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CASE SERIES AND REPORTS

Anterior laryngofissure approach in type III laryngotracheal cleft: a case report

Laringofissura anteriore nel cleft laringotracheale di tipo III: caso clinico

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SUMMARY

Laryngeal and laryngotracheal clefts are rare congenital malformations of the laryngobronchial tree. Their symptoms vary from mild cough to life threatening pulmonary aspiration and cyanosis. Type I and II clefts can be observed without surgical intervention, whereas type III and IV clefts usually require an anterior or lateral cervical approach. We present a case of type III laryngotracheal cleft seen in a 3-month-old male infant who died during revision surgery after an anterior laryngofissure approach. We discuss the difficulties in diagnosis, management and importance of anaesthesia for these rare anomalies in light of the current literature.

KEY WORDS: Larynx • Laryngeal cleft • Laryngotracheal cleft • Laryngofissure • Mortality

RIASSUNTO

I cleft laringei e laringotracheali sono rare malformazioni congenite dell'albero laringo-tracheo-bronchiale. La sintomatologia associata va dalla blanda tosse all'aspirazione e alla cianosi. I cleft di tipo I e II possono essere tenuti sotto osservazione senza intervenire chirurgicamente, mentre i tipi III e IV richiedono un approccio chirurgico anteriore o laterocervicale. Presentiamo il caso di un neonato di 3 mesi affetto da cleft laringotracheale di tipo III, deceduto in corso di revisione chirurgica dopo un approccio in laringofissura anteriore. Nel presente lavoro discutiamo, alla luce della letteratura, le difficoltà diagnostiche, le modalità di trattamento e le tecniche anestesiologiche relative a questa rara patologia.

PAROLE CHIAVE: *Laringe • Cleft laringeo • Cleft laringotracheale • Larinogofissura • Mortalità*

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Introduction

Congenital laryngeal anomalies are seen less than 0.5% of the population, and among these laryngeal clefts (LCs) account for 0.3-0.5%. These rare malformations are due to an embryological fusion defect of tracheo-oesophageal septum (TES). Early insult before 25 to 28 days of intrauterine life, which is the critical period for formation of TES, can cause incomplete formation of the septum. Defect in the fusion causes on-going relationship between the gastrointestinal and respiratory systems, which is the main cause of chronic cough or cyanosis during feeding and swallowing [1].

Since signs, symptoms and treatment options differ according to the severity of LCs, multiple classifications regarding the severity of disease have been proposed; that of Benjamin and Inglis is the simplest and most clinically applicable, which is used by most authors [2]. Type I is the interarytenoid cleft of soft tissue without involvement of cricoid cartilage. A type II cleft involves cricoid cartilage without involvement of the inferior lamina. Type III LC involves the entire posterior cricoid lamina with or without involvement of cervical trachea. Type IV is the total TES clefts with involvement of thoracic trachea. Type I is seen most often, while type IV was found in only 3% of cases in the series by Evans et al. [3]. The mortality rate is higher than than 90% for type IV clefts [1].

A 3-month-old male infant who died after revision anterior laryngofissure surgery for a type III LC due to anaesthesia complications is presented.

Case

A 3-month-old male infant was hospitalised in Mersin University paediatric intensive care unit due to frequent pulmonary infections after birth and cyanosis attacks after feeding. He was intubated for 7 days when consulted to our otorhinolaryngology department with the complaint

of a decrease in oxygen saturation after feeding with a nasogastric tube. Bedside examination was performed with a laryngeal blade. A posteriorly placed intubation tube was seen near the nasogastric tube. He had no additional illnesses or concomitant congenital malformations. Direct laryngoscopic examination in the operating theatre revealed a type III LC with involvement of the cricoid cartilage and first tracheal ring. A tracheotomy at the third tracheal cartilage was performed for protection and clearance of lower airways. After preparation of general anaesthesia and improvement of general health status and lung parenchyma, the infant underwent intervention for LC repair at the fourth month of life. An anterior laryngofissure approach was planned (Fig. 1). The LC was repaired with sternocleidomastoid muscle flap and three layers of closure (Fig. 2). In the postoperative period, proton pump inhibitor and antibiotic treatment including sulbactam-ampicillin was administered. He was discharged from hospital after recovery on fifth month of life with tracheostomy and nasogastric tube. Three months later, he was re-admitted with complaints of pulmonary infection. On direct laryngoscopic examination there was a cleft at interarytenoid area through the beginning of the cricoid cartilage. After preparation for general anaesthesia following treatment of pulmonary infection, he underwent revision surgery. The anterior laryngofissure approach revealed an approximately 1.5 cm defect at the previous suture line (Fig. 3). A sternocleidomastoid muscle flap and three layers of closure were used to repair the defect. During extubation, a sudden decrease in oxygen saturation to 60% was seen. Aspiration from tracheotomy was performed, and re-intubation and mechanical ventilation were administered by the anaesthesiologist. During aspiration, gastric fluid with saliva was seen although there was nasogastric tube during the surgical procedure. After deterioration of pulse oximetry levels, cardiopulmonary arrest developed. Although resuscitation was administered for one hour by the anaesthesiologist, spontaneous ventilation and cardiac beat could not be achieved and he was regarded dead.

