An unusual case of esophageal and laryngotracheal atresia

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Esophageal atresia with or without tracheoesophageal fistula is a relatively common congenital anomaly. However, esophageal atresia with associated laryngotracheal atresia, double tracheoesophageal fistula and cardiac malformations is an extremely rare condition. In this article we report a newborn infant with severe respiratory distress at birth who had both esophageal and laryngotracheal atresia with congenital cardiac malformations, in an attempt to bring attention to the clinical presentation, and emergent diagnostic and therapeutic approaches.

Key words: esophageal atresia, laryngeal atresia, tracheal atresia, congenital cardiac malformations.

Esophageal atresia (EA) with or without associated tracheoesophageal fistula (TEF) is a relatively common congenital anomaly. However, EA in combination with laryngotracheal atresia and double TEF is an extremely rare congenital anomaly and it is almost always fatal[1,2]. It may be found in association with other congenital anomalies. Rapid evaluation of the clinical signs and suspicion of the diagnosis at birth are of great importance for acute management[3]. In this article we report a preterm newborn infant with esophageal and laryngotracheal atresia with double TEF, in an effort to call attention to the clinical symptomatology and physical findings and to the importance of differential diagnosis and urgent management procedures.

Case Report

A preterm male infant of 32 weeks' gestation and birth weight of 1000 g was delivered by urgent cesarean section because of fetal distress to a 24-year-old gravida 1 para 0 mother. The pregnancy was complicated with polyhydramnios, and amniocentesis was performed for reducing amnionic volume a few hours before delivery. The Apgar scores were 2, 5 and 6 at 1, 5 and 10 minutes, respectively. The infant was noted to have an absent cry, cyanosis and respiratory distress. The attempt to intubate the infant failed but the lungs could be ventilated using bag-valve mask. The infant was immediately taken to the Neonatal Intensive Care Unit but attempts for intubation by senior neonatologists and then by the anesthesiology and otolaryngology consultants also failed. Efforts to place a nasogastric tube into the stomach were also unsuccessful. The infant was taken to the operating room for laryngoscopy. Direct laryngoscopy revealed complete occlusion of the airway immediately after the subglottic area, and esophagoscopy revealed EA. A contrast material was given to the esophageal pouch to show the relation between the trachea and esophagus radiologically. The X-ray showed a proximal EA with a thin TEF to the distal trachea and bronchi continuing with distal esophagus and stomach. A 2 FG endotracheal tube was inserted by tracheotomy to the distal trachea and the infant was transported back to the Neonatal Intensive Care Unit under mechanical ventilation. The X-ray examinations were also consistent with severe neonatal respiratory distress syndrome. Oxygen saturations by pulse oximetry were consistently low although maximum ventilatory support was applied. The infant died at the sixth hour of life. Postmortem X-ray examination confirmed the diagnosis (Fig. 1).

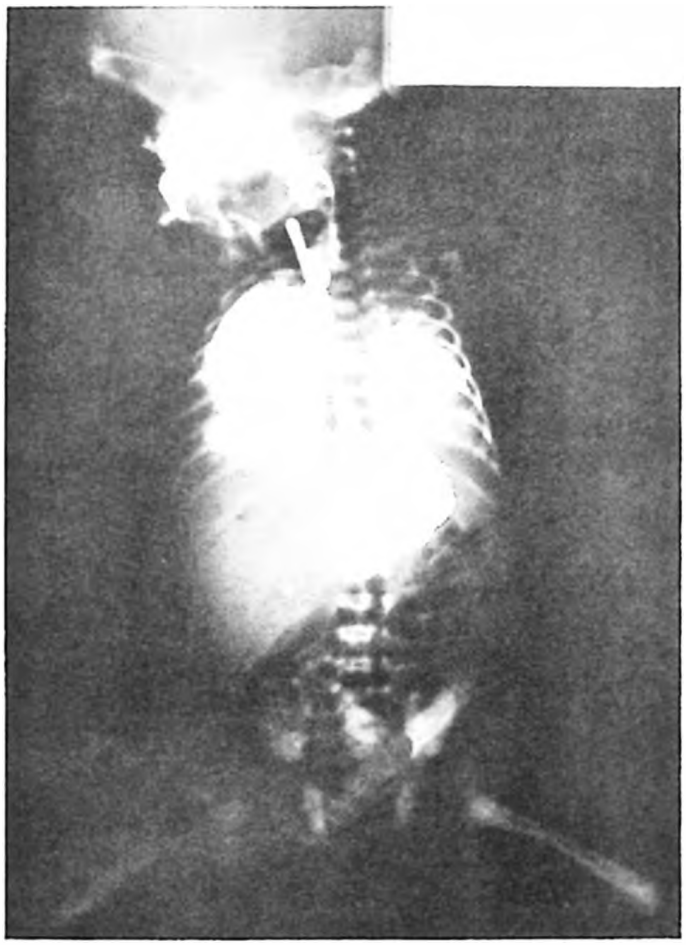

Fig. 1. Postmortem esophagography and bronchography of the newborn.

