Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2014; 2(11):755-758 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2014.v02i11.019

Congenital Proximal Tracheo-Esophageal Fistula Clinically Masquerading as Persistent Bronchial Asthma

Trilok Chand[1*], Deepak Rosha[2], K.K. Pandey[3], Kanika Agarwal[4], Rakesh K Yadav[4]
[1]Consultant, Department of Respiratory, Critical care and Sleep medicine, Indraprastha Apollo Hospitals, New Delhi, India
[2]Senior Consultant, Department of Respiratory, Critical care and Sleep medicine, Indraprastha Apollo Hospitals, New Delhi, India
[3]Senior Consultant, Department of Cardiothorasic surgery, Indraprastha Apollo Hospitals, New Delhi, India
[4]DNB Trainee, Department of Respiratory, Critical care and Sleep medicine, Indraprastha Apollo Hospitals, New Delhi, India

***Corresponding Author:**
Name: Dr. Trilok Chand
Email: rao272@gmail.com

Abstract: The congenital tracheo-esophageal fistula is a rare birth defect in which trachea and esophagus are communicated somewhere in their course, which leads to spillage of esophageal contents into airways and develops pulmonary complications. The H-type fistula is third most common type and less likely associated with other birth defects. Our case was a young female who presented with recurrent symptoms compatible with bronchial asthma but on routine investigations like CT-scan and bronchoscopy, she was found to have small tracheoesophageal fistula which was confirmed only by fluoroscopy guided bronchoscopy. She was treated by surgical correction of fistula and on follow-up visits she was found to be symptom free.
Keywords: Bronchoscopy, Fluoroscopy, Tracheo-esophageal fistula.

INTRODUCTION

A tracheoesophageal fistula (TEF) is a congenital or acquired communication between the trachea and esophagus. About half of patients have one or more associated congenital anomalies [1]. The H-type tracheoesophageal fistula is less often associated with other anomalies. During development, abnormal posterior deviation of the tracheoesophageal septum leads to incomplete separation of the esophagus from the laryngotracheal tube and results in a tracheoesophageal fistula [11]. Tracheoesophageal fistula is frequently associated with esophageal atresia; isolated tracheoesophageal fistula is less commonly reported. These patients usually present with episodes of chest symptoms due to recurrent aspiration in lungs.

Our case is a young girl who presented with recurrent episodes of shortness of breath and productive sputum, and she was diagnosed as proximal oblique (N-type) tracheoesophageal fistula which was corrected surgically.

CASE REPORT

A 16 years old female child delivered by full-term normal vaginal delivery, no visible birth anomalies, presented in our hospital with complaints of chronic productive cough and shortness of breath since birth and fever with increased symptoms for 3 days. She had been admitted and evaluated somewhere else and managed as a case of bronchial asthma but she was not relieved. Her parents gave a history of recurrent vomiting and cough after her feed since birth.

On examination she found to be thin built and of average height, with chest crepitations in bilateral basal areas and occasional rhonchi.

Her routine blood investigations showed high total leucocytes count (23400/cmm), normal renal and liver profile with increased total serum IgE (715 IU/ml). The malaria, dengue and H1N1 workup was negative. Patient was started on empirical antibiotics, bronchodilators and other symptomatic treatment.

Her blood and urine cultures were negative. There was no abnormal finding detected in chest X-ray. Patient's PFT was consistent with mild obstructive ventilatory defect with no bronchodilator reversibility.

A HRCT-Chest was ordered which showed bilateral central bronchiectasis with left lower lobe consolidation, esophagomegaly and doubtful

tracheoesophageal fistula (Fig. 1, 2). A bronchoscopy was done which showed a small opening in posterior wall of proximal trachea (Fig. 3). The BAL sample taken from bilateral lung fields grew alpha-hemolytic streptococci and non-pathogenic neisseria.

