Adult congenital tracheo-esophageal fistula with esophageal dysmotility and bronchiectasis

Manar T. El-Essawy, MD, FRCR.

ABSTRACT

تظهر النواسير المريئية القصبية في سن البلوغ نتيجةً لورم سرطاني، ولكنها نادراً ما تكون خلقية. نستعرض في هذا المقال حالة شاب يبلغ من العمر 26 عاماً ويعاني من السعال المزمن، وقد أظهرت الأشعة السينية حدوث توسع في المريء مع توسع في الفص العلوي من القصبة الهوائية. ولم يكن هناك دليل على وجود ورم خبيث، أو تعذر الارتخاء، أو مرض شاغاس، بالإضافة إلى عدم وجود تاريخ سابق بكل من: الإصابات الرضحية، أو الالتهابات، أو التدخل الجراحي. أثبتت الأشعة المقطعية وتنظير المريء وجود الناسور المريئي القصبي وذلك بخلاف ما ظهر في الأشعة السينية. ويؤكد هذا البحث بأن الناسور المريئي القصبي المجهول السبب أو الخلقي قد يظهر في سن البلوغ مع تغير في حركة المريء وتوسع في القصبة الهوائية.

Tracheo-esophageal fistulas (TEF) in adults are very rarely congenital in nature and most of the cases are secondary to neoplastic cause. We report a 26-year-old male with TEF that presented with chronic cough. An x-ray carried out for the patient showed upper lobe bronchiectasis and massive esophageal dilatation. There was no evidence of esophageal malignancy, achalasia, or Chagas' disease. There was no history of trauma, infection, or intervention. The CT demonstrated a small TEF, which was not visualized on esophagogram and confirmed by bronchoscopy. Our case demonstrated that idiopathic, or congenital TEF can be presented in adulthood with esophageal dysmotility and bronchiectasis.

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From the Department of Radiology and Diagnostic Imaging, King Saud University, King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Manar T. El-Essawy, Department of Radiology and Diagnostic Imaging, King Khalid University Hospital, PO Box 361680, Riyadh 11313, Kingdom of Saudi Arabia. Tel. +966 (1) 2251295. E-mail: manar970@hotmail.com

Tracheo-esophageal fistulas (TEF) is a condition, in which there is a communication between the alimentary and respiratory tracts. In adults, the presentation is predominantly associated with esophageal cancer.[1] Benign TEF in adults is rare.[2] Congenital TEF without esophageal atresia, otherwise known as the H-type, is exceptionally rare in adults. These patients often have an inborn esophageal motility disorder that is preserved even after the repair of the fistula. Non specific symptoms lead to a delay in diagnosis.[3] This paper highlights the need for thorough evaluation for occult esophageal respiratory fistula in any patient with chronic pulmonary symptoms of unknown etiology. Timely diagnosis and treatment will prevent serious complications and years of debilitating illness.

Case Report. A 26-year-old male patient presented to the Department of Radiology, King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia, 8 months ago because he experienced chronic cough, and he gave the history of similar recurrent complaints since childhood, which was treated with different types of medication without complete improvement. He also experienced bouts of dysphagia. He has no associated fever, history of injury, or exposure to trauma. The patient was not treated with any anti-tuberculous drugs in the past. He was not a smoker or an alcoholic. The chest x-ray was carried out for the patient which showed large gas filled tubular structure (**Figure 1**) in the mediastinum extending down to the upper abdomen that was seen continuous with the gastric shadow in keeping with the dilated esophagus, and there were also bronchiectatic changes in the upper zone of the lungs bilaterally more on the right side. Barium study (swallow) was performed that demonstrated marked dilatation of the esophagus, particularly in its upper part with almost normal caliber of its distal end at gastro-esophageal junction with no areas of narrowing, or any signs to suggest the diagnosis of achalasia. Esophageal manometry was performed that showed no abnormality in the function of the muscles of the lower esophagus, and no pressure abnormality along the lower esophageal sphincter. The fistula is

not visualized in the barium study. The chest CT was carried out showing a focal area of soft tissue disruption between trachea and proximal esophagus representing TEF formation (**Figure 2a**) opposite the sternoclavicular joint along with marked dilatation of the esophagus and bronchiectatic changes (**Figure 2b**) in the upper lobes bilaterally.

