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FULL PAPER

Exploring h-type tracheo-esophageal fistula: Unveiling variations in three cases– a detailed case report and insightful discussion

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H-type tracheo-esophageal fistula (HTEF) is a rare congenital anomaly characterized by a connection between the tracheal wall and anterior esophageal wall. HTEF occurs in approximately 4% of esophageal atresia. HTEF has unspecific symptoms, including cough, cyanosis during feeding, recurrent pneumonia, which contributes to delayed diagnostic, potentially leading to complications like aspiration pneumonia and respiratory distress. Airway management in these patients presents additional challenges, as inadvertent esophageal intubation is a plausible risk. This underlines the critical importance of an in-depth overview of HTEF to enhance awareness, early identification, and improve the overall management of this rare congenital anomaly. Increasing awareness among healthcare professionals is essential to facilitate early diagnosis, optimize patient care, and mitigate the risk of complications arising from delayed intervention. This case series will discuss about 3 cases of H-type tracheo-oesophageal fistula which is found accidentally.

KEYWORDS

Congenital anomaly; H-type TEF; tracheooesophageal fistula.

Introduction

Congenital tracheoesophageal fistulas (TEFs) are rare, occurring in approximately 1 in 2500 live births [1,2]. Type V ("H-type") is the rarest, consisting of an isolated TEF without esophageal atresia, constituting about 4% of all congenital TEFs [3]. H-type tracheo-oesophageal fistula (HTEF) is a rare congenital anomaly characterized by an abnormal connection between the trachea and esophagus without associated esophageal atresia. Unlike its more prevalent counterpart, esophageal atresia, H-type TEF manifests as an abnormal connection between the trachea and esophagus without accompanying esophageal malformations [4]. This enigmatic condition often eludes early detection due to its nonspecific clinical presentation, leading to potential delays in diagnosis and management [5].

First documented in medical literature, H-type TEF has perplexed clinicians with its rarity and subtle symptomatology [6]. Accounting for a small fraction of tracheo-oesophageal fistulas, ranging from 1.8% to

4.2%, this anomaly demands a heightened clinical suspicion for timely identification [7]. The overlapping symptoms with more common respiratory and gastrointestinal issues pose a formidable challenge in distinguishing H-type TEF from other conditions, contributing to its underdiagnosis [7,8].

The journey to diagnose H-type TEF is fraught with challenges. Its nonspecific presentation, often mirroring symptoms of other prevalent disorders such as gastro-oesophageal reflux disease, complicates the diagnostic landscape and even could be undetected until adulthood [9]. The classical triad of symptoms—severe choking and cough during feeds, abdominal distention, and recurrent pneumonia—may not consistently manifest, adding a layer of complexity to the diagnostic process [3].

Case report

Here we present 3 cases of H-type tracheo-oesophageal fistula which is found accidentally.

The first case

A 9-day-old baby with hydrocephalus who developed abdominal distention while intubated and receiving positive pressure ventilation during ventriculoperitoneal shunt placement. Intraoperative photograph showing distended abdomen after positive pressure ventilation (Figure 1). The baby was subsequently diagnosed with H-type tracheo-oesophageal fistula, which had been overlooked due to its nonspecific symptoms. The patient was then passed away on the operating table.

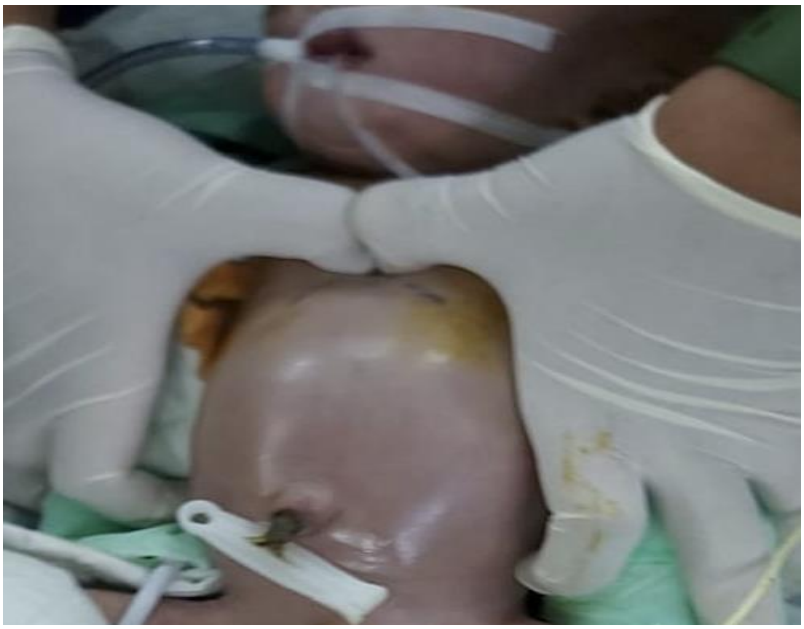

FIGURE 1 Abdomen is distended after positive pressure ventilation

The second case

A 2.5-month-old infant with cleft lip palate and secundum atrial septal defect who was going to undergo cheiloplasty. The infant presented without a history of choking and vomiting while drinking. There was occasional snoring during sleep but no shortness of breath. The patient has been exclusively fed breast milk using a pipette since birth. The initial complaint was loose stools occurring up to 6 times daily with a liquid consistency for the past two weeks. Fever episodes were intermittent and not accompanied by vomiting. After intubated, sound generated by ventilation was heard by

gastric area during abdomen auscultation. Upon further evaluation, it was discovered that the infant also had an H-type tracheo-oesophageal fistula. Eventually the surgical procedure was continued via open NGT and further work up was conducted.