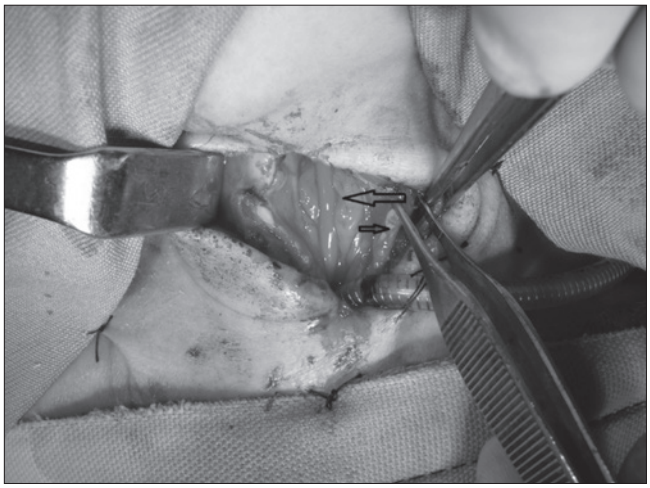

Fig. 1. Perioperative appearance of the patient during laryngofissure approach. The long arrow shows the cleft area with a nasogastric tube behind it. The short arrow shows dissected thyroid cartilage.

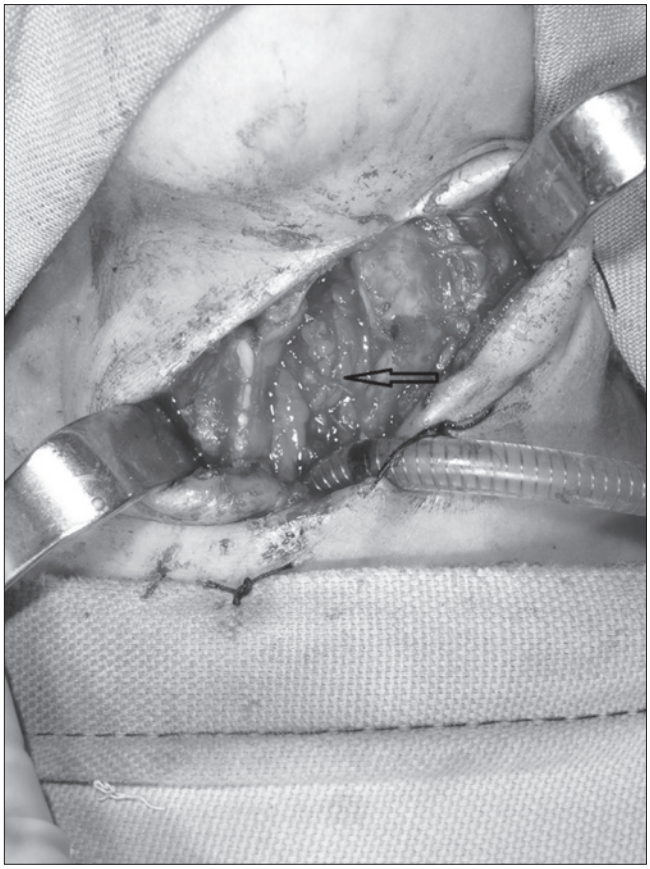

Fig. 2. Postoperative appearance of the patient. The arrow shows the three layer closed cleft area, while the nasogastric tube is behind the surgical wound.