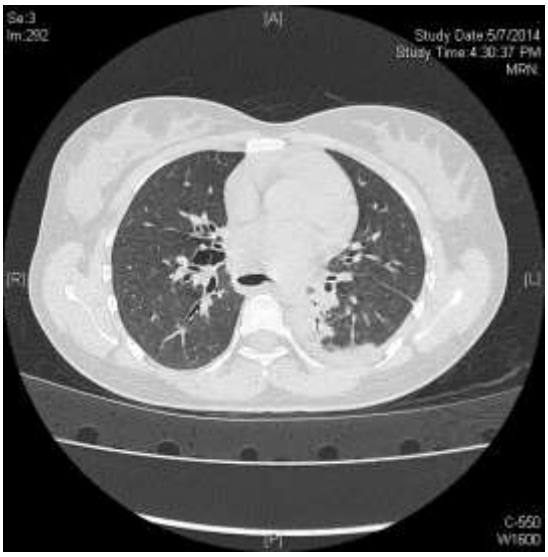

Fig. 1: HRCT shows bilateral central bronchiectasis

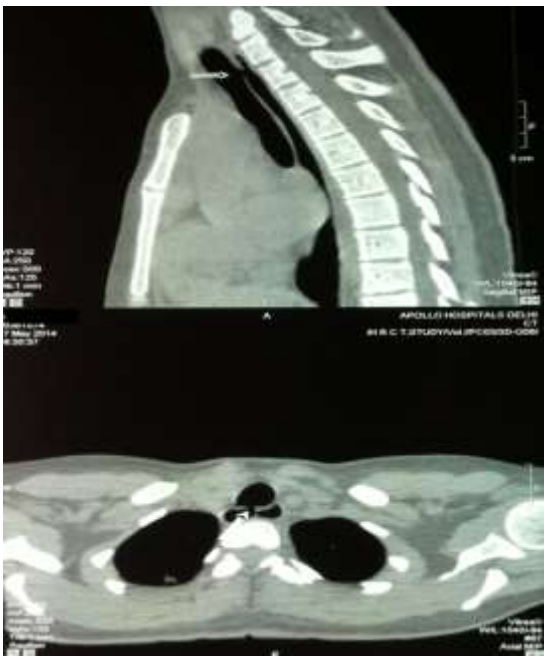

Fig-2: CT-Scan showing TEF left , lower lobe consolidation and esophagomegaly

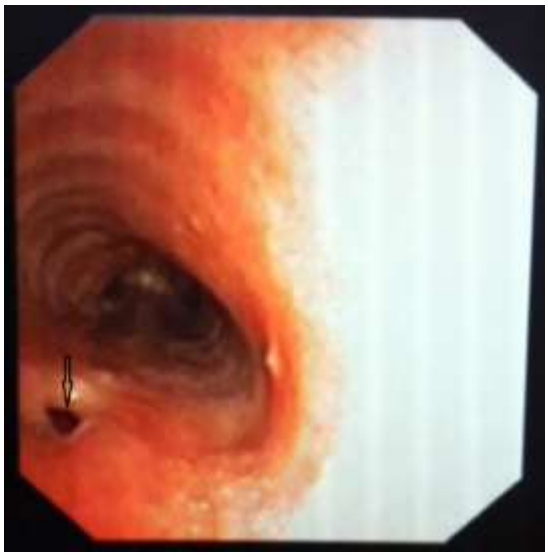

Fig. 3: Bronchoscopy shows a fistulous opening in posterior wall of trachea

For confirmation of tracheoesophageal fistula, upper GI endoscopy was done but it did not reveal any fistulous opening in esophagus (N-type fistula). A gastrograffin study was ordered which was also found negative for tracheoesophageal fistula (Fig. 4). Patient's fluoroscopy guided bronchoscopy was done and a thin catheter with soft tip was inserted through the bronchoscope and advanced in fistulous opening. Dye was injected through the catheter and fluoroscopic images were taken which showed dye to be present in esophagus as well as in stomach (Fig. 5) and this confirmed the evidence of tracheoesophageal fistula. It was a very small and oblique opening in esophagus, which was probably the reason why fistula was, missed both in UGIE and gastrograffin study.

