© Pediatr Med Rodz 2019, 15 (1), p. 81–84
DOI: 10.15557/PiMR.2019.0014

Muhammad Hazim Abdul Ghafar[1], Chow Xiao Hong[2], Priatharisiny Velayutham[2], Farah Hani Hassan[2]

Received: 13.06.2018
Accepted: 27.11.2018
Published: 31.05.2019

Laryngeal cleft type IV: a rare entity

Rozszczep krtani typu IV – rzadka jednostka chorobowa

[1] Department of Otorhinolaryngology – Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, Kota Bharu, Kelantan, Malaysia
[2] Department of Otorhinolaryngology – Head & Neck Surgery, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia
Correspondence: Muhammad Hazim bin Abdul Ghafar, Department of Otorhinolaryngology – Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, Kubang Kerian 161500 Kota Bharu, Kelantan, Malaysia, tel.: 013-3034060, e-mail: drhazim1986@gmail.com

Abstract

Laryngeal cleft is a rare disease. The manifestation of the disease and its management vary depending on the severity of the cleft. Laryngeal cleft should be considered in the differential diagnosis, especially in neonates presenting with stridor and recurrent aspiration. The outcomes in patients with laryngeal cleft have improved significantly over the years due to increased awareness of the disease, early detection, good intensive care and advancement in surgical techniques. We present a unique case of laryngeal cleft type IV. To the best of our knowledge, there is no reported case in the author's country, Malaysia.

Keywords: laryngeal cleft, congenital malformation, children

Streszczenie

Rozszczep krtani to rzadka nieprawidłowość. Zarówno objawy, jak i sposób leczenia różnią się w zależności od stopnia ciężkości anomalii. Rozszczep krtani należy uwzględnić w diagnostyce różnicowej, w szczególności u noworodków, u których występują świst krtaniowy i nawracająca aspiracja. Większa wiedza na temat choroby, wczesne wykrywanie, dobra intensywna opieka i rozwój technik chirurgicznych sprawiły, iż w ostatnich latach wyniki leczenia u pacjentów z rozszczepem krtani znacząco się poprawiły. W pracy przedstawiono wyjątkowy przypadek rozszczepu krtani typu IV. Zgodnie z naszą wiedzą w kraju autorów – Malezji – nie zgłoszono dotąd przypadku omawianej anomalii.

Słowa kluczowe: rozszczep krtani, wada wrodzona, dzieci

INTRODUCTION

Laryngeal cleft is considered a rare congenital malformation. It accounts for 0.5–1.6% of laryngeal abnormalities with an annual incidence rate of approximately 1 in 10,000 to 20,000 livebirths[1–3]. The disease shows a male predilection with a male to female ratio of 1.2:1 to 1.8:1[1].
Since its first description by Richter in 1792, the disease has been underdiagnosed for centuries. Pezzettigotta et al. proposed three reasons: 1) minor laryngeal cleft may be asymptomatic; 2) endoscopic diagnosis is difficult and the lesion is easily missed; 3) severe clefts can lead to child's death before a diagnosis can be made[1].

CASE REPORT

A male infant was born at full term via emergency caesarean section for transverse lie, with a good birth weight of 2.24 kg. Significant antenatal history included maternal anaemia in pregnancy and polyhydramnios. Despite good Apgar score (7 at 1 minute and 9 at 5 minutes), the child was admitted to Special Care Nursery (SCN) for close observation due to nasal flaring and grunting post-delivery. At 3 hours of life, the child showed evidence of worsening respiratory distress and non-invasive ventilation (NIV) via nasal continuous positive airway pressure (NCPAP) was required at 6 hours of life to maintain saturation. He was then referred to the tertiary paediatrics centre for further management.
Echocardiogram demonstrated a small patent foramen ovale and a small patent ductus arteriosus not in failure, thus the cardiac lesion was managed conservatively. Since the chest radiograph showed coiling of the Ryles tube with no gastric bubble seen, an upper gastrointestinal tract contrast study was done. It revealed distal short segmental narrowing of the oesophagus close to the gastroesophageal junction, causing partial obstruction with abnormal stomach configuration and small bowel loop distribution.
To further evaluate the child, an oesophagoduodenoscopy was performed, and revealed the absence of posterior wall at the level of vocal fold and extending up to the carina. A diagnosis of laryngotracheoesophageal cleft type IV was made, and a gastric transection at the level of the body of stomach with proximal gastrostomy to the left and distal gastrostomy to the right was performed in the same setting for feeding purposes.
The child was kept intubated in the neonatal intensive care unit (NICU) for a total duration of about two weeks, with multiple failed extubations and desaturation episodes complicated with pneumonia. In view of prolonged ventilation period, a second-look bronchoscopy was performed. Ventilation was achieved via fibre-optic nasal intubation technique in the contralateral bronchus, using size 2.5 endotracheal tubes. Bronchoscopy showed anomalous larynx with short epiglottis, absent vocal folds with arytenoids, absent subglottis and tracheal rings. The tracheobronchial tree was also anomalous with separated right upper and lower bronchial openings, small and oedematous left main bronchus opening, no sharp carina, and an oesophageal opening at the distal end of the combined lumen. A conversion procedure to type II laryngotracheoesophageal cleft was performed via a right thoracotomy approach. Tracheoesophageal reconstruction was achieved by using pleura as interpositioning tissue. The child received broad spectrum cephalosporins for recurrent pneumonia with blood cultures showing no growth.
Postoperatively the child was ventilated in NICU and feeding was gradually restarted. However, on day 53 of life the child developed sepsis with deranged coagulation profile. Since then the child experienced persistent desaturation which led to bradycardia. The child eventually died on day 56 of life.

