CASE REPORTS

H-TYPE TRACHEOESOPHAGEAL FISTULA IN A CHILD WITH DYSMORPHISM: VATER OR NOT?

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ABSTRACT

Tracheo-Esophageal fistula (TEF) with or without esophageal atresia occurs in approximately 1 in 3500 births. In around half of the cases there are associated anomalies while esophageal atresia/TEF occurs in isolation in the remainder. Congenital H-type tracheoesophageal fistula (TEF) in adults is a rare presentation and can test the diagnostic acumen of a surgeon, endoscopist, and the radiologist. These undetected fistulas may present as chronic lung disease of unknown origin because repeated aspirations can lead to recurrent lung infections and bronchiectasis. Congenital TEFs should be considered in the diagnosis of infants and young adults with recurrent respiratory distress and/or infections. Here, we present the successful management of this rare case in an adult patient.

Congenital H-type TEF in children is a rare presentation and search revealed no such case reported in local literature. Here, we present the diagnosis and successful management of a child with congenital H-type TEF with vertebral and limb defects.

Keywords: Adult, Congenital tracheoesophageal fistula, H-Type Tracheoesophageal fistula, Late presentation, VACTERAL, VATER.

INTRODUCTION

Congenital TEFs are unlikely to be missed at birth as in more than 95% of the cases they are associated with esophageal atresia. However, the H-type TEFs can be missed in infancy if high index of suspicion is not kept for repeated chest infections, bouts of cough and recurrent pneumonias[1]. Presentation of H-type TEFs in later life is very rare, with only 16 cases described in adults in medical literature[2]. Here, we report a case of H-type TEF in a 10 year old boy with skeletal and vertebral anomalies but not all features of VACTERAL or VATER syndrome.

CASE REPORT

A 10 years old boy was brought to hospital with history of persistent cough and fever since 3 months of age with hardly any period of wellbeing in between. Cough was productive, episodic, usually triggered by taking meals, especially liquids and frequently resulting in vomiting. Sputum was mucopurulent, foul smelling but easy to expectorate. He was repeatedly hospitalized since infancy for chest infections and was given repeated courses of oral and parenteral antibiotics. Though there was partial relief with medications but symptoms used to reoccur after some time. However, there was no history of prolonged fevers, night sweats or diarrhea and no family history of tuberculosis or asthma.

Old documents review revealed multiple consultations from general practitioners, pediatricians, adult physicians and pulmonologists and he had been investigated for gastro-esophageal reflux disease (GERD), tuberculosis, foreign body lung, cystic fibrosis, HIV infection, immune deficiencies, congenital heart disease, and interstitial lung diseases. He had undergone repeated imaging studies of chest which included x-rays, computerized tomography and magnetic resonance imaging scans, which were unremarkable. He had been treated for GERD

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throughout infancy, had taken 9 months anti tuberculosis treatment twice at 4 and 7 years of age, had taken multiple antibiotic courses and was on intermittent oral and inhaled bronchodilators / corticosteroids, mucolators, nebulizing agents and leukotriene inhibitors till date.

On examination he was afebrile, had cough without respiratory distress and was maintaining saturations above 94% in air. His height (19 kg) and weight (116 cm) were well below 3rd centile and frontal-occipital circumference was 47 cm. He had left sided torticollis and congenitally absent left thumb (fig-1). Auscultation revealed bilateral fine crepitations and rhonchi with normal heart sounds and no cardiac murmur. Rest of the examination was unremarkable. Base line investigations revealed normal cell counts and Chest x-ray showed fine reticular shadowing, normal heart size and right sided scoliosis of lower cervical spine (fig-2). Though he had congenital thumb hypoplasia and cervical scoliosis but no other associations of VACTERAL or VATER syndrome were found clinically or radio-logically.

communication, as trachea and carina was outlined (fig-2). Unfortunately, procedure could not be completed due to intractable cough and thus fistula tract could not be outlined. Both rigid and flexible bronchoscopy was carried out under general anesthesia and fistula opening was

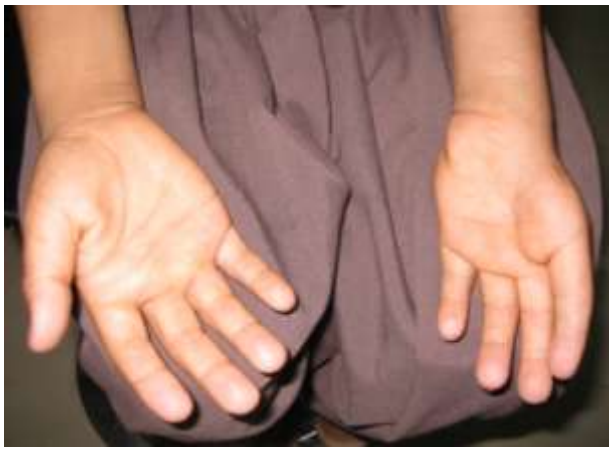

Figure-1: Left thumb hypoplasia.

located in posterior wall of trachea 16 cm distal to the incisors (fig-3). Thus final diagnosis of H-type TEF was made and thoracic surgeon's consultation was sought.