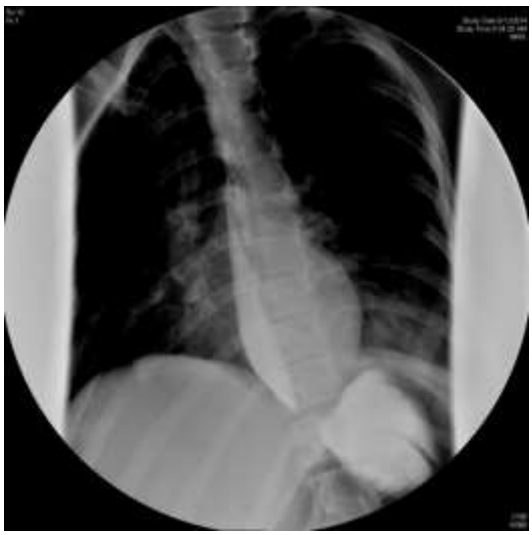

Fig. 4: Normal Gastrograffin study

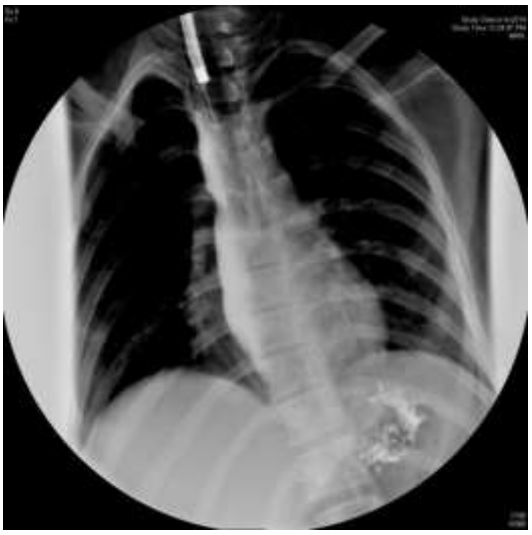

Fig. 5: Fluoroscopy guided bronchoscopy and dye injection into fistula

This patient was managed by surgical repair of tracheoesophageal fistula. Surgery was done by a cervical approach and a muscle patch was applied at fistulous site, patient was kept fasting with continuous Ryle's tube aspiration for a week and nutritional support was given by total parenteral nutrition. After a week her nasogastric tube was removed and gradually she was started on oral liquids and then solid food. She got discharged in stable condition and on follow up visit after one month she did not have any cough and all her other symptoms had also resolved.

DISCUSSION

Congenital Tracheoesophageal fistula (TEF) is a birth defect in which the trachea is connected to the esophagus. In majority of cases, TEF is associated with esophageal atresia and isolated TEF are less in frequency [2]. The esophagus and trachea are derived from the primitive foregut. During the fourth and fifth weeks of embryologic development, the trachea forms as a ventral diverticulum from the primitive pharynx (caudal part of the foregut) [3]. A tracheoesophageal septum divides the foregut into a ventral portion, the laryngotracheal tube and a dorsal portion the esophagus. The incomplete separation of the esophagus from the laryngotracheal tube results in a tracheoesophageal fistula. Incidence of tracheoesophageal fistula along with esophageal atresia is 1 per 2500 to 3500 live births [4, 5]. There is only a minimal hereditary or genetic component with incidence in twins and those with family history being only approximately 1% and there is no gender or racial predilection [5].

There are different types of tracheo-oesophageal fistula with or without esophageal atresia. The most common is proximal esophageal atresia with distal fistula (85%). There may be isolated esophageal atresia (8%), or isolated fistula that is H-type tracheoesophageal fistula (4%). The least common are double fistula with intervening atresia (1%) and proximal fistula with distal atresia (1%) [6, 7]. The H-type tracheoesophageal fistula 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.

The majority of patients with congenital tracheoesophageal fistulas (TEFs) are diagnosed immediately after birth or during infancy, because more than 98% of them are associated with esophageal atresia that results in potential life-threatening complications [8].

Clinically, they present with episodes of coughing, choking and cyanosis, aggravated by heavy meals. Recurrent aspiration leads to frequent chest infections and symptoms related to bronchiectasis.