The third case

A one-month-old infant with a serial congenital anomalies including rupture lumbosacral myelomeningocele, tethered cord syndrome, bacterial meningitis, non-communicans hydrocephalus secondary to aquaductal stenosis, left CTEV, small secundum ASD, suspected type 2 Chiari Malformation, and H-type tracheo-oesophageal fistula. Similar to case number 2, the suspicion of HTEF in this patient also arise after intubation, which was then confirmed by diagnostic procedure. Identification of fistula located at the level of T1 vertebra (Figure 2).

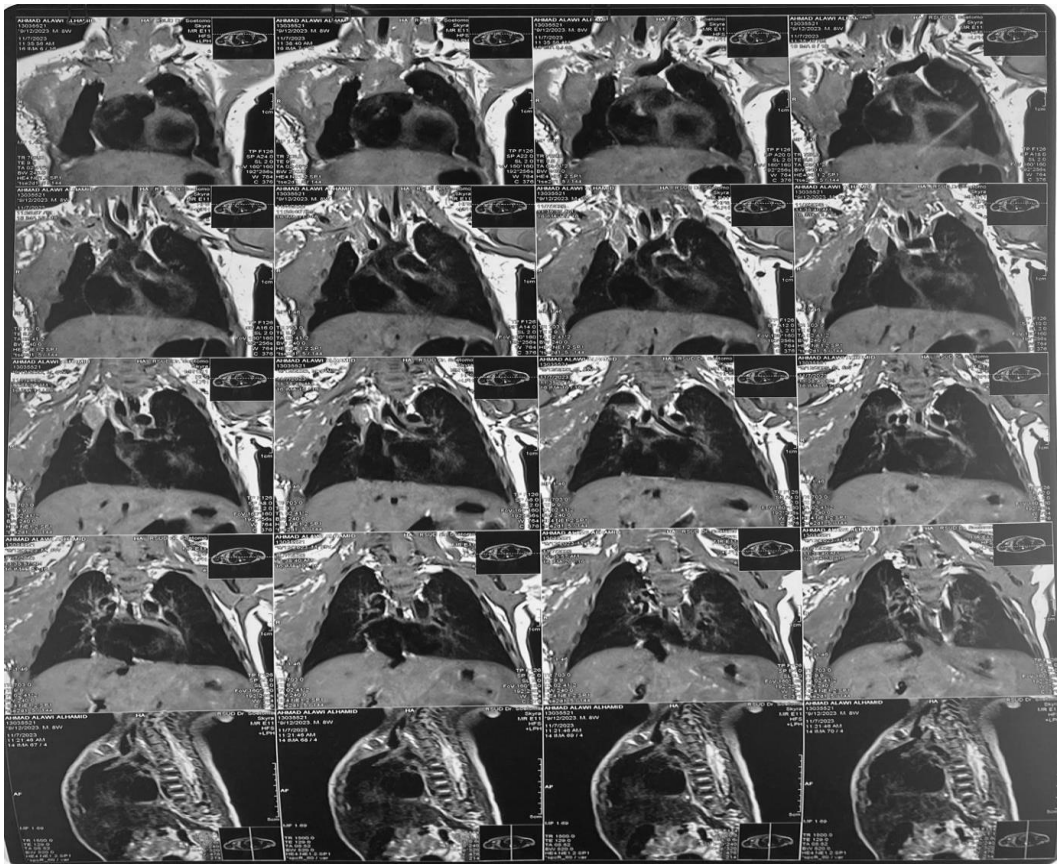

FIGURE 2 MRI shows a congenital tracheo-esophageal fistula type E (H-type) at the T1 level.

Discussion

Diagnosing HTEF poses several challenges due to its nonspecific symptoms and rarity [4]. In addition, the overlap of symptoms with more common medical conditions further complicates the diagnosis [9]. One of the main challenges in diagnosing HTEF is its nonspecific presentation [10]. Pressure changes between both structures can cause entry of air into the oesophagus, or entry of oesophageal content into the trachea [11,12]. Clinical manifestations—cyanosis, cough and choking with feeding, recurrent chest infections and persistent gastrointestinal distension—are related to the presence of the fistulous connection [13]. The symptoms, such as recurrent respiratory tract infections, chronic cough, and choking during feeding, are often mistaken for gastro-oesophageal

reflux disease or other common medical conditions. This often leads to a delay in diagnosis, which can have serious consequences for the affected child [14].