Discussion. Failure of separation of the esophagus and trachea in the early stage of embryonic development results in congenital fistula between the bronchus/trachea and the esophagus, and is usually associated with esophageal atresia, and most of the cases usually presents in infancy.[4] In adults, a fistula between the esophagus and the respiratory tract is usually the result of a malignant disease.[1]

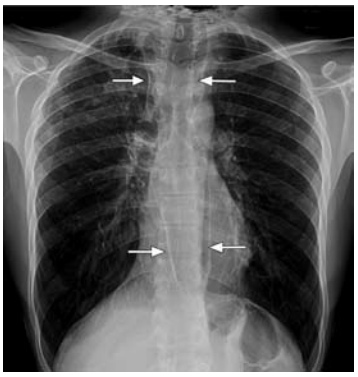

Figure 1 - Plain x-ray of the chest posterior to anterior view showed diffuse dilatation of the esophagus that appears as a tubular air filled structure seen in the mediastinum (arrows) that is seen extending downward along the diaphragm connected with the gas shadow of the stomach.

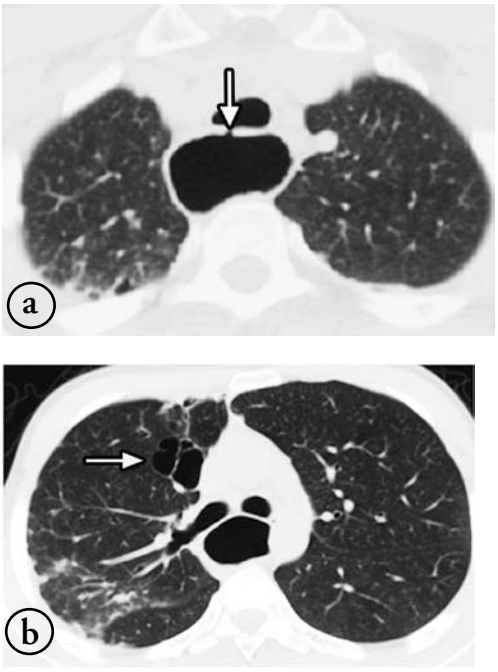

Figure 2 - An axial CT scan of the chest lung window demonstrates: **a**) soft tissue defect between trachea and esophagus (white arrow) with marked dilatation of the esophagus with **b**) foci of bronchiectasis (arrow) in the right lung with area opacification resulting from chronic recurrent aspiration and infection.

Non-malignant TFE fistulas are relatively rare, and may be congenital, or acquired as a complication of inflammatory disorders, foreign body ingestion, trauma,[5] and prolonged intubation of the trachea.[6] Infrequent etiologies include Barrett's ulceration of the esophagus, ulceration of heterotopic gastric mucosa, and an iatrogenic complication of endoscopic variceal sclerotherapy.[7] Adult congenital TEF without accompanying esophageal atresia is a rare anomaly, however, due to the lack of esophageal atresia, they are compatible with life and may present in adult life, the highest incidence occurring in the third decade with no gender preponderance. This anomaly may cause various symptoms, such as chronic respiratory infections, cough, and coughing bouts when eating or drinking, and even hemoptysis.[8] The tracheal opening is usually above the esophageal opening, giving the fistula the appearance of a slanted "H".[8] The H type of TEF usually present in adults could be due to the delay in the rupture of an occlusive membrane that covers the tract until adulthood, or due to the obliquity of the tract with the esophageal end lower than the tracheal end that closes during swallowing, or might be due to the presence of the mucosal fold at the esophageal end that acts as a check valve and prevent aspiration. The contraction, closure, and the obliquity of the tract can all be reasons for the late appearance of the symptoms until later in adulthood[2] as found in the present patient.

In patients with TEF, respiratory symptoms are more prevalent than the gastrointestinal symptoms. Most have cough on swallowing, due to spillage of gastric contents into the respiratory tract that is predispose to recurrent respiratory tract infections, and could result in bronchiectasis and lung abscesses.[9] The adult congenital TEF more commonly occurs between the right lower lobe bronchus and the middle third of the esophagus.[10] Other places include left lower lobe bronchus, bronchus intermedius, and right upper lobe bronchus. The present case was peculiar because a fistula existed between the middle part of the trachea and the esophagus. Another intriguing feature of our case was the presence of bronchiectasis in the upper, rather than the lower and middle lobes. We hypothesize that the bronchiectasis is a very unusual complication,[11] has a predilection in the posterior segment of the right upper lobe, which could have been due to the aspiration of food contents, while the patient was lying supine and on his side.[10]

