

Congenital tracheoesophageal fistula: A rare and late presentation in adult patient

Waseem M. Hajjar, Ahmed Iftikhar, Sami A. Al Nassar, Salah M. Rahal

Thoracic Surgery
Department, Thoracic
Surgery Unit, King
Khalid University
Hospital, College of
Medicine, King Saud
University, Riyadh,
Saudi Arabia

Address for correspondence:

Dr. Waseem M. Hajjar,
Thoracic Surgery
Department, Thoracic
Surgery Unit, King Khalid
University Hospital,
College of Medicine, King
Saud University, Riyadh,
Saudi Arabia.
E-mail: washajjar@yahoo.
com

Submission: 12-06-11
Accepted: 01-07-11

Abstract:

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.

Key words:

Adult, congenital tracheoesophageal fistula, late presentation

Most patients with congenital tracheoesophageal fistulas (TEFs) are diagnosed immediately following birth or during infancy, because more than 98% of them are associated with atresia of the esophagus that results in potential life-threatening complications.^[1] So, early surgical intervention is needed to correct these anomalies. H-type of TEFs which present in adults are very rare, with only 16 cases described in the English literature.^[2] They are not associated with esophageal atresia and produce minimal symptoms; so, diagnosis is usually late. Here, we report a case of H-type TEF in a young adult.

Case Report

A 31-year-old male, non-smoker, presented to us with a history of attacks of chronic intermittent cough since childhood. These attacks usually last for two to three days. Sometimes initiated and aggravated after heavy meals. It was also associated with recurrent chest infections. No significant history of hemoptysis, shortness of breath, difficulty of swallowing, or choking was reported. Systemic examination was unremarkable. He used inhalers and cough suppressants as required.

Chest X-ray showed bilateral apical pleural thickening with fibroatelectatic changes at the right upper lobe. Barium studies showed significant dilatation of the whole esophagus, more at the proximal 3rd, with free passage of the contrast into the stomach without any stricture or narrowing at the distal end or at gastroesophageal junction; these findings were suggestive of some neurological causes or esophageal motility disorder. For further

evaluation, the patient underwent esophageal manometry study and upper gastro-intestinal (GI) endoscopy. Esophageal manometry showed aperistaltic esophagus, but the lower esophageal sphincter could not be assessed. Upper GI endoscopy revealed the presence of smooth tracheoesophageal opening (fistula) in the upper 3rd of the anterior wall of the esophagus at 25 cm from the incisor teeth [Figure 1]; however, biopsy and wash aspiration from this fistula was negative for tuberculosis (TB) culture.

Computed tomography (CT) scan of chest confirmed the presence of the TEF at the level of sternoclavicular joint along with dilated esophagus. Also, there was evidence of cystic, cylindrical, and varicose bronchiectatic changes in both the right upper, middle lobes, and in the lingula. There were also subpleural fibroatelectatic changes in the posterior segment of the right upper lobe, most likely due to recurrent aspirations [Figure 2a-b].

Patient explored in the operating room through cervical approach along the anterior border of the left sternocleidomastoid muscle. Fistula was identified between the posterolateral wall of the trachea and the anterior wall of the esophagus. Adhesions separated and the fistula isolated completely and then closed by endo-GI stapler. Esophageal side reinforced with muscular patch to buttress this repair and to minimize the postoperative esophageal leak and to prevent long-term recurrence. Postoperative recovery was uneventful. Patient started oral fluids on the second postoperative day, the drain removed on the third day, and he was discharged home on fourth postoperative day. Patient remained

Access this article online

Quick Response Code:



Website:
www.thoracicmedicine.org

DOI:
10.4103/1817-1737.91553

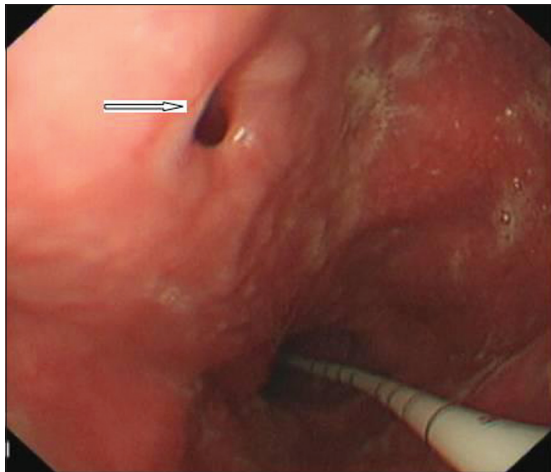


Figure 1: Tracheoesophageal fistula (TEF) in the the anterior wall of the esophagus, "see arrow"

asymptomatic and well at 12-month regular follow-up, as he underwent during this follow-up a barium swallow study which showed normal swallowing process and no hold up of the contrast with no evidence of gastroesophageal reflux and there was no evidence of recurrence of the TEF; however, he remained under regular follow-up with the gastroenterologist team for the esophageal motility disorder which has improved significantly clinically and radiologically.