Discussion

Laryngeal clefts are rare malformations and their incidence is challenging to accurately estimate since type I clefts can be asymptomatic or misdiagnosed for years; secondly, submucosal cleft, which is the absence of interarytenoid musculature with presence of the mucosa, can hide physical findings [4]. There is no known prenatal diagnostic tools, and only polyhydramnios can be a non-specific finding that makes diagnosis more difficult [1]. Signs and symptoms of LCs differ according to the severity of disease; most are respiratory in nature including chronic cough, aspiration, cyanosis and recurrent pulmonary infection associated with feeding. More serious symptoms such as severe respiratory infection and distress, stridor needing hospitalisation and even entubation as in our case are usually seen in type III and IV LCs [1 4]. Rarely, type III LCs can be misdiagnosed as asthmatic attack and treated accordingly with delayed diagnosis [5]. Posterior place-

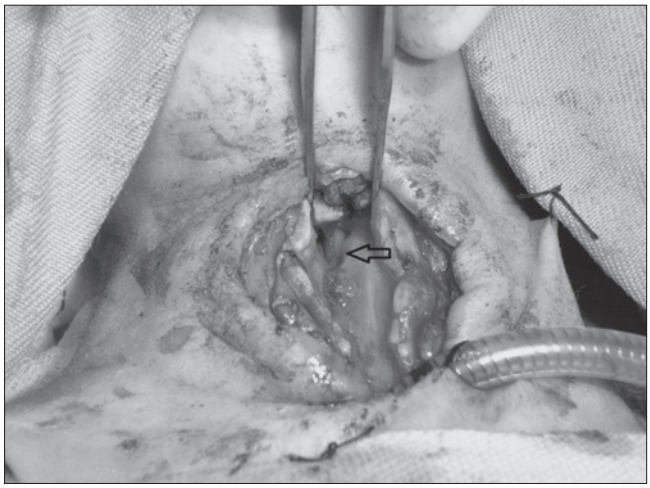

Fig. 3. Re-opened cleft area during revision with a laryngofissure approach. The arrow shows the cleft area with the nasogastric tube behind it.

ment of an endotracheal intubation tube and multiple unsuccessful oesophageal intubations can be seen as in our case [1].

Although LCs can be seen alone, concomitant multiple congenital malformations may also exist. Opits-Frias syndrome (oculo-gential-laryngeal or G syndrome) with airway cleft, cleft lip, cleft palate, swallowing dysfunction, imperforate anus and hypospadias can be seen in patients with LC. Pallister-Hall syndrome is another syndrome consisting of airway cleft, congenital hypothalamic hamartoblastoma, hypopituitarism, imperforate anus, and postaxial polydactyly that may be present with LC. VATER association (vertebral defects, anal atresia, tracheo-oesophageal fistula, oesophageal atresia, renal dysplasia) also commonly includes LC [6]. Overall, the most commonly accompanying anomalies are gastrointestinal; oesophageal atresia and tracheo-oesophageal fistula is the most common seen in 20-27% of cases, followed by genitourinary anomalies [1]. In our case there was no accompanying congenital malformation.

Microscopic examination of the larynx under general anaesthesia and palpation of interarytenoid area is the gold standard diagnostic tool for LCs. Barium swallow study and video fluoroscopy may miss aspiration of contrast agent for intermittent aspirating cases [6]. Functional endoscopic evaluation of swallowing is helpful in diagnosis as it provides the chance of evaluation of anatomic structure during swallowing including vocal cord motion and location of aspiration [4]. Pneumonia or peribronchial cuffing can be seen as a result of persistent chronic aspiration in chest X-ray [4].

Just as symptoms of LCs vary between cases, treatment options also differ according to severity of disease. Observation without surgical treatment can be a suitable option for type I and II cases with mild symptoms. For medical management, feeding therapy involving thickening of liquids and food consistency can be used. Gastro-oesophageal reflux can be seen with LCs and can cause oedema of mucosa postoperatively. Proton pump inhibitor for prevention is mandatory and endoscopic Nissen fundoplication can be an additional procedure for control of reflux [4 7]. In our case, high dose proton pump inhibitor was administered in the perioperative period. For symptomatic cases of type I and II clefts, or unresponsive to medical treatment endoscopic repair, can be considered. Flapping and suturing techniques of interarytenoid area are widely used [6]. More recently, injection laryngoplasty to the interarytenoid region can be an alternative technique for type I cases [8]. For type III and IV LCs, open cervical approaches are preferred. A posterior approach is used with a vertical incision along the anterior border of sternocleidomastoid muscle for achieving lateral pharyngotomy after retraction of major vessels. It does not provide visual access as good as that is provided by an anterior approach, and recurrent and superior laryngeal nerves are at great risk for damage. The anterior approach is the most commonly used and safe procedure done via a horizontal incision above the tracheostomy line if present. After retraction of strap muscles, a laryngofissure incision through thyroid cartilage is made. The cleft area can be repaired with interposing grafts such as sternocleidomastoid muscle flaps, pleura, pericardium, strap muscle, periosteum with two or three layers of closing the wound [4 6]. We also preferred an anterior cervical approach with laryngofissure incision and sternocleidomastoid muscle flap used as an interposing graft. Tracheotomy is widely used for type III and IV clefts for prevention of lung against aspiration after enteral feeding. Gastrostomy may also be performed for enteral feeding [4]. We also performed a tracheotomy before an open anterior cervical approach.

