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Case Report

Diagnosis and Management of Isolated Tracheoesophageal Fistula: A Case Report

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Abstract Isolated tracheoesophageal fistula (TEF) or H-type TEF is a rare congenital malformation of the esophagus in which a fistula between the lower trachea and the membranous part of the esophagus may lead to choking precipitated by feeding. Because of its rareness, diagnosis and surgical management of this anomaly can be challenging. A 2-month-old male infant presented with frequent cough and occasional choking during breastfeeding, beginning from his 3 days of life. At 2 weeks of age, the infant developed tachypnea and fever and was diagnosed with aspiration pneumonitis. His symptoms improved after feeding via an NG tube. An esophagogram at the local hospital suspected a tracheoesophageal fistula with a faint shadow of the fistulous tract, 2 centimeters above the clavicular level. With a high index of suspicion, the child was scheduled for a rigid tracheo-bronchoscopy. After the fistula, located 2 centimeters above the carina, was identified and catheterized, a transcervical division of the fistula was performed. The child had an uneventful postoperative course and could catch up with standard growth within six months of follow-up.

Keywords: Tracheoesophageal fistula, Esophageal malformation, H-type

Introduction

Tracheoesophageal fistula (TEF) is a rare congenital anomaly characterized by abnormal communication between the trachea and the esophagus. It occurs in approximately 1 in 2,500 to 4,500 live births.[1-3] The most common type of TEF is esophageal atresia, with a distal tracheoesophageal fistula accounting for around 85% of cases.[4] Isolated tracheoesophageal fistula, without esophageal atresia, is a much rarer entity, representing less than 5% of all tracheoesophageal malformations.[5,6] Vogt's classification of tracheoesophageal fistula (1929) classified this type as Type 4, later re-classified in the Gross schema (1953) as Type E. However, considering its anatomical feature, it is known as H-type or N-type TEF.[6]

H-type TEF typically presents respiratory symptoms such as coughing, choking, or cyanosis during feeding. These symptoms are often subtle and can be mistaken for other respiratory conditions, leading to delays in diagnosis.[7] Diagnosing H-type TEF can be challenging

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and often requires a combination of imaging studies and endoscopic procedures.

In this case report, we present a 2-month-old infant with an isolated tracheoesophageal fistula who presented with an intermittent cough during feeding. We discuss this rare condition's diagnostic challenges, surgical management, and postoperative outcomes. This case highlights the importance of considering H-type TEF in the differential diagnosis of infants with respiratory symptoms, particularly those associated with feeding.

Case Presentation

A 2-month-old boy presented with a history of intermittent coughing and vomiting during feeding, beginning at 3 days of age. His gestational age was 38 weeks. He occasionally experienced cyanosis during coughing episodes but did not have abdominal distension or dysphagia. At 2 weeks of age, he developed dyspnea following a coughing episode, and the severity of his cough progressively worsened. He was initially taken to a primary-care hospital, where he was diagnosed with aspiration pneumonitis and treated with intravenous antibiotics. Due to suspicion of a tracheoesophageal fistula (TEF), an esophagogram was performed, which revealed the presence of contrast material in the trachea and bilateral bronchial trees during swallowing, suggestive of a possible thin TEF tract fistula at the lower cervical level above the thoracic inlet (Figure 1). The patient was treated with intravenous antibiotics for aspiration pneumonia at the primary-care hospital for 2 weeks, then was referred to our hospital at 2 months of age for further evaluation and management.