DISCUSSION

The exact pathogenesis of laryngeal cleft is still poorly understood. One of the proposed hypotheses is that the underlying pathology of laryngeal cleft is associated with prematurely arrested development of the tracheoesophageal septum and, eventually, the lack of fusion of the two lateral centres of chondrification of the cricoid cartilage at week 8 of development[4,5].
Over the past 50 years, several classification systems have been proposed, such as Armitage in 1984 and Evans in 1985. However, the Benjamin and Inglis's classification[6] introduced in 1989 is most widely used. It is the only classification system that differentiates partial and total cleft of the cricoid cartilage and cervical and tracheothoracic cleft, which forms the basis for the choice of treatment and determining prognosis[1]. It describes four types of laryngeal cleft: type 1 is a supraglottic interarytenoid defect that extends inferiorly no further than the level of the true vocal folds; in type 2, the cricoid lamina is partially involved, with extension of the cleft below the level of the true vocal folds; type 3 is a total cricoid cleft that extends completely through the cricoid cartilage with or without further extension into the cervical trachea; and type 4 extends into the posterior wall of the thoracic trachea and may extend as far as the carina[6].
Patients with laryngeal clefts may present with either swallowing symptoms such as aspiration or cyanosis during feeding; pharyngolaryngeal symptoms such as toneless or weak cry and pharyngeal hypersecretions; respiratory symptoms such as stridor, respiratory distress at birth and recurrent pneumonia[1–3]. A literature review shows a great variability of clinical presentations: Glossop et al.[7] claimed that the pulmonary impact is mild to none in type I clefts as compared to Chien et al.[8] and Slonimsky et al.[9], who reported that cough, recurrent aspirations and pneumonia are the most common symptoms in type I cleft. Ojha et al. observed that choking on feeding was the most common symptom in type I clefts (19%), followed by aspiration on thin fluids (9%), chronic cough (8%) and hoarse voice (2%)[10].

DOI: 10.15557/PiMR.2019.0014

In another report, Myer et al. found that 21% of patients in the series had no symptoms in connection with laryngeal cleft and were diagnosed by accident[11].

Studies have shown that laryngeal cleft may be associated with other congenital abnormalities such as laryngomalacia, tracheomalacia, tracheoesophageal fistula, subglottic stenosis and gastroesophageal reflux. Other associated malformations may be gastrointestinal (16–67%), such as atresia with oesophageal fistula and imperforate anus; genitourinary (14–44%), such as hypospadias and kidney malformation; cardiovascular (16–33%), such as patent ductus arteriosus and ventricular septal defect; craniofacial (5–15%), such as cleft lip and palate; and bronchopulmonary (2–9%), such as bronchial stenosis and lung hypoplasia. Conditions such as vocal cord paralysis, laryngeal mobility disorder, reactive airway disease and central neurogenic swallowing disorder should be included in the differential diagnosis of these patients. A higher incidence of laryngeal cleft is also reported with Pallister–Hall syndrome, Opitz–Frias syndrome, VACTERL association and CHARGE syndrome[1–3,6–14].

Extensive works have been conducted to establish a comprehensive diagnostic and management algorithm for laryngeal cleft[8,10–12]. It is recommended that all patients with a clinical history consistent with laryngeal cleft should be thoroughly investigated, which includes a careful history and physical examination, chest X-ray, modified barium swallow (MBS) and/or fiberoptic endoscopic evaluation of swallowing (FESS), and endoscopic examination under general anaesthesia.

