An uncommon presentation of double H type of tracheoesophageal fistula

Abstract
An eight-year-old child came with complaints of recurrent pneumonia since early childhood. At birth he was diagnosed to have Tracheo-esophageal fistula (TEF), for which he was operated. He was treated as asthma without much relief. Computed tomography of thorax showed Double H type of fistula with pneumonia and bronchiectasis.

Keywords: double H type TEF, asthma, recurrent pneumonia, bronchiectasis

Introduction
Tracheoesophageal fistula (TEF) is an abnormal connection between the trachea and the esophagus. Most TEF abnormalities are diagnosed at birth since 98% of them are associated with esophageal atresia. The incidence is approximately 1 in 4000 live births and in some cases, it may associate with VACTERL (Vertebral, Anal, Cardiovascular, Renal and limb) anomalies. H type of tracheoesophageal fistula is rare and comprises only 4% of the cases of TEF. In the H type, the fistula runs from the posterior wall of trachea downwards to the anterior wall of esophagus. Double H type of fistula is even less common with only few cases reported. There is no associated esophageal atresia. These cases are difficult to diagnose early because of nonspecific symptoms. Untreated it leads to multiple complications such as recurrent pneumonia, lung abscess, ARDS, acute lung injury, poor nutrition, bronchiectasis from recurrent aspiration, respiratory failure and death. Here we present a case of an eight-year-old who presented with recurrent lower respiratory tract infection mimicking asthma and was found to have double H type of TEF.

Case report
An 8-year-old boy came with history of recurrent cough, breathing difficulty since he was 8 months of age. Present episode was for 5 days with fever, purulent sputum and breathlessness with wheeze. The child came for evaluation of refractory asthma to pulmonary physician. His mother had polyhydramnios in pregnancy. She delivered him by full term normal vaginal route. Following birth, he had tachypnea and breathing difficulty and nasogastric tube could not be passed. He was diagnosed with Tracheo-esophageal fistula and was operated. Surgical closure of upper fistula was done by pediatric surgeon. Post-operative period was uneventful.

He was then asymptomatic for 8 months. At 8th month of age he was first diagnosed with pneumonia, admitted and treated with antibiotics. After 1 year of previous episode he again had respiratory infection for which he was treated. Thereafter, since 5 years of age he had been having recurrent episodes of cough with purulent sputum, breathlessness with wheeze every 2 months for which he was diagnosed to have Asthma and treated with inhaled bronchodilators and inhaled steroids with not much relief. On examination, he was febrile with pulse oximetry saturation 90%. He had tachycardia and tachypnea. He was poorly built for his age. Respiratory examination showed bilateral diffuse polyphonic rhonchi and coarse crepitations. Other system examination was within normal limits.

Since the above presentation was not typical of Atopy and Asthma, Computed Tomography of thorax with contrast was asked. It showed presence of Tracheo-oesophageal fistula (TEF) with bilateral pneumonia and bronchiectasis. Double H type of fistula was seen with upper fistula forming a pouch and the lower fistulous connection persisting between trachea and esophagus. Diagnosis of recurrent respiratory infection secondary to TEF was made. This case is unique because recurrence of TEF post-surgical correction is uncommon. It also highlights the importance of a detailed birth history and its clinical correlation.

Discussion
TEF occurs due to abnormal development of Tracheo-oesophageal ridges in fetus. There is failure of lateral septation of the two tracts, namely digestive and respiratory, resulting in formation of abnormal communication between the two. It usually occurs between fourth to fifth week of development. It was first described in 1929 by Negus. It was then an incidental postmortem finding in an adult who died of lung carcinoma. Isolated H type of TEF is a rare disorder and is hence challenging to the diagnose. H type of fistula is not associated with esophageal atresia. If the defect is large, then these cases present early with respiratory symptoms due to aspiration and abdominal
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