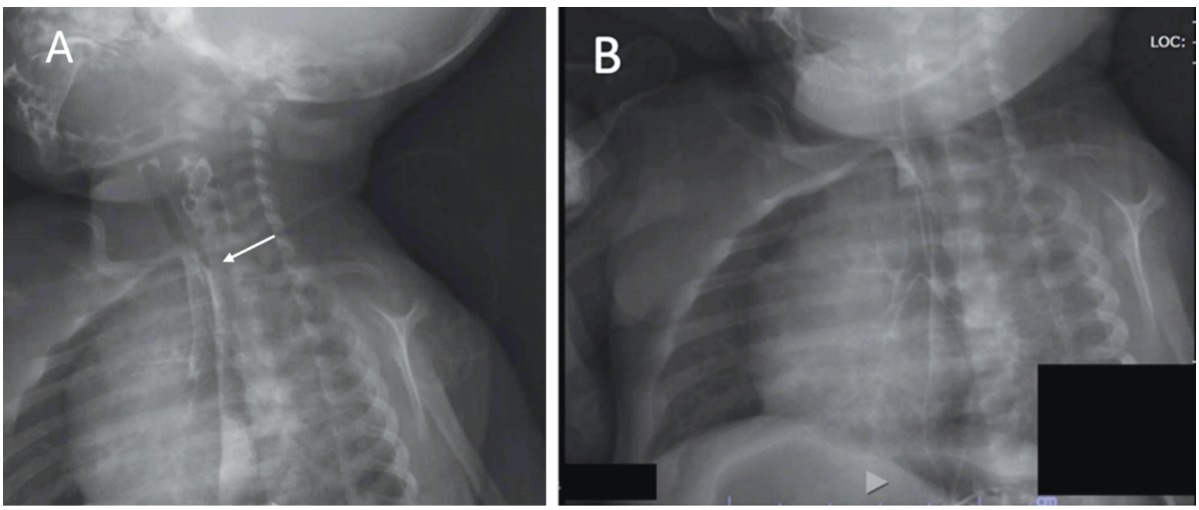

Figure 1 Water-soluble contrast esophagograms showing A) the presence of contrast in the trachea and B) the bronchi. The suspected fistula was at the lower cervical level, above the thoracic inlet (white arrow).

On admission to our hospital, the patient was in stable condition, afebrile, and without dyspnea. He was maintained on nasogastric (NG) tube feeding to prevent aspiration pneumonia. His body weight was 4.4 kg (< 3rd percentile for Thai children), and his height was 54.5 cm (< 3rd percentile for Thai children). A review of the previous esophagogram revealed inconclusive findings, and a repeat esophagogram was not performed to minimize the risk of aspiration. A rigid tracheo-bronchoscopy was scheduled in the operating room with a standby for surgical intervention if a TEF was identified. The tracheo-bronchoscopy was performed by the ENT team and revealed a tracheoesophageal fistula located 2 centimeters above the carina, near the cervical esophagus. A guidewire was inserted through the fistula from the trachea (Figure 2) to facilitate intraoperative identification. After the fistula was identified and catheterized, an esophagoscopy was performed by the Pediatric surgical team to confirm the tip of the wire.

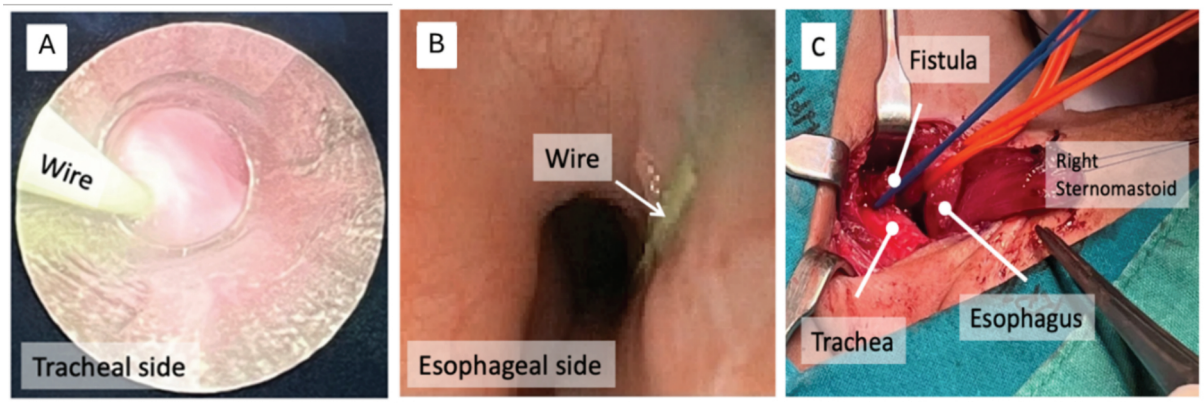

Figure 2 Endoscopic and operative findings in this patient. A) The wire was passed via a rigid tracheo-bronchoscopy. B) An esophagoscopy showed a wire within the esophageal lumen C) A craniocaudal view of the operative field, showing the fistula identified by palpating the wire.

