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Olgu Sunumu

Management of H-type tracheoesophageal fistula in children: A report of 3 cases

Çocuklarda H tipi trakeoözofageal fistülün yönetimi: Üç olgu sunumu

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ABSTRACT

H-type tracheoesophageal fistula (TEF) is a relatively uncommon congenital anomaly that can be difficult to identify and sometimes, challenging to repair. It is the Gross E type of esphageal atresia (EA) and constitutes 4% of all EA cases. Three infants with TEF were treated between 2003 and 2012. The diagnostic workup, surgical technique, and postoperative course of patients who underwent repair of H-TEF were reviewed. Conventional esophagram demonstrated the fistula in 2 of the patients and a cineradiographic procedure was performed to outline the H-TEF in the last patient. In all 3 cases the location of the fistula was confirmed by tracheoscopy. The closure of the fistula was made by cervical route in 2 cases and by thoracotomy in the remainig patient with distal located fistula. A high index of suspicion for an H-TEF should be maintained in the presence of neonatal respiratory symptoms. Since H-TEFs are known to be complicated with lower respiratory tract infection, early referral of these patients to pediatric surgeons and accurate and timely surgical treatment should be realized.

Key words: Tracheoesophageal fistula, H-type, tracheoscopy

ÖZET

H tipi trakeoözofageal fistül (TÖF) ender bir anomali olup, tanı ve onarımda bazen zorluklar içermektedir. Bu klinik durum özefageal atrezilerin (ÖA) Gross sınıflamasına göre E-tipi olup tüm ÖA olgularının %4'nü oluşturmaktadır. Kliniğimizde 2003 ile 2012 yılları arasında H-tipi TÖF'lü üç olgu tedavi edilmiştir. Olgularımızda tanı yöntemleri, cerrahi teknik ve ameliyat sonrası gidişat gözden geçirilmiştir. Tanıda konvansiyonel özofagografi iki olguda yararlı olurken, son olguda ise sineözofagografi yararlı olmuştur. Tüm olgularda trakeoskopi ile fistülün yeri doğrulanmıştır. İki olguda servikal yaklaşım yolu ile, distal yerleşimli fistül olan son olguda ise torakotomi yolu ile fistül onarımı uygulanmıştır. Neonatal respiratuvar bulguların varlığında H tipi TÖF akılda tutulmalıdır. Alt solunum yolu infeksiyonları ile komplike olduklarından bu olgular çocuk cerrahına erkenden refere edilmeli ve uygun ve zamanlı cerrahi onarım uygulanmalıdır.

Anahtar kelimeler: Trakeoözofageal fistül, H-tipi, trakeoskopi

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INTRODUCTION

Despite the elapse of more than a century since the original description of congenital H-type tracheoesophageal fistula (H-TEF) by Lamb, difficulties and pitfalls in establishing the definitive diagnosis continue to exist [1]. Although symptoms are usually present from birth, the diagnosis is frequently delayed because of nonspecific and sometimes intermittent symptoms, and because of false negative results of diagnostic tools.

H-type tracheoesophageal fistula is the Gross E-type of esophageal atresia (EA) and constitutes 4% of all EA cases. Classically it is characterized by a clini-

cal triad consisting of history of choking and cyanosis after feeding during infancy, gaseous distention of the gastrointestinal tract and recurrent lower respiratory tract infections [2,3].

In the current study, our aim is to review the diagnostic workup, associated disorders, surgical technique, and postoperative course of patients who underwent surgical treatment of H-TEF in our department.

MATERIALS and METHODS

A retrospective study was conducted with children having H-TEF treated in our hospital between month 2003 and month 2012. Age, sex, duration of symptoms, clinical features, preoperative diagnostic tests, treatment, and postoperative complications were noted.

RESULTS

During the study period one female, and two male infants were analyzed. Mean ages at presentation and surgical treatment were 63.0 days (14-150 days) and 131.7 days (35-270 days), respectively. Total number of 117 EA patients were seen during the study period. Average duration of follow-up in patients with H-TEF was 39 months (2-62 months). There is Any case of mortality, morbidity or recurrence of the fistulas was not detected.