Patient was admitted and put on broad spectrum intravenous antibiotics. Oral feeding

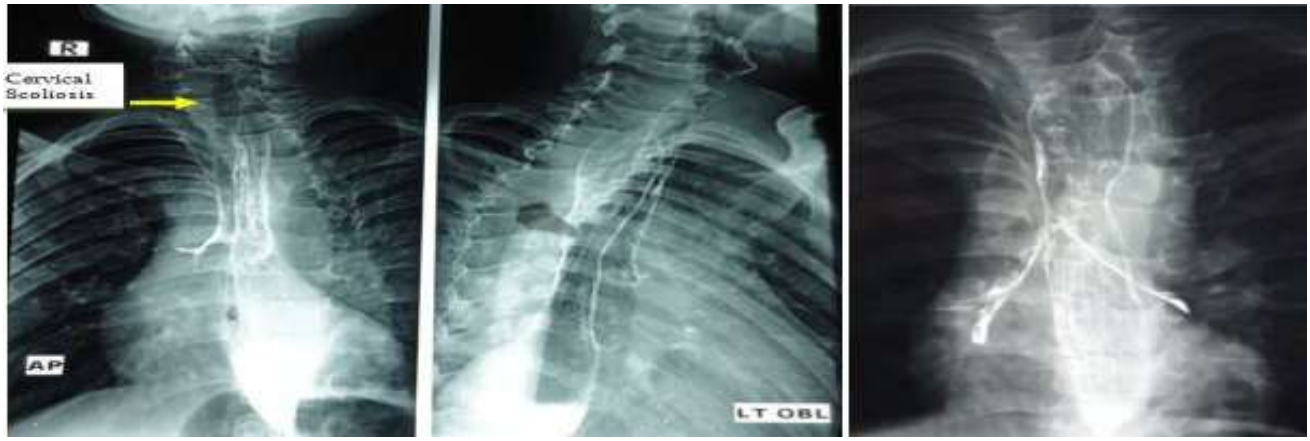

Figure-2: Barium swallow showing outlining of trachea due to barium spill through fistula tract and right sided scoliosis of lower cervical spine.

This symptomatology of disease onset in early infancy, recurrent chest infections and association of bouts of cough with oral intake prompted us to consider H-type TEF. Barium swallow under fluoroscopy was done which confirmed the presence of a definite

was stopped 48 hours before surgery to avoid aspiration. Patient was operated under general anaesthesia through a cervical approach. Esophagus was approached and fistula was located on the right side beneath the upper part of the mediastinum (fig-4). Fistula was divided

and repair of esophagus was done in 2 layers with interrupted sutures of vicryl 3/0. Repair of esophageal mucosa and muscularis was done separately. Tracheal repair was done in single layer and strap muscle was interposed in between the repair.

Post Op recovery was uneventful. Oral feeding was started on 5th post Op day and patient was discharged on 8th post Op day. Four weeks follow up showed a weight gain of 1.5 kilograms and there were no cough or any respiratory symptoms at rest or with oral intake.

DISCUSSION

H-type fistula is a rare, isolated form of TEF[3]. H-Type TEFs usually present in infancy with recurrent respiratory symptoms, coughing, cyanosis and aspiration syndromes[4]. However, early diagnosis of this disorder is occasionally missed and a small number may remain undiagnosed until adulthood[2,5]. These undetected fistulas usually present in late childhood or adulthood with chronic lung disease and bronchiectasis due to repeated aspirations[2].

greater tolerance for solid foods than liquids in these patients[7]. This might be explained by the oblique course of the fistula with tracheal orifice usually proximal to esophageal orifice resulting in occlusion of fistula's lumen due to compression of anterior esophageal wall against the fistula by food bolus. A chronic cough aggravated by heavy meals and recurrent chest infections are common presenting symptoms as seen in our patient. Bouts of coughing when swallowing liquids are pathognomic for this condition[8]. Similar presentation prompted us to consider H-type TEF in our patient.

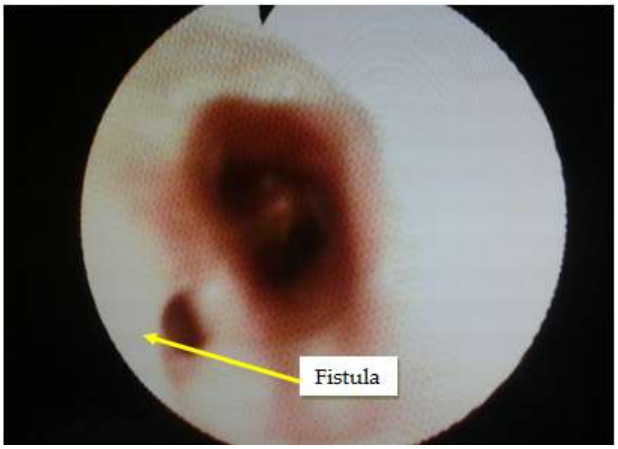

Figure-3: Tracheal opening of fistula.

