A congenital proximal tracheoesophageal fistula 14 years after surgical repair of esophageal atresia with distal tracheoesophageal fistula

Zeynep Tamay[1], Nermin Güler[1] Gürkan Kılıç[1], Feryal Gün[2]
Aleaddin Çelik[2], Ensar Yekeler[3]
[1]Division of Allergy and Chest Diseases, Department of Pediatrics, and Departments of [2]Pediatric Surgery and [3]Radiology, İstanbul University, Istanbul Faculty of Medicine, İstanbul, Turkey

SUMMARY: Tamay Z, Güler N, Kılıç G, Gün F, Çelik 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.

Congenital esophageal atresia with proximal and distal tracheoesophageal fistula occurs very rarely. This report describes late diagnosis of a congenital proximal fistula in a 14-year-old girl who underwent surgical repair of a congenital esophageal atresia with distal tracheoesophageal fistula on her first day of life and suffered recurrent bouts of lower airway infections and chronic cough, with the diagnosis of asthma in later childhood.

Key words: children, congenital, esophageal atresia, tracheoesophageal fistula.

Congenital esophageal atresia (EA) and tracheoesophageal fistula (TEF) are common congenital anomalies, occurring in 1/2500 to 1/4500 births[1,2]. TEF results from the failure of the digestive and respiratory tracts to separate from each other during the embryonic stage of development[3]. Esophageal atresia with two fistulas (both proximal and distal TEF) occurs very rarely, in less than 0.3%[4].

This report describes late diagnosis of a congenital proximal fistula in a 14-year-old girl who underwent surgical repair of a congenital EA with distal TEF on her first day of life and who suffered recurrent bouts of lower airway infections and chronic cough, with the diagnosis of asthma in later childhood.

Case Report

A 14-year-old girl was referred to the Department of Pediatric Chest Diseases and Allergy with complaints of chronic cough and intermittent dyspnea.

Her history revealed that she had a primary repair of a congenital EA with TEF on the first day of her life. The postoperative course was uneventful; she was discharged on the 10th postoperative day, while feeding by mouth regularly. During her regular outpatient follow-up visits until five years of age, she had one choking event (on a piece of orange) at 16 months of age, which was removed by rigid esophagoscopy, and one dilatation procedure at the same time because of slight narrowing at the area of anastomosis demonstrated by barium swallow examination of the esophagus. Another barium swallow and 24-hour esophageal pH monitoring examination performed at three and five years of age revealed normal findings.

After an asymptomatic interval she had recurrent bouts of pneumonia and wheezy episodes requiring two hospitalizations when she was seven years old and three hospitalizations in the previous year. A chest computed tomography (CT) performed at 10 years of age revealed right-sided bronchiectasis. She had been using inhaled corticosteroids and bronchodilators intermittently since the age of seven with the diagnosis of asthma.

On physical examination, her weight was between the 75th and 90th percentile and height was at the 97th percentile. On auscultation, she had bilateral crackles and rhonchi at lower zones. Laboratory investigations including

whole blood count, serum biochemistry and acute phase reactants were within the normal limits. Chest high resolution CT demonstrated bilateral fields of segmentary cylindrical bronchiectasis and bilateral bronchial infiltrations at basal zones. Pulmonary function test showed a slight restrictive pattern [forced expiratory volume in 1 sec (FEV1) (%predicted): 67; FEV1/forced vital capacity (FVC) (%): 107; peak expiratory flow (PEF) (%predicted): 69; and no change in FEV1 15 minutes after administration of inhaled beta-2 agonist]. Peripheric blood lymphocyte subsets, serum quantitative immunoglobulin and subgroup levels and sweat test were normal. Barium swallow investigation and 24-hour pH monitoring for gastroesophageal reflux revealed normal findings. A dynamic radionuclide gastroesophageal scintigraphy with technetium-99m showed a fistulous opening between the trachea and the esophagus and radionuclide activity at right lung. Although the standard barium swallow was found normal, repeated barium swallow in prone position confirmed a proximal TEF at midline, in the posterior trachea (Fig. 1). During surgical procedure, the fistula determined by a guidewire with bronchoscopy was found at the supraclavicular level and repaired.

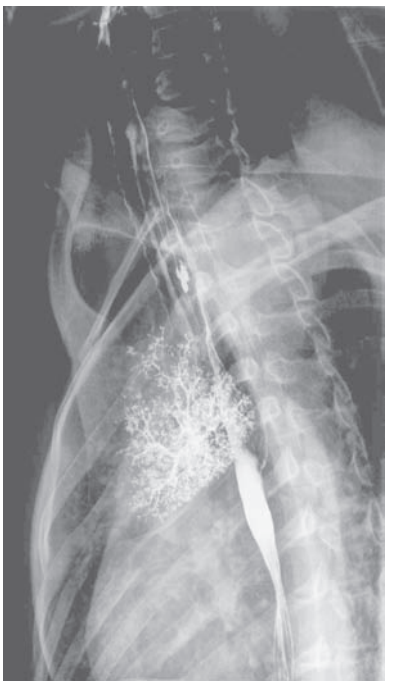

Fig. 1. Barium swallow examination in the prone oblique position shows aspiration to the right lung due to tracheoesophageal fistula.

Discussion

Most infants with EA with TEF have proximal atresia with distal TEF. They are easily diagnosed in the first days of life with apparent symptoms and findings and undergo surgical repair. Since two fistulas with EA occur very infrequently, proximal fistula may be overlooked during primary surgical repair, as in the present case. An undiagnosed proximal fistula shares the same clinical features with an isolated H-type fistula, which is usually diagnosed later, even in adulthood[4-7].