Case 1: A term male baby weighing 3.7 kg was born through normal vaginal delivery. He became distressed after every feeding with choking, cyanosis and episodes of abdominal distention. He was admitted with the diagnosis of bronchopneumonia at the age of 5 days. Associated anomalies included inguinal hernia and hypertrophic lingual frenum. Subsequent esophagram with the 8F orogastric tube in place showed an H-TEF in the upper esophagus (T2-T3 level) with an intact esophagus from the oropharynx to the stomach (Figure 1). Intervening Candida septicemia which was treated medically detained definitive surgical treatment to the age of 3 months. During operation, rigid bronchoscopy was performed, and the fistula was identified in the posterior trachea, approximately 2 cm below the epiglottis. Then a 4 F ureteral catheter was placed through the fistula. He was operated through a right cervical approach. By carefully palpating the catheter TEF was identified and oversewn on both the tracheal and esophageal ends with interrupted polydioxanone sutures and an omohyoid muscle flap was interposed in between to prevent recurrent fistula formation. Synchronous repair of inguinal hernia and hypertrophic lingual frenum was also performed. An esophagram performed on the 7th postoperative day revealed no evidence of a leak from either repair. The infant was switched to full oral feedings and postoperative recovery was uneventful. During a follow-up period of 62 months, any morbidity or recurrence of TEF was not detected.

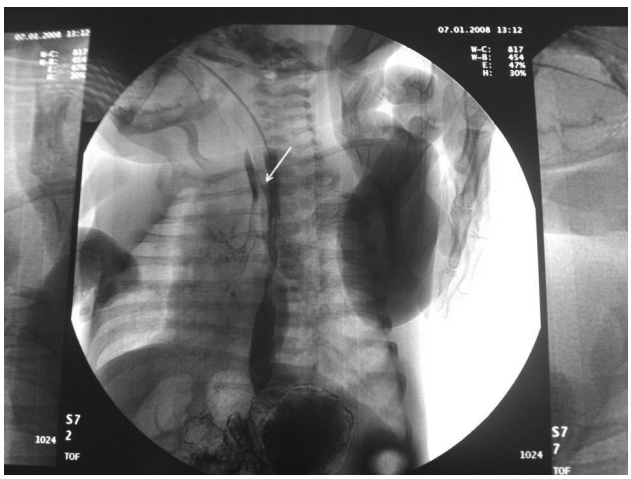

Figure 1: Conventional esophagram revealing an OG tube, the presence of an H-type fistula between the esophagus and trachea and contrast lining the bronchial tree (Arrow: H-type fistula).

Case 2: A term male weighing 3.45 kg was born with caesarean section. At the age of 25 days he was admitted with a history of respiratory distress, persistent coughing and choking with every feed dating back to birth. Antenatal history, physical examination, and chest radiograph were unremarkable. An esophagram showed an H-TEF at the level of second dorsal vertebrae, and an intact esophagus all the way down to the stomach. On day 35, before surgery, with a rigid bronchoscope tracheoscopic examination was performed which revealed an H-TEF located in the

posterior aspect of the trachea 2.5 cm. distal to the vocal cords and the fistula was catheterised with a 4 F ureteral catheter. After successful identification and division of the fistula through a right cervical approach, both ends of H-TEF were repaired. A sternocleidomastoid muscle flap was fixed between the trachea and esophagus to reinforce the repair. Postoperatively he did well and was symptom free. The esophagraphy taken on the first postoperative year showed an otherwise intact passage with an esophageal diverticulum at the level of T1 (Figure 2). Mean follow-up period was 53 months without any morbidity.

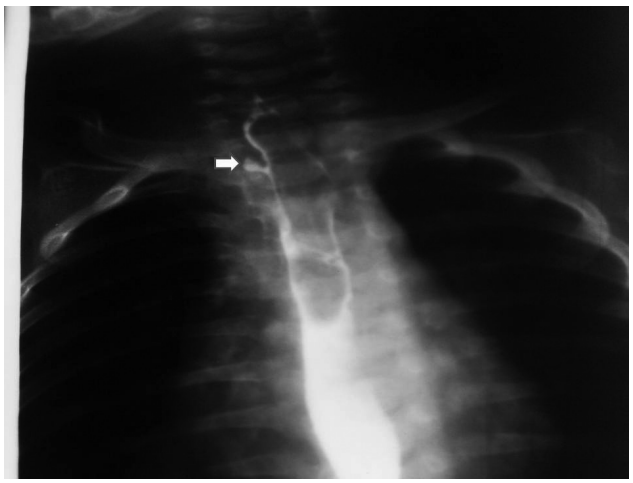

Figure 2. Control esophagram taken on the first postoperative year showing an esophageal diverticulum with an otherwise intact esophagus (Arrow: esophageal diverticulum).