Anaesthesia is very critical during surgery for management of LCs. For endoscopic procedures, an endotracheal intubation tube can hinder the surgeon from studying at a well visualised area. Usage of tubeless anaesthesia and insufflation with spontaneous breathing and jet ventilation can be used [6]. After induction with inhalational agent, maintaining the general anaesthesia with propofol or remifentanyl, without using jet ventilation is also a possible option. In this situation, a modified cut-off endotracheal tube can be used near the laryngoscope blade to support ventilation [4]. Ferrari et al. recommends tubeless total intravenous anaesthesia (TIVA) with spontaneous ventilation during endolaryngeal management of LCs [9]. For open procedures, anaesthesia risks are more complicated. LC cases are in children whose lung functions are affected due to chronic aspiration. During reverse anaesthesia, children are prone to aspiration of gastric contents and thick salivary secretions to preoperatively affected pulmonary parenchyma [10]. In the series by Kluger et al.

with 133 cases, five children died postoperatively due to aspiration [11]. In data in 2011 from United Kingdom, pulmonary aspiration was the most common cause of anaesthesia-related death accounting for 50% of mortality [12]. In our case, bronchospasm and sudden deterioration of pulse oximetry was observed during reverse anaesthesia and extubation from tracheotomy. Although a post-mortem study could not be performed, we believe that gastric content and salivary aspiration to previously affected lung parenchyma was the cause.

In conclusion, LCs are rare malformations of the laryngobronchial tree. Some LCs can be life-threatening and early diagnosis is important to protect lung parenchyma from chronic aspiration. Type III and IV cases are prone to open surgical procedures. In the reverse anaesthesia and extubation period, these patients can aspirate secretions to previously diseased pulmonary parenchyma. Frequent aspiration of tracheotomy is important for releasing secretions. Extubation after ensuring that spontaneous ventilation and aspiration reflexes are achieved is critical for prevention of life threatening complications.

References

1 Chitkara AE, Tadros M, Kim J, et al. *Complete laryngotracheoesophageal cleft: complicated management issues.* Laryngoscope 2003;113:1314-20.

2 Benjamin B, Inglis A. *Minor congenital laryngeal clefts: diagnosis and classification*. Ann Otol Rhinol Laryngol 1989;98:417-20.

3 Evans KL, Courtney-Harris R, Bailey M, et al. *Management of posterior laryngeal and tracheoesophageal clefts.* Arch Otolaryngol Head Neck Surg 1995;121:1380-5.

4 Johnston DR, Watters K, Ferrari LR, et al. *Laryngeal cleft: evaluation and management*. Int J Pediatr Otorhinolaryngol 2014; 78:905-11.

5 Parsons DS, Herr T. *Delayed diagnosis of a laryngotracheoesophageal cleft.* Int J Pediatr Otorhinolaryngol 1997;39:169-73.

6 Clark K. *Anterior repair of posterior laryngeal and laryngotracheal clefts*. Operat Tech Otolaryngol Head Neck Surg 1999;10:42-52.

7 Kamata S, Ihara Y, Usui N, et al. *Surgical management for posterior laryngeal cleft developing subglottic airway stenosis*. J Ped Surg 2005;40:15-6.

8 Cohen MS, Zhuang L, Simons JP, et al. *Injection laryngoplasty for type 1 laryngeal cleft in children.* Otolaryngol Head Neck Surg 2011;144:789-93.

9 Ferrari LR, Zurakowski D, Solari J, et al. *Laryngeal cleft repair: the anesthetic perspective*. Paediatr Anaesth 2013;23:334-41.

10 Kelly CJ, Walker RWM. *Perioperative pulmonary aspiration is infrequent and low risk in pediatric anesthetic practice.* Paediatr Anaesth 2015;25:36-43.

11 Kluger MT, Short TG. *Aspiration during anaesthesia: a review of 133 cases from the Australian anaesthetic incident monitoring study (AIMS).* Anaesthesia 1999;54:19-26.

12 Cook TM, Woodall N, Frerk C: Fourth National Audit Project. *Major complications of airway management in the UK: Results of the fourth national audit project of the Royal college of anaesthetists and the difficult airway society. Part 1: anaesthesia.* Br J Anaesth 2011;106:617-31.

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