

TRACHEOESOPHAGEAL FISTULA - H TYPE: A RARE ENTITY

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ABSTRACT

Tracheoesophageal fistula is a common cause of recurrent pneumonia in childhood. H-type tracheoesophageal fistula is a rare entity with better prognosis if diagnosed earlier. It needs to be differentiated from its mimickers that share similar clinical presentation like cystic fibrosis, tuberculosis, primary immunodeficiency disorders, other congenital anatomical anomalies of airways. Anticipation and prompt recognition can prevent development of complications. Rigid tracheobronchoscopy is considered to be an investigation of choice for confirmation of diagnosis and surgical repair is the main modality of treatment.

KEYWORDS: Tracheoesophageal fistula, TEF, pneumonia, prognosis, treatment.

CASE REPORT

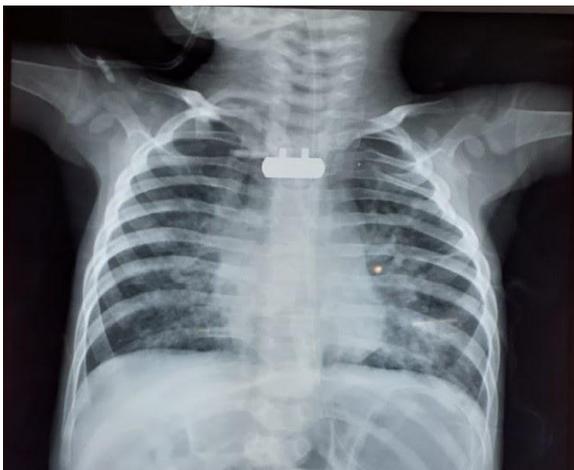
A 1 year male child presented with on and off fever and cough which used to increase after feeds since birth, associated with difficulty in breathing and failure to thrive. The child had history of multiple admissions for the same complaints. There was no history of nasal regurgitation, bluish discoloration, suck rest suck cycle. On general examination the child had frontal bossing, thin lustreless hair, loss of buccal pad of fat with no dysmorphism. Anthropometry was suggestive of acute on chronic malnutrition. The child was febrile, tachypneic with saturation of 92% at room air. The respiratory system examination showed increased work of breathing, bronchial breath sounds with coarse crepts on auscultation. There was no marked improvement in the symptoms of the child despite antibiotics upgradation. Tuberculin skin test, gastric aspirate for Gene Xpert, Covid RT-PCR and sweat chloride test came negative. HIV test of mother and father was done which was also reported negative. Serial chest x-rays showed change in the location of nodular opacities from left to right and then again to left. Cect thorax was suggestive of aspiration pneumonia with a possibility of TEF. Bronchoscopy further confirmed the presence of H-type Tracheoesophageal fistula. The child underwent Bronchoscopy+neck exploration+fistula closure+repair of trachea and esophagus+gastrostomy+bilateral intercostal drainage tube placement. Blood Investigations revealed:

Lab Parameters	18/01/21	01/02/21
Hb	12.2	10.4
TLC	19670	13400
DLC	N68L36	N54L40
Plt	3.65L	2.56L
OT/PT	61.7/36.8	40/23
TB/DB	0.2/0.1	0.2/0.1
TP/Alb	7.2/2.5	7.5/3.0
ALP	182.1	179
Ur/Crt	18/0.3	13/0.3
Na/K	140.7/4.9	137/4.3

Chest X-rays



Day 1 of admission
Nodular opacities in right perihilar region



Day 5 of admission
Nodular opacities in bilateral lower lung fields



Day 11 of admission
Nodular opacities in right perihilar region



Day 17 of admission
Opacities more in left perihilar and lower lung fields

Cect Thorax



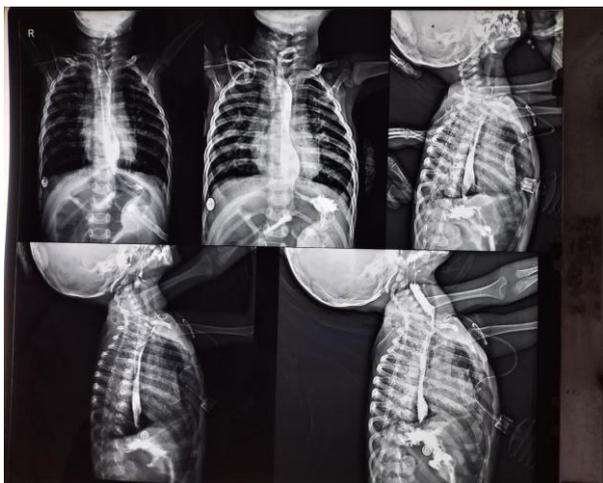
Symmetrical patches of consolidation with surrounding ground glass opacification in posterior segment of bilateral upper lobes



Symmetrical patches of consolidation with surrounding ground glass opacification in superior segment of bilateral lower lobes



Bronchoscopy showing fistula between trachea and esophagus.



Post-operative esophagogram showing no leak.

DISCUSSION

Tracheoesophageal fistula is an abnormal communication between esophagus and trachea with an incidence of 1 in 2500-3000 live births and male: female ratio of 1.26:1. Fifty percent of infants with this condition are non syndromic and the rest have associated anomalies. It's incidence is affected by environmental factors like Methimazole in early pregnancy, maternal diabetes, Thalidomide exposure, chromosomal anomalies like trisomy 18,21. It also has association with syndromes like CHARGE, VACTERAL. The H-type of tracheoesophageal fistula occurs with an incidence of about 4%. The fistula starts from the membranous trachea and runs caudal to enter the esophagus. It is caused by a defect in the lateral septation of foregut into esophagus and trachea and the fistula tract is thought to derive from a branch of embryonic lung bud that fails to undergo branching because of defective epithelial mesenchymal interaction. It is clinically characterized by frothing and bubbling at the mouth and nose and also episodes of coughing, cyanosis and respiratory distress which are exacerbated by feeding. It might come to medical attention later in life with chronic respiratory problems including refractory bronchospasm and recurrent bronchopneumonia. This H-type is more difficult to diagnose clinically. If the fistula is long and oblique, the symptoms may be minimal, and the condition may not be identified for many years. Anticipation and prompt

recognition can prevent development of complications. The diagnosis can be made by insertion of oro/naso-esophageal Replogletube, esophagogram and confirmed by bronchoscopy. Surgical ligation and primary end to end anastomosis of the esophagus is the current standard surgical approach. The prognosis for H-type fistula is generally good. Long term complications following fistula repair in infancy include: dysphagia, GERD with and without esophagitis, respiratory tract infections, asthma, wheeze, persistent cough, Barrett esophagus, esophageal cancer.

CONCLUSION

Tracheoesophageal fistula is a common cause of recurrent pneumonia in childhood. H-type tracheoesophageal fistula is a rare entity with better prognosis if diagnosed earlier. So a high index of clinical suspicion with classical symptoms can help with the diagnosis. Rigid tracheobronchoscopy is considered to be an investigation of choice for confirmation of diagnosis and surgical repair is the main modality of treatment.

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