Case 3: A term female was born with caesarean section weighing 2.2 kg. With symptoms of persistent wheezing, she was admitted with the diagnosis of bronchopneumonia at the age of 9 months. Her history revealed that the wheezing episodes dated back to birth and she had been repeatedly hospitalized with the diagnosis of endobronchial tuberculosis. Physical examination was unremarkable except an intense growth retardation with a weight percentile below 3 percent. Conservative esophagraphy failed to outline the fistula and H-TEF was diagnosed during a cine esophagographic examination which revealed the fistula in mid-esophagus but a seemingly intact esophagus from the oropharynx to the stomach (Figure 3). Rigid bronchoscopy was performed, showing a fistula with a diameter of 6-7 mm in the posterior trachea 4 cm proximal to the carina, and it was catheterized with a 4 F ureteral tube. Due to the location of the fistula, a right extrapleural posterolateral thoracotomy, and successful division of both ends of the H-TEF were performed (Figure 4). In order to prevent recurrent fistula formation, flap of parietal pleura was interposed between the esophagus and trachea. Postoperative recovery was uneventful. The infant remained intubated for 3 postoperative days. Esophagraphy performed on the 7th postoperative days revealed no evidence of leak and diet was

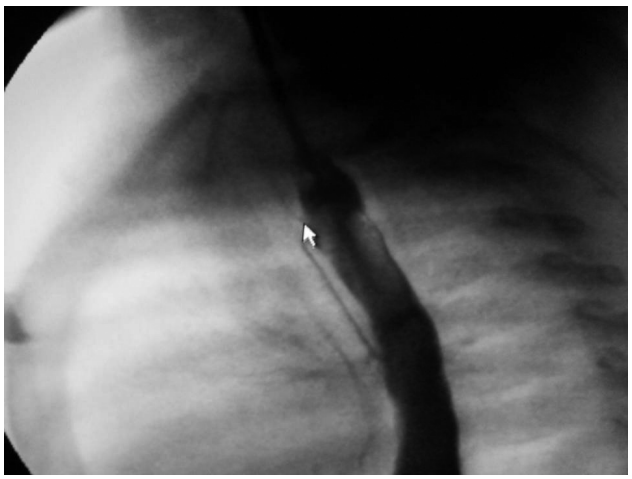

Figure 3. A cineradiographic procedure showing a clear fistula filled with contrast that courses cephalad from the esophagus into the trachea (Arrow: H-type fistula).

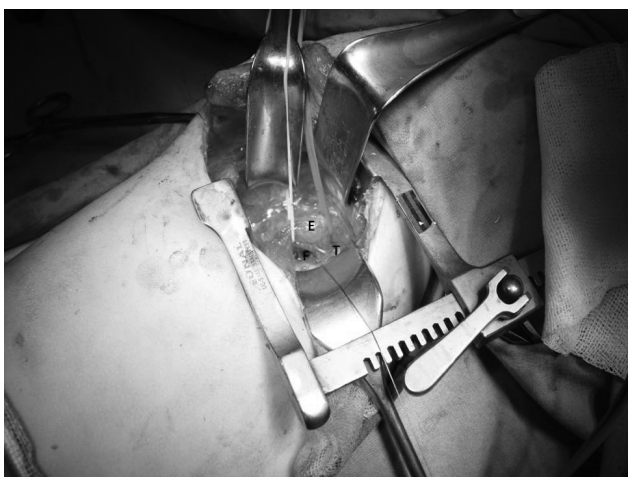

Figure 4. Intraoperative view of a child with H-TEF located 4 cm proximal to carina. Note the ureteral catheter inside the fistula serving as an intraoperative marker (E: esophagus, F: H-type fistula, T: trachea).

advanced to full oral feedings. Two months after the surgery, she was still under regular follow-up. During the follow-up period she gained weight appropriately and remained symptom free.

DISCUSSION

H-type tracheoesophageal fistula was first described in 1873 and firstly reported on a postmortem specimen in 1929 [1,4]. Imperatori performed a successful transcervical repair of the fistula in 1939 [5]. Extrapleural transthoracic approach to repair H-TEF was introduced in 1948 [6]. Subsequent reports dealing with various aspect of this anomaly have appeared in the literature [7-25].

This is a rare anomaly that occurs approximately once in 100.000 births [7]. A pediatric surgeon is unlikely to treat a lot of cases during his career due to the rarity of this clinical entity. The etiology is not known with certainty, but it is thought to result from incomplete separation of the trachea and esophagus in the early embryologic development [8,9]. The term H-TEF refers to an oblique communication between the posterior wall of the trachea and the anterior wall of the esophagus, presenting as an 'H' form.

