesis. Other associated congenital anomalies are present in 50%-70% of patients. These anomalies include cardiovascular (35%); gastrointestinal (24%); neurologic (10%); skeletal (13%); and those of vertebral, anal atresia, cardiac, tracheal, esophageal, renal, and limb association (25%).[1]

Early diagnosis of H-type tracheoesophageal fistula can often be difficult to make because of nonspecific symptoms that can be attributed to more common etiologies such as gastroesophageal reflux and swallowing problems or other malformations such as laryngeal cleft. Presenting symptoms include pneumonia, coughing while feeding, and cyanosis.[2]

Esophagram is the modality of choice for diagnosis of H-type tracheoesophageal fistula. Frequently, a

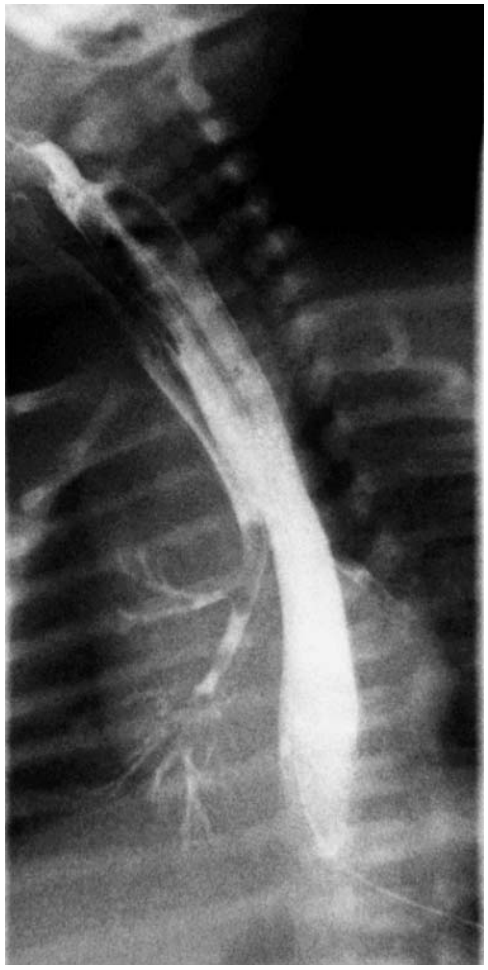

Figure 1. Standard contrast material swallow esophagram demonstrates oral contrast within the tracheobronchial tree. Prior images (not shown) demonstrated no oropharyngeal aspiration.

small fistula may not be visualized by standard esophagram technique. If the suspicion is still high, tracheobronchoscopy is the definitive diagnostic study and should be performed with plans for correction under the same anesthetic if identified. Classically, H-type tracheoesophageal fistulas are located at the level of the thoracic inlet. The esophageal end of the fistula is generally located inferiorly in relation to the tracheal component. Oropharyngeal aspiration may appear similar to contrast seen within the tracheobronchial tree and care should be taken to distinguish the two.

In the setting of high clinical suspicion for H-type tracheoesophageal fistula that cannot be confirmed with standard esophagram, a pull-back tube esophagram can be used to increase the sensitivity for small fistulas. A pull-back tube esophagram is performed by first inserting a nasogastric tube into the patient's