The NG tube was inserted, and the right cervical approach was utilized for the surgical division of the TEF. The esophagus and trachea were identified (Figure 2) and carefully dissected to locate the fistula. The TEF was successfully divided, and the tracheal side was closed with a vertical polypropylene suture line. The esophageal side was closed with a transverse suture line using absorbable polyglactin sutures (Figure 3). An interposition muscle graft from the sternocleidomastoid muscle was placed between the esophagus and trachea to minimize the risk of recurrence.

The patient's postoperative course was uneventful. He has had an indwelling NG tube for 7 days, and the feeding via the NG tube was started one day postoperatively, and oral feeding was initiated on postoperative day 8. He was extubated on postoperative day 3 and discharged home on postoperative day 10. Searching for associated anomalies by urinary tract ultrasonography reported negative results. At the follow-up visit, his body weight had increased to 6 kilograms one month after surgery (a gain of 1.6 kg) and 9 kilograms six months after surgery (a gain of 4.6 kg). He also tolerated oral feeding without difficulty.

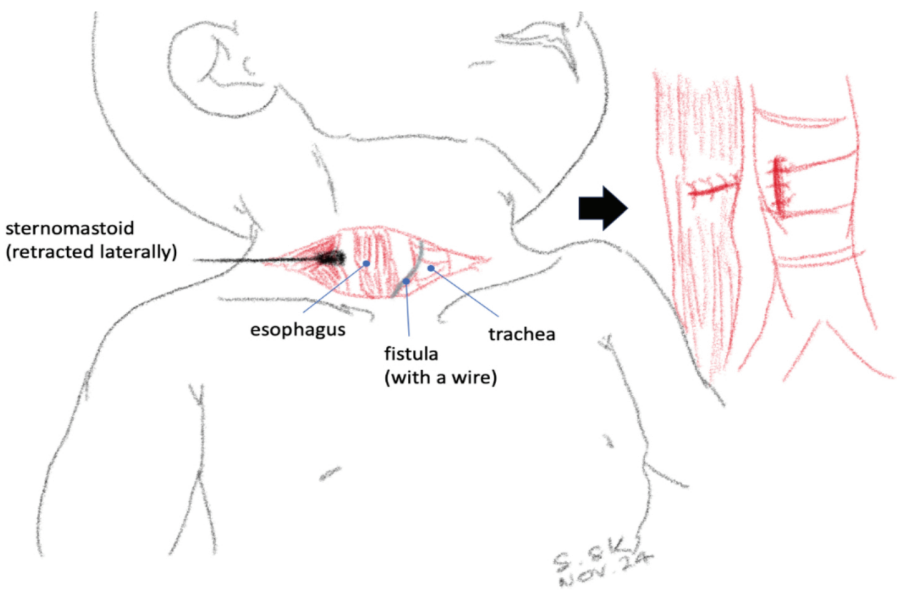

Figure 3 Drawing pictures explaining the surgical anatomy of this patient. Through a transverse incision on the right neck above the clavicle, the esophagus was identified by retracting the right sternomastoid to the lateral side. The fistula could be identified by palpating the wire passed from the trachea to the esophagus. Note that the suture lines on the trachea and the esophagus were not in the exact alignment, which reduced the chance of recurrence.