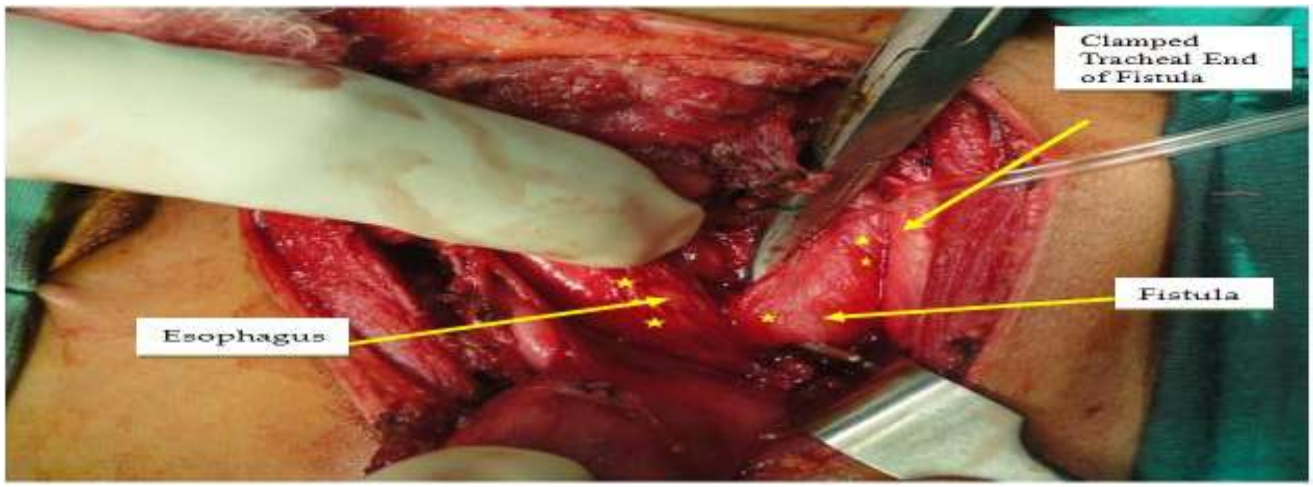

Figure-4: Per-operative details showing esophagus(***), fistula(*) and the tracheal end of fistula(**).

Congenital H-type TEFs represent the least severe form of abnormal trachea-esophageal communication and its presence in later childhood or in adults is uncommon to the extent that no one surgeon or institution is likely to encounter more than a few cases[6]. The symptomatology is usually trivial and there is

Radiological investigation of choice is barium swallow under fluoroscopy which highlights the fistula's tract. Same was employed in our case. A CT scan of the chest may define the

extent of irreversible damage of the lung parenchyma and can also tell the exact location of the fistula. Bronchoscopy and Esophagoscopy can confirm the diagnosis by demonstrating the fistula opening[5]. Bronchoscopy also helps in identifying the exact location of the opening with respect to the carina or the vocal cords which helps in deciding whether or not a cervical approach for fistulectomy is appropriate. Esophagoscopy also helps for direct visualization of TEF fistulas, but occasionally they can be missed as they are located in the upper 3rd and on the anterior wall[9]. Esophagoscopy was not employed in our patient as the facility was not available at our Centre.

The definite treatment is surgical repair and closure of the TEF[10]. Different surgical approaches have been described. For proximally located fistula cervicotomy is done and in cases of distal fistulas, located at the level of the carina, thoracotomy is usually preferred[2]. Right sided cervicotomy was done in our patient.

H-type TEF is associated with other malformations in about 30% of cases which include VACTERL/VATER syndrome, CHARGE syndrome, Goldenhar syndrome, esophageal stenosis, and syndactyly[9]. Though our patient had congenital thumb hypoplasia and cervical scoliosis in addition to TEF but other associations like anal atresia, cardiac defects or renal malformations of VACTERAL or VATER syndrome were not found.

Early diagnosis and treatment help in preventing the long-term debilitating respiratory symptoms associated with the fistula. Delay in diagnosis is usually related to the minute symptoms in some patients, low index of suspicion by the physicians, and unsatisfactory radiological methods as suffered by our patient. Keeping a high index of suspicion and timely application of radiological and endoscopic procedures helps in an early diagnosis of H-type congenital TEF.

CONFLICT OF INTEREST

This study has no conflict of interest to declare by any author.

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