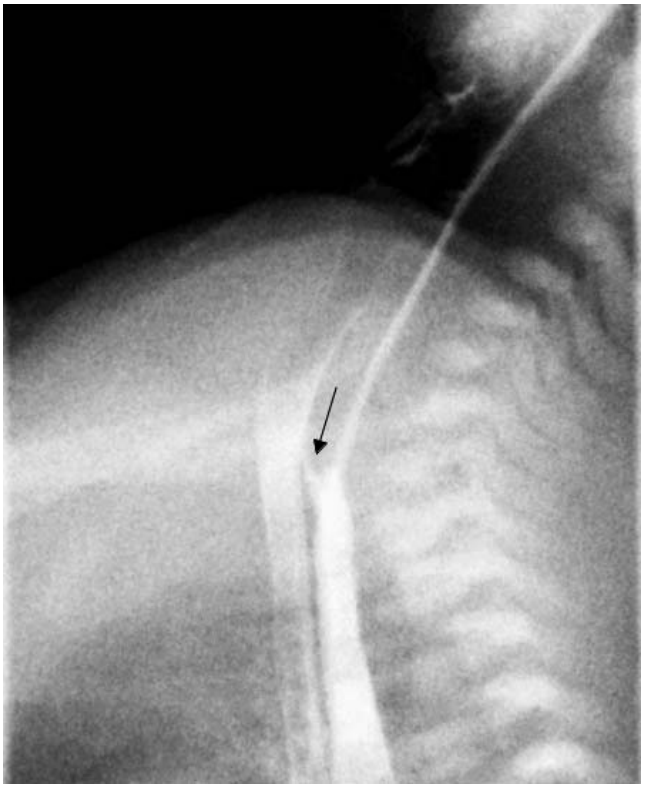

Figure 2. Pull-back tube esophagram demonstrates a small fistulous tract (arrow) at the level of the thoracic inlet between the esophagus and trachea compatible with an H-type tracheoesophageal fistula. Note the inferior location of the esophageal end in relation to the tracheal component.

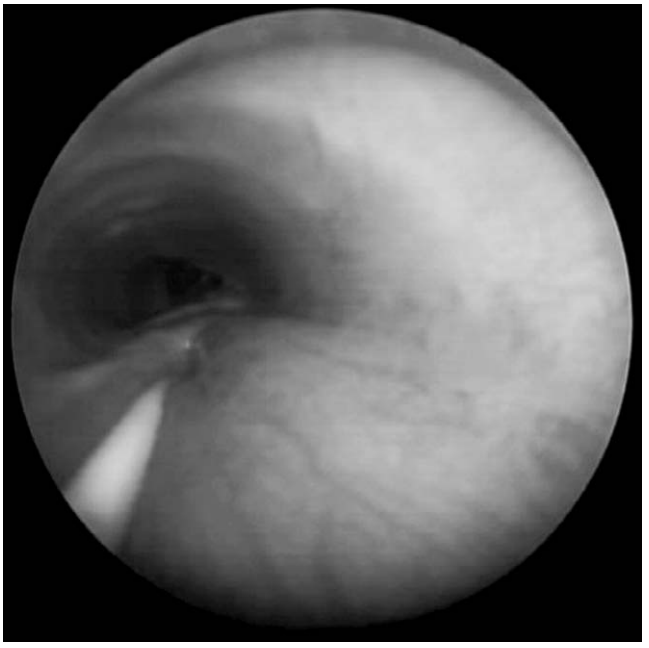

Figure 3. Direct laryngotracheal bronchoscopy with #2 Fogarty catheter placed within the fistula.

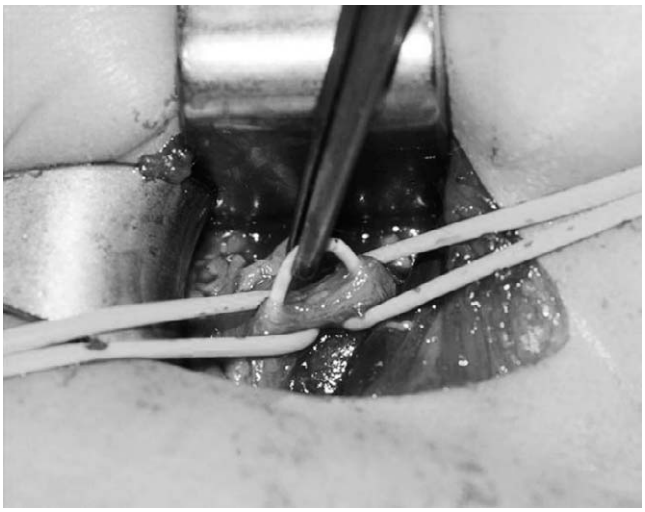

Figure 4. Intraoperative photograph shows fistula tract encircled by 2 vessel loops and Fogarty catheter being removed through an incision in the anterior wall of the tracheoesophageal fistula.

stomach. Contrast material is then injected through the nasogastric tube at different levels of the esophagus. Contrast-enhanced studies have the potential risk of aspiration pneumonia and pulmonary injury. The study should be performed using isotonic water-soluble material and with adequate neonatal emergency resuscitation at hand.[3]

H-type tracheoesophageal fistulas are managed surgically with ligation of the fistula, usually from a transcervical approach. In rare cases, a transthoracic surgical approach is needed, depending on the location of the fistula. For this reason, accurate documentation of the location of the fistula is extremely important for preoperative planning. Long-term complications of tracheoesophageal fistula include recurrent pneumonia, obstructive airway disease, airway hyperreactivity, gastroesophageal reflux disease, and esophageal stenosis.[4]

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