H-type tracheoesophageal fistula represents less than 5% of individuals with an anomaly within a TEF/EA spectrum [10]. Cases with H-TEF in the current study constitute 2.6% of all the children with the anomaly of TEF/EA spectrum treated in our clinic during the study period. Relative low percentage of H-TEF patients (2.6%) in this series with regard to TEF/EA spectrum as whole, may be attributed to discrepancies pertaining to the demographic characteristics of the study population and genetic factors. Patients with H-TEF rarely present with typical manifestations in the newborn period although many infants have a clear history of coughing and choking with oral feedings or episodes of pulmonary aspiration [11]. The diagnosis is often delayed for months or years. A time lag extending up-to 63 years for confirmation of the diagnosis has been reported [12]. The oldest age of the patient at the time of surgical treatment in our study was 9 months comparable to those reported previously [13-15].

Many diagnostic methods have been used to establish the diagnosis for a suspected H-TEF. Classical esophagram is usually a reliable method to identify the anomaly though often difficult, requiring multiple attempts for confirmation of the diagnosis. Former two patients have been successfully diagnosed by way of this diagnostic modality in the current study. Due to inconclusive findings in the conventional esophagram in that the fleeting wisp of contrast might might have been missed, a cineradiographic procedure was performed to demonstrate the H-TEF in case 3 in our study (Fig. 3). Noninvasive diagnostic modalities in the diagnostic work up include magnetic resonance imaging (MRI) and scintigraphic examination. Due to potential hazards of conventional invasive investigations, MRI and radionuclide imaging using Tc-99m sulfur colloid have been also suggested for diagnosis and localization of an H-TEF especially in critically ill premature infants [16,17].

Careful assessment of trachea and esophagus by endoscopic examination has been recommended as an important adjunct to surgical repair of H-TEF. This method has the advantage of being diagnostic, in that its establishes the level of fistula, allows placement of a catheter across the fistula and identifies an additional fistula [10]. However bronchoscopic cannulation of the fistula is not always successful. An innovative technique of transillumination using a flexible pediatric bronchoscope to localize H-type fistula has been also suggested [18]. Identifying the level of fistula with respect to the carina, the vocal cords and epiglottis is crucial. On all three patients bronchoscopy was firstly performed and a fistula tract noted in the trachea was catheterized with a 4 F ureteral tube. Repair of H-TEF is typically performed through cervical route along the anterior border of sternocleidomastoid muscle. However, for the repair of fistulas below the level of T2 and T3 or when a damaged lung should be managed with simultaneous pulmonary resection, a thoracotomy operation is suggested [2,14,15,19]. A thoracoscopic approach has also

been described in the management of patients with H-TEF, but this has not become a standard practice yet [20-22]. In a study evaluating the endoscopic treatment of H-TEF using electrocautery and Nd:YAG laser, the latter was found to be better than electrocautery for the obliteration of the fistula [23]. In the current study, the fistulae were repaired successfully via a right cervical approach in two patients with TEF located in the proximal trachea whereas a formal thoracotomy was performed in case 3 with fistula 4 cm proximal to the carina.

Conventional thoracotomy may be associated with significant late sequelae including scapula alata, scoliosis, and excessive scarring [24]. There is a risk of damage to vital structures including carotid artery, internal jugular vein, injury to thyroid gland and recurrent laryngeal nerve during surgical intervention by cervical route [7]. Except a minute esophageal diverticulum observed in case 2, none of our patients had long-term difficulty with swallowing, respiration, and phonation.

H-type TEF is associated with other malformations in about 30% of the cases, including VACTERL/VATER, CHARGE syndrome, Goldenhar's syndrome, esophageal stenosis, and syndactyly [25]. None of the children with H-TEF in our study exhibited malformations described above and only case 1 had inguinal hernia and hypertrophic lingual frenum which were synchronously repaired at the time of definitive surgical treatment of the fistula.

Nevertheless, this report has some limitations. The retrospective design, limited number of patients and relatively short follow-up period may undermine the strength of this study. Rarity of this anomaly may necessitate conduction of multicentered studies on this subject. Further prospective studies involving more patients with longer follow-up periods may provide additional information on the management of these lesions.

In conclusion, a high index of suspicion for an H-TEF should be maintained in the presence of neonatal respiratory symptoms. Delay in diagnosis may be due to minute symptoms in some patients, low index of suspicion by the physicians, and unsatisfactory radiological methods. Repeat esophagrams and bronchoscopies may be required for diagnosis. Early referral of these patients to pediatric surgeons and accurate and timely surgical treatment is suggested. A long-term follow-up is necessary because of the risk of recurrence.

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