Patients with congenital TEF have esophageal dysmotility with inefficient or absent peristalsis along

with reduced lower esophageal sphincter pressure, resulting in reflux and esophagitis.[12] These changes lead to diffuse dilatation of the esophagus as found in the presented case. Achalasia was not present in our patient, which was proven by distal esophageal manometry, however, abnormal peristalsis, dysmotility, and dilatation of the esophagus were present. The most effective tool in the diagnosis of congenital TEF is rigid endoscopy, but sometimes it can miss the lesions that have a flap membrane as it covers and conceals the fistula.[8] At times, even an esophagram may not be helpful because of the lower positioning of the esophageal opening relative to the tracheal opening. This angulation precludes contrast flow from the esophagus into the trachea. Nonetheless, esophagram is still beneficial in the diagnosis of dilatation, achalasia, and dysmotility associated with the fistula. Bronchoscopy, can be advantageous at times as in the case of our patient where the fistula was not seen on esophagoscopy. A CT with thin cuts is seldom helpful, unless the fistula is so conspicuous, as in our case, that it can be readily visualized. In rare cases, TEF cannot be diagnosed by any diagnostic modality, and is only established at the time of surgery, which is the most definitive therapeutic option.

A high degree of suspicion is required to reach a proper diagnosis. Although there is no severe progressive clinical deterioration due to a fistula of limited size, closure is important to avoid chronic pneumonia and disability.[8]

In conclusion, a diagnosis of tracheo-esophageal fistula should be suspected in any patient who presents with persistent unexplained cough. Such a diagnosis should be questioned, and further assessment should be tailored when symptoms are recurrent or persistent, inspite of medical treatment. The rarity of this entity, as well as the relatively nonspecific symptoms and signs makes the diagnosis challenging.

References

1. Dogan R, Farsak B, Yilmaz M, Tok M, Güngen Y. Congenital broncho-oesophageal fistula associated with bronchiectasis in adults. Report of two cases and review of the literature. ***Respiration*** 1999; 66: 361-365.
2. Kohler B, Köhler G, Riemann JF. Spontaneous esophagotracheal fistula resulting from ulcer in heterotopic gastric mucosa. ***Gastroenterology*** 1988; 95: 828-830.
3. Mangi AA, Gaissert HA, Wright CD, Allan JS, Wain JC, Grillo HC, et al. Benign broncho-esophageal fistula in the adult. ***Ann Thorac Surg*** 2002; 73: 911-915.
4. Smith BD Jr, Mikaelian DO, Cohn HE. Congenital bronchoesophageal fistula in the adult. ***Ann Otol Rhinol Laryngol*** 1987; 96: 65-67.
5. Ranganathan SC, Ramesh PM. Congenital tracheo-oesophageal fistula in adult. ***Pulmon*** 2006; 8: 62-64.
6. Gerzic Z, Rakic S, Randjelovic T. Acquired benign esophagorespiratory fistula. Report of consecutive cases. ***Ann Thorac Surg*** 1990; 50: 724-727.
7. Diehl JT, Thomas L, Bloom MB, Dresdale AR, Harasimowicz P, Daly BD, et al. Tracheoesophageal fistula associated with Barrett's ulcer: the importance of reflux control. ***Ann Thorac Surg*** 1988; 45: 449-450.
8. Tamay Z, Güler N, Kiliç G, Gün F, Celik A, Yekeler E. A congenital proximal tracheoesophageal fistula 14 years after surgical repair of esophageal atresia with distal tracheoesophageal fistula. ***Turk J Pediatr*** 2008; 50: 299-301.
9. Bhandari A. Congenital malformations of the lung and the airway. In: Bell LM, Panitch HB, eds. Pediatric Pulmonology. The Requisites in Pediatrics. Philadelphia (PA): Elsevier Mosby; 2005. p. 40-43.
10. Kurashige M, Kusachi S, Kato O, Kamiya K, Ito K, Atobe T. Congenital bronchoesophageal fistula in a 39-year-old male. ***Nippon Kyobu Geka Gakkai Zasshi*** 1985; 33: 922-927.
11. Little DC, Rescorla FJ, Grosfeld KW, Scherer LR, Engum SA. Long-term analysis of children with esophageal atresia and tracheoesophageal fistula. ***J Pediatr Surg*** 2003; 38: 852-856.
12. Behnia M, Tarver RD. Unilateral bronchiectasis and esophageal dysmotility in congenital adult tracheoesophageal fistula. ***Internal Medicine*** 2001; 40: 620-623.

Ethical Consent

All manuscripts reporting the results of experimental investigations involving human subjects should include a statement confirming that informed consent was obtained from each subject or subject's guardian, after receiving approval of the experimental protocol by a local human ethics committee, or institutional review board. When reporting experiments on animals, authors should indicate whether the institutional and national guide for the care and use of laboratory animals was followed.