At autopsy, the larynx was normal until the level of vocal cords, after which the lumen was completely occluded as a consequence of laryngeal atresia. After an atretic segment of proximal trachea which was 1 cm in length, the distal trachea was normal. As for the esophagus, proximal atresia with proximal and distal TEF was present: the atretic proximal esophagus (3 cm in length) was connected to the distal trachea through a TEF of 0.1 cm in diameter. The tracheal bifurcation and main bronchi were opened to the distal esophagus by a very large fistula as if the esophagus was in continuity with the trachea (Vogt type IIc). The heart showed a complex combination of congenital malformations: left persistent superior vena cava, dilated right atrium, primum type atrial septal defect 0.4 cm in diameter and total anomalous pulmonary venous return. Left atrium was very small owing to total anomalous pulmonary venous return.

Discussion

Esophageal atresia with or without TEF is a relatively common congenital anomaly. However, EA with associated laryngotracheal atresia and with congenital cardiac malformations is an extremely rare condition[1,2]. Laryngotracheal atresia is almost always incompatible with life, more than 80 cases have been reported in the literature with only a few showing long-term survival[3].

Esophageal atresia can be found in association with laryngeal anomalies such as laryngeal stenosis and atresia, especially in Vog type IIb. The type of EA in our case was classified as Vogt type IIIc as it was associated with an extended form of laryngeal atresia and double TEF[4].

Experimental studies show that EA is probably caused by disorders in a system of folds in the tracheoesophageal space rather than abnormalities of a tracheoesophageal septum. In the past it was generally accepted that embryological differentiation of the esophagus from the trachea occurs when lateral ridges fuse in the midline forming a septum. But it has been shown that the differentiation of the foregut into the esophagus and trachea is a process of reduction in size of a foregut region called the tracheoesophageal space. This reduction is caused by a system of folds that develop in the primitive foregut. These folds approach but do not fuse. On the basis of this data it was concluded that malformations of the trachea or esophagus with fistula can be explained by abnormalities in the formation of the folds or their developmental movements[5]. In an adriamycin-treated animal model it was concluded that failure of the tracheal bud to develop normally from the primitive foregut is the main event which leads to the tracheoesophageal anomalies[6]. In another fetal rat model, exposure of fetal rat embryos to adriamycin has led to abnormal development of the notochord, including prolonged attachment to or fusion with the foregut and abnormal branching. Traction on the foregut by the notochord produces occlusion of its lumen and may result in its complete interruption. Separation of the notochord from the foregut would appear to be a prerequisite for the normal development of the foregut into its derivatives: the esophagus and trachea[7].

The prenatal presentation of infants with esophageal and laryngotracheal atresia has ranged from high-risk complicated pregnancies to completely uneventful ones. Polyhydramnios is one of the most significant prenatal findigs[3,5].

Although laryngotracheal atresia is a very rare congenital anomaly, this diagnosis should be suspected in any newborn with respiratory distress, absence of audible crying and difficult or impossible endotracheal intubation in the delivery room. Attempts at intubation will reveal an absent glottic or subglottic opening. No air movement with respiratory efforts is typically present and should alert the clinician to the possibility of laryngeal or tracheal atresia (TA). If the esophagus is normal and is intubated by accident when it is assumed that endotracheal intubation has been performed, insertion of the suction catheter to a greater depth than expected can be an indication of esophageal intubation. But, esophageal intubation generally allows relatively adequate ventilation in the short term through a TEF. Stabilization may also be achieved by bag-valve mask as in our case. Further respiratory difficulty or suspicion of the diagnosis generally leads to endoscopic examination of the infant or emergent neck exploration to identify the trachea and attempt a tracheotomy. Laryngoscopy, broncoscopy and esophagoscopy often confirm the diagnosis. More information can be obtained by a contrast study through the esophagus[3,5,8].

Esophageal and laryngotracheal atresias are usually complicated by many other congenital anomalies which worsen the prognosis. In a review of 89 patients with EA and TEF, genitourinary anomalies were present in 21%, cardiovascular in 19%, gastrointestinal in 10%, central nervous system in 9%, musculoskelatal in 8%, chromosomal in 5% and head and neck in 6% of cases. It is concluded that the survival rate of newborns with EA/TEF is high, especially in the absence of associated anomalies[9]. In another review, 52.4% of the patients with EA had associated congenital malformations. Early gestational age and lower birth weights are significantly correlated with higher rates of malformations[10]. Investigations for possible associated anomalies should be considered for all patients with EA, TA and TEF[11].

Infants with EA, with or without a TEF, are frequently of low birth weight. With advances in neonatal, respiratory, surgical and anesthetic care, more infants with very low birth weight are surviving. The most important risk factors are respiratory distress syndrome, pneumonia and major cardiac anomalies[12]. The therapy of the neonate with EA includes primary or delayed end-to-end anastomosis and different esophageal substitutes such as gastric, small intestinal and colonic transpositions, with good results[13,14]. Unfortunately, however, further management of laryngotracheal atresia has yielded no long-term survival. Intubation of the esophagus with either an endotracheal or tracheostomy tube provides a temporary airway. An attempt for a definitive repair is difficult none of the surgical approaches previously described has achieved adequate survival since a satisfactory tracheal prosthesis has not yet been developed[3,15].

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