Although symptoms of an undiagnosed proximal fistula may include choking, coughing and recurrent respiratory tract infections, there may be long and asymptomatic intervals, which may be explained by the nature of the fistula[6]. The contractions of the muscle wall of the fistula or the oblique direction of the fistula tract from the trachea to esophagus may protect the airway from aspiration of foods during swallowing[6-9]. In the present case, up until seven years of age, the child had no apparent symptoms requiring admission to hospital except a choking period due to esophageal stricture, which is an expected postoperative complication with an occurrence rate of 6 to 40% in repaired patients[10].

Recurrent respiratory tract infections and wheezing are commonly reported in infants after primary repair, but become less frequent over time[2,7]. Bronchiectasis is a very unusual complication[7,11,12]. Asthma is reported in nearly one-fourth of children with repaired EA and TEF[13,14]. In our patient, late onset of recurrent bouts of pneumonia and wheezing resulting in bronchiectasis, unresponsiveness to anti-asthmatic treatment, and pulmonary function tests incompatible with the diagnosis of asthma raised the possibility of a recurrent TEF or a second congenital fistula.

Recurrence of the TEF occurs in approximately 8-9% of cases, most often in the immediate postoperative period or early in life[2,12]. Late recurrence or re-communication following the primary repair is very rare[15]. In our patient, recurrent TEF was not considered. The primary fistula, repaired in the newborn period, was at the carinal level. A second fistula, which was preoperatively demonstrated by radionuclide scanning, was determined at the supraclavicular level by a guidewire with

bronchoscopy during the surgical procedure. In the pre-operative period, although standard barium swallow failed to show the fistula in the first attempt, the second procedure performed in the prone position confirmed the diagnosis. The presence of a TEF can also be demonstrated by three-dimensional CT scanning and virtual bronchoscopy[16].

This case report emphasizes the importance of performing bronchoscopy prior to initial TEF repair. Preoperative esophagoscopy and bronchoscopy may be performed in every EA to determine the site of distal TEF, to exclude an upper pouch fistula and to document the presence of tracheomalacia. It also emphasizes that recurrent respiratory symptoms in a patient with repaired EA with TEF should alert the physician to the possibility of a second or recurrent TEF. In such cases, a detailed surveillance of the upper gastrointestinal system is recommended to exclude TEF before attributing the cause to asthma.

REFERENCES

1. Harmon CM, Coran AG. Congenital anomalies of the esophagus. In: O'Neil JA Jr, Rowe MI, Grosfeld JL (eds). Pediatric Surgery. St Louis: Mosby; 1998: 941-967.
2. Spitz L. Esophageal atresia: lessons I have learned in a 40-year experience. J Pediatr Surg 2006; 41: 1635-1640.
3. Bhandari A. Congenital malformations of the lung and the airway. In: Bell LM, Panitch HB (eds). Pediatric Pulmonology, The Requisites in Pediatrics. Philadelphia: Elsevier Mosby; 2005: 40-43.
4. Peat SJ, Paes T, Howard ER, Jones PI. Congenital tracheo-oesophageal fistula in an adult. Br J Surg 1989; 76: 412.
5. Azoulay D, Regnard JF, Magdeleinat P, Diamond T, Rojas-Mirand A, Levasseur P. Congenital respiratory-esophageal fistula in the adult. Report of nine cases and review of the literature. J Thorac Cardiovasc Surg 1992; 104: 381-384.
6. Enoksen A, Lovaas J, Haavik PE. Congenital tracheo-oesophageal fistula in the adult. Report of a case and review of the literature. Scand J Thorac Cardiovasc Surg 1979; 13: 173-176.
7. Karnak I, Senocak M, Hicsonmez A, Buyukpamukcu N. The diagnosis and treatment of H-type tracheoesophageal fistula. J Pediatr Surg 1997; 32: 1670-1674.
8. Chernoff WG, White AK, Ballagh RH. Tracheoesophageal fistula: a case report. Int J Pediatr Otorhinolaryngol 1993; 27: 173-182.
9. Risher WH, Arensman RM, Ochsner JL. Congenital bronchoesophageal fistula. Ann Thorac Surg 1990; 49: 500-505.
10. Engum SA, Grosfeld JL, West KW, Rescorla FJ, Scherer LR 3rd. Analysis of morbidity and mortality in 227 cases of esophageal atresia and/or tracheoesophageal fistula over two decades. Arch Surg 1995; 103: 502-508.
11. Chetcuti P, Myers NA, Phelan PD, Beasley SW. Adults who survived repair of congenital esophageal atresia and tracheo-oesophageal fistula. BMJ 1988; 297: 344-346.
12. 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.
13. Agrawal L, Beardsmore CS, MacFayden UM. Respiratory function in childhood following repair of oesophageal atresia and tracheo-oesophageal fistula. Arch Dis Child 1999; 81: 404-408.
14. Couriel JM, Hibbert M, Olinsky A, Phelan PD. Long-term pulmonary consequences of oesophageal atresia with tracheo-oesophageal fistula. Acta Paediatr Scand 1982; 71: 973-978.
15. Alvear DT, Cresson SL, Kirkpatrick JA. Recurrence of tracheoesophageal fistula. Am J Dis Child 1973; 126: 660-661.
16. Le SDV, Lam WW, Tam PK, Cheng W, Chan FL. H-type tracheo-oesophageal fistula: appearance on three-dimensional computed tomography and virtual bronchoscopy. Pediatr Surg Int 2001; 17: 642-643.