The diagnosis of esophageal atresia and TEF sometimes requires a high degree of clinical suspicion. Classically, these patients during neonatal or infancy period present with copious, fine, white, frothy bubbles of mucus in the mouth and nose. Radiological workup includes chest X-ray which may be normal or shows some parenchymal shadows related to recurrent aspiration and sometimes esophageal dilatation, CT-scan of chest can reveal extent of the lung parenchymal shadows and is helpful in localizing the fistula. Bronchoscopy helps to visualize the fistula directly and exact localization of its origin with respect to the vocal cords and carina. Esophagoscopy also helps for direct visualization of TEF fistulas, but occasionally they can be missed on esophagoscopy as they are located in the upper 3 rd and on the anterior wall [9] as in our case it was missed in endoscopy. Gastrograffin studies sometimes used to track the fistulous opening. Interestingly in our case both esophagoscopy and gastrograffin study were negative for trachea-esophageal fistula.

In our case fistula was confirmed only by fluoroscopy guided bronchoscopy and catheter insertion in fistulous opening, which was interesting and probably not mentioned in any other case report or literature.

Treatment of choice for congenital trachea-esophageal fistula is surgical correction. If size of fistula is substantial or associated with esophageal atresia then symptoms are severe and it warrants early surgical correction to prevent pulmonary complications. The fistula is approached mostly through cervical route along the anterior border of the sternocleidomastoid muscle and secondly through thoracotomy when the fistula is located at the level of the carina or when to deal with damaged lung at the same time that might

need pulmonary resection [9, 10]. Other supportive management includes broad-spectrum antibiotics for pulmonary infection.

Patients with small size or oblique course of fistula usually are less symptomatic and present late in their childhood or adult life, but pulmonary complications have already taken place due to recurrent aspiration. Prognosis is often depending on size of the fistula and presence of associated congenital anomalies.

Our case was a young female who had less symptoms but recurrent episodes and she was treated as bronchial asthma. We detected the fistula but it couldn't be confirmed by standard means like esophagoscopy and gastrograffin study. It was diagnosed by a fluoroscopy guided bronchoscopy and catheter insertion, which was probably rarely tried earlier. This patient was treated by surgical correction of fistula and discharged in stable condtion.

CONCLUSION

Congenital tracheoesophageal fistulae are not so common but consequences of TEF lead to common pulmonary complications. Early diagnosis and treatment is paramount to prevent pulmonary complications and growth retardation. Small size fistula sometimes missed by standard diagnostic tools, so we recommend the fluoroscopy guided bronchoscopy in case of high suspicion.

REFERENCES

1. Ein SH, Shandling B, Wesson D, Filler RM; Esophageal atresia with distal tracheoesophageal fistula: associated anomalies and prognosis in the 1980s. J Pediatr Surg., 1989; 24:1055–1059.
2. Moore KL; Before we are born: Essentials of embryology and birth defects. Elsevier Health Sciences Division, Kent, UK, 2002.
3. Moore KL. The respiratory system. In The developing human: Clinically oriented embryology. 4th edition, Philadelphia: Saunders, 1988: 207–210.
4. Greenwood RD, Rosenthal A; Cardiovascular malformations associated with tracheoesophageal fistula and esophageal atresia. Pediatrics, 1976; 57(1): 87-91.
5. Shaw-Smith C; Oesophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: review of genetics and epidemiology. J Med Genet., 2006; 43(7): 545-554.
6. Martin LW, Alexander F; Esophageal atresia. Surg Clin North Am., 1985; 65(5):1099–1013.
7. Foglia RP; Esophageal disease in the pediatric age group. Chest Surg Clin North Am., 1994; 4(4): 785–709.
8. Black RJ; Congenital tracheo-oesophageal fistula: in the adult. Thorax, 1982; 37(1): 61-63.
9. Genty E, Attal P, Nicolas R, Roger G, Triqlia JM, Garabeduan EN *et al*.; Congenital tracheoesophageal fistula without esophageal atresia. Int J Pediatr Otorhinolaryngol., 1999; 48(3): 231-238.
10. Zacharias J, Genc O, Goldstraw P; Congenital tracheoesophageal fistulas presenting in adults: Presentation of two cases and a synopsis of the literature. J Thorac Cardiovasc Surg., 2004; 128(2): 316-318.
11. Clark DC; Esophageal Atresia and Tracheoesophageal Fistula. Am Fam Physician. 1999; 59(4): 910-916.