The first case highlights the challenges in diagnosing HTEF due to its nonspecific symptoms. The diagnosis was delayed because the symptoms were attributed to the baby's underlying condition of hydrocephalus [15]. Healthcare professionals need to maintain a high index of suspicion for HTEF and consider it in the differential diagnosis for infants presenting with respiratory symptoms and gastrointestinal issues [13]. On the second case, infant presented with recurring respiratory tract infections and chronic cough, which are typical symptoms of HTEF. A fistula between the trachea's posterior wall and the esophagus's anterior wall allowed food and saliva regurgitation, leading to pulmonary aspiration and subsequent respiratory infections [12]. However, it is important to consider alternative explanations for the presenting symptoms and not immediately jump to HTEF diagnosis while it is true that this condition can present with symptoms such as recurrent respiratory tract infections and chronic cough, these symptoms can also be attributed to other common conditions, such as gastro-oesophageal reflux disease [16].

H type tracheoesophageal fstula without oesophageal atresia is a rare entity seen in 1 in 50,000 to 80,000 live births and accounts for 4–5% of all congenital oesophageal anomalies [17]. With a prevalence of 1.8% to 4.2%, H-type tracheo-oesophageal fistula is uncommon [7]. Due to its rarity, many healthcare providers may not consider HTEF a possible diagnosis, leading to further delays in diagnosis and treatment [10]. In addition, the classical triad of symptoms associated with HTEF may not always be present or recognized, such as paroxysm of severe choking and cough during feeds, abdominal distention, and recurrent pneumonia. Its clinical symptoms are similar to other respiratory disease, such as cyanotic aspiration during feeding, recurrent respiratory symptoms, and abdominal distention [10,18].

Another challenge in diagnosing HTEF is the difficulty in performing diagnostic tests. Early diagnosis of HTEF is crucial to prevent potential complications and ensure appropriate management. Delayed diagnosis of HTEF can lead to serious consequences, such as recurrent respiratory tract infections and aspiration pneumonia [3,9]. Early diagnosis of HTEF is crucial in guiding appropriate management, preventing further complications, and improving outcomes [11]. However, it is important to consider an opposing argument when discussing the diagnosis and management of HTEF. While early diagnosis is desirable, balancing unnecessary interventions' potential benefits and harms is crucial [4,19]. In previous reports, the most commonly used methods to detect H-type TEFs have been the barium swallow test, followed by bronchoscopy and chest CT scans. The least common methods are three-dimensional airway reconstruction, esophagoscopy, salivagram, and MRI [12].

Both overdiagnosis and underdiagnosis is a problem in diagnosing HTEF. Over diagnosis commonly related to the non-specific nature of the symptoms which potentially lead to unnecessary medical procedures, such as invasive diagnostic or surgical procedures, which may carry further risks and complications [20,21]. On the other hand, there is a risk of under-diagnosis because of the HTEF rarity and overlapping with other common medical conditions. This can result in delayed treatment and potentially worsen the prognosis for affected infants [10].

Delayed diagnosis and treatment can lead to recurrent respiratory tract infections, chronic lung damage, malnutrition, and even death [6]. Therefore, healthcare providers must maintain a high suspicion for HTEF in infants presenting with nonspecific symptoms such as chronic cough, recurrent respiratory

infections, and feeding difficulties. To improve the diagnostic process, healthcare providers should consider conducting further investigations such as imaging studies like contrast esophagram or bronchoscopy [13].

High rates of co-associated anomalies were reported with H-type fistulae, with Daniel *et al.* noting 50% of their cohort having another abnormality [22,23]. The most commonly reported anomalies were cardiac lesions, followed by allied disorders such as VACTERL syndrome (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fstula, renal anomalies, and limb abnormalities) and CHARGE sequence (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities), as found in the third cases [14,15].

However, it is worth considering that not all cases of HTEF present with the classical triad of symptoms [4]. Most cases are mild and may go undetected during the neonatal period, only to be diagnosed in adulthood [9]. This raises questions about the true prevalence and significance of HTEF as a condition. The rarity of this condition also contributes to the challenges in diagnosis. Healthcare providers may not initially consider HTEF a possibility, leading to delays in diagnosis and treatment [16].

In cases of suspected type HTEF found during mechanical ventilation, we recommend stopping mechanical ventilation, inserting an NGT, waking the patient and continuing with supporting examinations because providing positive pressure ventilation for type H TEF may harm the patient [24-26].

Conclusion

H-type tracheo-oesophageal fistula is a rare congenital anomaly that can be difficult to diagnose due to its nonspecific symptoms and rare incidence. Early diagnosis is crucial to prevent complications and improve outcomes for affected infants. Clinician need to have a high index of suspicion based on the presenting symptoms. While early detection may be beneficial in some cases, weighing the potential risks and benefits is essential to ensure appropriate intervention. In addition to a thorough clinical evaluation, further investigations can help confirm the diagnosis. Early diagnosis is crucial for proper management and prevention of complications. Imaging studies such as contrast esophagram or bronchoscopy can provide visual evidence of the fistula and help guide treatment decisions.

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Authors' Contributions

All authors contributed to data analysis, drafting, and revising of the paper and agreed to be responsible for all the aspects of this work.

Conflict of Interest

The authors declare that they have no conflict of interest in this study.

Data Availability

The article contains all the necessary data to support the results; no supplementary source data is needed.

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