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Case Report

Laryngeal cleft type IV: One pathology, two different presentations

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ABSTRACT

Laryngotracheoesophageal cleft is a rare congenital malformation where a communication exists between the aero-digestive tracts that is associated with high morbidity and mortality. In this case series, we describe our experience with two neonates evaluated in our institution with two diverse initial presentations and symptomatology of type IV laryngotracheoesophageal clefts. One patient presented with acute distress and respiratory failure due to bowel contents obstructing the trachea and the other presented with mild subcostal retractions and inspiratory stridor without overt respiratory failure. Most importantly, not every patient with type IV laryngotracheoesophageal clefts will present with fulminant respiratory difficulty. This prompted a literature review.

1. Introduction

Laryngeal cleft is a rare congenital malformation that arises due to arrested medial fusion of the lateral aspects of the laryngotracheal groove and cricoid cartilage [1]. When the cleft extends past the cricoid cartilage and into the trachea a direct communication with the esophageal lumen is established and it is more commonly termed laryngotracheoesophageal cleft (LTEC). The etiology of this defect is considered to be multifactorial [2]. LTEC is associated with other malformations and conditions, mostly of the aerodigestive tract such as laryngomalacia, tracheoesophageal fistula (TEF), tracheobronchomalacia and gastroesophageal reflux (GERD) [3,4]. Laryngeal clefts have been categorized according to the Benjamin and Inglis classification (Fig. 1) [5]. This classification is based on the caudal extension of the cleft and ranges from type I to type IV. Symptoms usually range from mild aspiration and stridor for type I and II, severe aspiration and respiratory distress for type III, and overt cardiorespiratory failure with a high morbidity and mortality index for type IV [6–8]. In this study, we describe two neonates with LTEC type IV who presented with different initial symptomatology, and based on our literature review, discuss how the same high grade malformation can present with different symptom severities.

2. Case presentation

2.1. Case 1

A preterm, adequate for gestational age, baby boy was born via induced vaginal delivery at 35 weeks of gestational age to a G1P1A0 24-year-old female with no history of systemic illness. A prenatal ultrasound at 28 weeks of gestational age showed reduced abdominal circumference, hypoplastic left lung, and herniated abdominal contents into the left hemithorax. A left diaphragmatic hernia was diagnosed, and patient was referred to the high-risk pregnancy clinics at our tertiary hospital. Neonatology service and pediatric surgery were consulted prenatally who recommended immediate endotracheal intubation after birth to prevent air entry into the bowel. Pregnancy was complicated by preeclampsia at 35 weeks and labor was induced. At birth, the infant had decreased breath sounds bilaterally, point of maximal impulse shifted to the right side, and a scaphoid abdomen with absent bowel sounds. APGAR was 6 and 8 at 1 and 5 minutes, respectively. The patient was immediately intubated with at 3.5 endotracheal tube in the delivery room by a neonatologist with adequate CO2 return. During intubation, medical staff noted an abnormal laryngeal anatomy. The patient was transferred to NICU and General Otolaryngology – Head and Neck Surgery service (OTO-HNS) was consulted.

During examination, the patient had no abdominal distention, trachea was palpated at midline and abundant mechanical ventilator air leak was heard coming from the oral cavity. Patient was found initially unstable with episodic desaturations and bradycardia episodes. A flexible laryngoscopy was initially performed which showed a discrete separation between esophagus and larynx at the esophageal inlet highly suggestive of LTEC. Patient was immediately taken to the operating room for rigid bronchoscopy and laryngoscopy. In the operating room, pulmonary CO2 return disappeared with minimal endotracheal tube manipulation so the decision was made to perform a tracheostomy due

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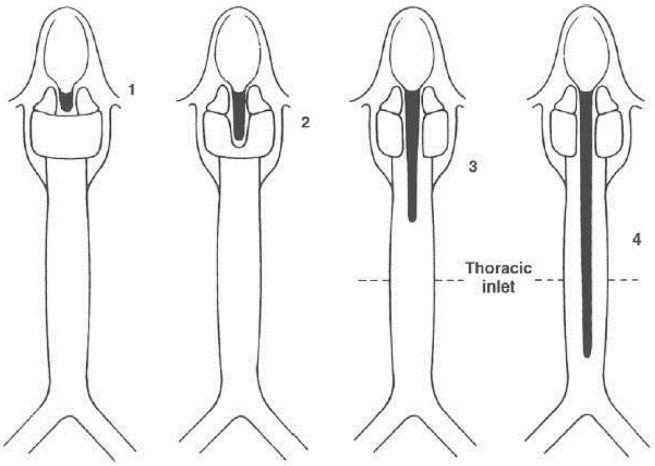

Fig. 1. Benjamin and Inglis' laryngeal clefts original classification. (1) Type 1, supraglottic interarythenoid cleft. (2) Type 2, partial cricoid cleft that does not extend completely below the posterior lamina of the cricoid cartilage. (3) Type 3, total cricoid cleft with or without extension to the cervical tracheoesophageal wall. (4) Type 4, involves a mayor part of the intrathoracic tracheoesophageal wall.
B. Benjamin, A. Inglis. Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol. 98 (1989) 417–420.