Discussion

Congenital H-type TEFs represent the least severe form of abnormal tracheoesophageal communication, and its presence in adults is uncommon to the extent that no one surgeon or institution is likely to encounter more than a few cases.^[3] The presence of TEF also disrupts in utero the normal development of the myenteric plexus of the esophagus, leading to disordered peristalsis and impaired lower esophageal sphincter function.^[4] This explains the dilated esophagus along with aperistalsis seen in our case. Also, a consistent demonstration of oblique course of the fistula with tracheal orifice cephalad to esophageal orifice might function as a valve during swallowing, compressing anterior esophageal wall against the fistula and occluding its lumen with either the passage of bolus of food or peristaltic wave explains the trivial symptoms and greater tolerance of solid foods than liquids in these patients.^[5] Another plausible explanation is that initial mild symptoms are not properly investigated until complications appear.^[6]

A chronic cough aggravated by heavy meals and recurrent chest infections are common presenting symptoms. Bouts of coughing when swallowing liquids are pathognomic for this condition.^[7]

The initial radiological investigation consists of plain chest X-ray that may show esophageal gaseous distension, but a thin barium swallow, particularly if done with patient in prone position, often confirms the diagnosis. A CT scan of the chest is helpful to assess the extent of irreversible damage of the lung parenchyma and shows also the exact location of

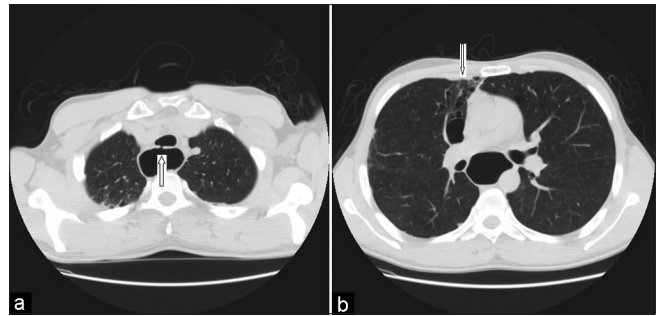


Figure 2: (a) CT scan chest demonstrating TEF at the anterior wall of the esophagus, "see arrow," (b) CT scan chest demonstrating bronchiectatic changes in the right middle lobe, "see arrow"

the fistula. Bronchoscopy allows the fistula to be visualized directly and to identify the exact location of its origin with respect to the carina or the vocal cords, as this relation to these vital structures is to be accurately established. This determines whether or not a cervical approach of the fistula is appropriate for surgical intervention. Esophagoscopy also helps for direct visualization of TEF fistulas, but occasionally they can be missed on esophagoscopy as they are located in the upper 3rd and on the anterior wall.^[8]

Other differential diagnosis of these fistulas should be excluded like the acquired one due to chronic inflammatory process or even tuberculosis.

The treatment is surgical repair and closure or stapling of the TEF. 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.^[8,2]

The early diagnosis and the treatment are essential to prevent 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. Both radiological and endoscopic procedures are complementary in diagnosis of H-type congenital TEF with high index of suspicion.

References

1. Black RJ. Congenital tracheo-oesophageal fistula in the adult. *Thorax* 1982;37:61-3.
2. 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;128:316-8.
3. Bedard P, Girvan DP, Shandling B. Congenital H-type tracheoesophageal fistula. *J Pediatr Surg* 1974;9:663-8.
4. Kovesi T, Rubin S. Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. *Chest* 2004;126:915-25.
5. Acosta JL, Battersby JS. Congenital tracheoesophageal fistula in the adult. *Ann Thorac Surg* 1974;17:51-6.
6. Negus VE. Specimen: Oesophagus from a middle-aged man

showing a congenital opening into the trachea. Proc R Soc Med 1929;22:527.

7. Azoulay D, Regnard JF, Magdeleinat P, Diamond T, Rojas-Miranda A, Levasseur P. Congenital respiratory-esophageal fistula in the adult. J Thorac Cardiovasc Surg 1992;104:381-4.
8. Genty E, Attal P, Nicolas R, Roger G, Triqlia JM, Garabedian EN, *et al.* Congenital tracheoesophageal fistula without esophageal

atresia. Int J Pediatr Otorhinolaryngol 1999;48:231-8.

How to cite this article: Hajjar WM, Iftikhar A, Al Nassar SA, Rahal SM. Congenital tracheoesophageal fistula: A rare and late presentation in adult patient. Ann Thorac Med 2012;7:48-50.

Source of Support: Nil, **Conflict of Interest:** None declared.

Announcement

iPhone App



Download
**iPhone, iPad
application**

FREE

A free application to browse and search the journal's content is now available for iPhone/iPad. The application provides "Table of Contents" of the latest issues, which are stored on the device for future offline browsing. Internet connection is required to access the back issues and search facility. The application is Compatible with iPhone, iPod touch, and iPad and Requires iOS 3.1 or later. The application can be downloaded from <http://itunes.apple.com/us/app/medknow-journals/id458064375?ls=1&mt=8>. For suggestions and comments do write back to us.