Discussion

An isolated tracheoesophageal fistula typically presents with cough or choking with oral feeding, especially with liquid meal. Aspiration pneumonitis can result from the bronchial aspiration of the food content. Although our patients presented with those classic symptoms of tracheoesophageal fistula, other differential diagnoses of infant aspiration, including gastroesophageal reflux disease (GERD), oropharyngeal incoordination, esophageal web, and foreign body obstructing the esophagus should be ruled out. Isolated TEF can present in late childhood or even in an adult with recurrent pneumonia.[8,9]

When congenital tracheoesophageal fistula was suspected, the video-fluoroscopic esophagogram was a modest initial radiologic investigation to simultaneously study esophageal anatomy and physiology. Fortunately, the study demonstrated a fistula between the trachea and the esophagus, just above the clavicle. It should be emphasized that the contrast used in the study should be low osmolar water-soluble contrast to prevent a risk of lung injury caused by aspiration during the study.[10,11] Before the surgical management, searching for associated congenital anomalies, such as those in the cardiovascular, genitourinary, vertebral, and gastrointestinal systems, was part of the evaluation for the VACTERL association.[12,13]

Identification of the fistula is crucial to operative planning, as a low-lying fistula below the thoracic inlet requires a thoracotomy. Although a contrast swallowing esophagogram gave a sensitivity of TEF detection up to 63-87% in some studies, there was a report of a fistula missing in more than 50% of a series.[7,14] In cases where the TEF cannot be demonstrated on contrast esophagography, multiple repetitions of the study or the use of special techniques that may enhance the reflux of contrast into the fistula, such as pulling back tube esophagogram or distal balloon technique, were reported.[15] Moreover, the infant's position is important; they should be placed in the prone Trendelenburg position to increase diagnostic sensitivity.[13] With an awareness of the limitations of the radiologic study, direct visualization of the fistula by an endoscopy study is recommended. An esophagoscopy alone has even lower sensitivity in fistula detection because the opening in the esophageal side is usually small, subtle in the mucosa, and caudal to the tracheal side (Figure 2). A combination of rigid tracheo-bronchoscopy and esophagoscopy is the current gold standard in fistula identification.[7] Rigid tracheo-bronchoscopy is the most suitable equipment for identifying TEF and passing a guide wire through the fistula. In our case, an esophagogram revealed the presence of contrast material in the trachea and bilateral bronchial trees during swallowing, suggesting a possible thin TEF tract at the lower cervical level above the thoracic inlet. However, it could not definitively diagnose TEF, so a double endoscopy technique was used to confirm the diagnosis and pass a guidewire through the fistula. This helped determine the surgical approach and localization of the fistulous tract during surgical exploration. An alternative technique to double endoscopy is the use of real-time fluoroscopy to detect the wire passed through the tracheal side to the course of the esophagus and stomach.[13] Although the investigation suggested that the pathology is above the thoracic inlet, backup preparation for a thoracotomy is advisable.

We chose a transverse incision on the right clavicular fossa parallel to the clavicle for cosmetic purposes. The esophagus was approached by retracting the right sternomastoid laterally. Care should be taken not to injure the vagus nerve and the recurrent laryngeal nerve. Choices of fistula occlusion are division, clipping, or ligation. However, a nationwide survey from Italy reported that clipping or ligation was associated with a higher recurrence rate.[7] It could be emphasized that the suture lines on the trachea and the esophagus should not be in the same direction to reduce anastomotic kissing that might cause recurrence. An intercalation with a muscle flap is a good technique to reduce this complication. As a low-pressure contrast study has limited sensitivity, most surgeons do not perform a routine esophagogram during the postoperative period.[7] Potential postoperative complications include stridor and hoarseness caused by neurapraxia of the recurrent laryngeal nerve, which is usually transient. However, some institutes recommended a routine laryngoscopy after surgery.[7] In our case, the patient had no postoperative complications and could drink milk, so a routine postoperative esophagogram was not performed.

In conclusion, we presented a case of isolated TEF successfully corrected with surgery. Although the esophagogram was not very clear, a high index of suspicion led to an endoscopy and localization of the lesion. The fistula was successfully managed using the cervical approach division.

Ethical Approval

In this case, the Human Research Ethics Committee of the Faculty of Medicine, Prince of Songkla University, has approved the report and use of clinical materials.

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