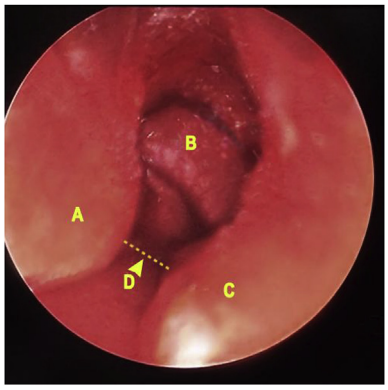

Fig. 2. Rigid bronchoscopy performed on the patient from case #1. Laryngotracheoesophageal cleft with small bowel loops occluding tracheoesophageal lumen. (A) Left arytenoid (B) Small bowel loops (C) Right arytenoid. (D) Laryngotracheoesophageal cleft.

to inadequate ventilation via endotracheal intubation and unable to tolerate neck extension for rigid bronchoscopy. The trachea was identified midline without evidence of malformation or any other anterior abnormality during the procedure. After airway was secured, a bronchoscopy was performed which showed an omega shaped epiglottis with a 3mm gap between the arytenoid cartilages extending down through the cricoid cartilage and posterior tracheal wall down to the carina, consistent with a LTEC type IV. Segments of small bowel were visualized inside the tracheoesophageal lumen proximal to the tracheostomy insertion site (Fig. 2).

The patient returned to the NICU on mechanical ventilation, however, he had cardiovascular arrest and was pronounced dead 48 hours after birth.

2.2. Case 2

A term, adequate for gestational age, baby girl was born at 37 weeks of gestation to a 19-year-old G3P1A1 with no history of systemic illness. A sonographic prenatal evaluation of the fetus showed absent gastric bubble and polyhydramnios with a high index of suspicion for

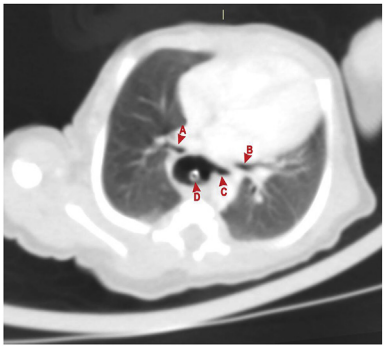

Fig. 3. CT scan performed on the patient from case #2 showing LV type IVb. (A) Right mainstem bronchus (B) Left upper lobar bronchus (C) Left lower lobar bronchus (D) Nasogastric tube in common trachea-esophageal cavity.

esophageal atresia as well as ambiguous genitalia. At birth, the neonate was found hypotonic, pale, and with poor respiratory effort. Stimulation along with bag-mask ventilation was provided and adequate peripheral saturation was obtained. APGAR scores of 4 and 8 at 1 and 5 minutes were documented.

The child was admitted to NICU, NGT was placed successfully and the patient was started on high flow nasal cannula. Abdominal X-ray and uncomplicated NGT placement suggested no signs of esophageal atresia, as previously suspected. Later on the same day, evaluation by NICU team found the patient had developed mild subcostal retractions with inspiratory stridor. FiO2 was increased and pediatric surgery service was consulted who recommended thoracic CT scan for better evaluation of the aerodigestive tract. Imaging showed a direct communication between trachea and esophagus with a cleft measuring 2.4cm long by 0.8cm extending from the proximal region of trachea/esophagus down to the level of carina and right mainstem bronchus (Fig. 3).

The OTO-HNS service was consulted and initial evaluation at NICU found patient with bilateral vocal cord paresis with vocal cords in the lateral position with otherwise normal anatomy without evidence of clefting. The patient was then taken to the operating room for rigid bronchoscopy and laryngoscopy. Examination showed a direct communication of the larynx, trachea, and esophagus extending down to level of carina and right mainstem bronchus as previously identified on CT scan. The neonate was extubated successfully after the procedure and returned to NICU for further workup and management. Functional imaging studies showed a patent foramen ovale, small patent ductus arteriosus, and ambiguous genitalia with a normal 46XX karyotype. Due to unavailability of an extracorporeal membrane oxygenation machine and a specialized surgical team to treat this patient at our institution, arrangements were made, and the patient was transferred to Cincinnati Children's Hospital where corrective surgical intervention was performed.

3. Discussion

Despite its rarity, two cases of LTEC type IV presented to our institution in short succession. The incidence of all grades of laryngeal clefts is estimated to be between 1 in 10,000 to 1 in 20,000 living births with a male:female ratio of 5:3 [2,3,9]. Laryngeal clefts represent between 0.2% and 1.5% of congenital anomalies of the larynx [2]. Type IV clefts represent the least common subtype and carries the highest morbidity and mortality rates [7]. LTEC type IV extends into the thoracic inlet and can be further subdivided into type IVa when it reaches the carina and type IVb if it extends into mainstem bronchi [5,10].

Laryngeal clefts are caused by a failure of two early embryologic processes: formation of the tracheoesophageal septum and fusion of the dorsal region of the cricoid cartilage [11]. Normally, the