Chest X-ray may demonstrate pulmonary infiltrates secondary to aspiration[12]. Both MBS and FESS are valuable tools to assess aspiration; each with its own advantages and limitations. Although MBS has the advantage of being a non-invasive test, it offers inferior resolution of the detailed laryngeal anatomy. On the other hand, FESS offers direct visualisation of the larynx during swallowing, which allows for a better assessment of the mobility of the vocal cords, the amount of pooling and penetration of both secretions and liquid/food substances, and the location of aspiration[8]. FESS may also help identify neurological aspiration where there is lateral spillage and aspiration over the aryepiglottic fold as opposed to posterior to anterior aspiration. However, Ojha et al. reported that children under 4 years are often distressed during the FESS procedure with episodes of crying, rendering the conclusion about baseline feeding difficult to interpret[10]. It is important to note that that the results of MBS and FESS may be normal in patients who aspirate intermittently, and thus a normal result in either test does not definitively rule out laryngeal cleft. However, Rahbar et al. advocate that MBS and FESS offer complimentary diagnostic information and should be performed in the work-up of laryngeal cleft[12].

Intraoperative endoscopy remains the gold standard in the diagnosis of laryngeal cleft[8,10,12,14]. Direct laryngoscopy is first performed to rule out tracheoesophageal fistula, subglottic stenosis and other associated anomalies mentioned previously. Subsequently, a blunt laryngeal probe is used to part the arytenoid and remove redundant laryngeal and/or oesophageal mucosa prolapsing into the laryngeal cleft[12]. The interarytenoid area is then palpated using the probe to assess the depth of the interarytenoid groove. If the cleft extends below the vocal cords, the cricoid is palpated for evidence of dehiscence. If the cricoid cartilage is not palpable with the probe, the cleft is considered at least type III and extends into the cervical or thoracic trachea[14].

The management of laryngeal clefts ranges from medical management to surgical intervention. The two aims of medical management are: 1) to maintain optimal respiration, prevent pulmonary complications of recurrent pneumonia and to ensure adequate nutrition; 2) to treat comorbid medical conditions, especially gastroesophageal reflux disease (GERD), which significantly improves surgical outcomes. This approach includes maintaining an upright position during feeding, feeding with thick liquids and the use of proton pump inhibitors (PPIs)[8,13]. Ojha et al. reported that 36% of their patients who received conservative management showed overall resolution[10]. In another report, Chien et al. observed that 80% of their patients showed no significant improvement with conservative measures alone, and required surgical intervention[8].

Surgical intervention is indicated for type I clefts that fail medical treatment. Cleft type II, III and IV will almost certainly require surgical management since aspiration is inevitable. There are two surgical approaches to the reconstruction of laryngeal clefts – endoscopic approach and open surgical repair. For type I, II and selective type III clefts, endoscopic techniques may be considered. Open surgical management is advocated for most type III and IV clefts[13]. Open surgical repair may be achieved via cervical or thoracic approaches. Three types of cervical approaches have been proposed: 1) the lateral approach with pharyngotomy; 2) the lateral approach with posterior pharyngotomy; 3) the anterior translaryngolateral approach. Three types of thoracic approaches have been reported: 1) right posterolateral thoracotomy combined with right cervical approach; 2) partial upper sternotomy combined with cervical incision; 3) anterior cervicothoracic approach[1]. Several techniques for cleft closure have been described, including a two-layer closure with either symmetric or asymmetric flaps and, also, complete separation of the tracheal and oesophageal lumens. Various types of interposition grafts have been used to provide tension-free closure and avoid the risk of breakdown and fistula, including pleura, pericardium, sternocleidomastoid muscle flaps, strap muscle, jejunum, temporalis fascia and tibial periosteum[1,13].

There is no consensus regarding the timing and approach for surgical repair of laryngeal cleft. It depends on the severity of symptoms, associated abnormalities, and the type of cleft[12]. In the present case, the cleft was initially repaired from type IV to type II via an open surgical procedure using the right thoracotomy approach and pleura as interposition graft. We have arranged to re-assess the child via direct

 DOI: 10.15557/PiMR.2019.0014

laryngoscopy and bronchoscopy with a view to a second-stage surgical procedure either by an endoscopic or open approach. Unfortunately, the child had succumbed to other co-morbidities prior to the surgery.

Several possible complications of surgical repair include laryngeal nerve injury, granulation tissue formation, oesophageal stricture, wound dehiscence, anastomotic leak, supraglottic stenosis and deepening of the initial cleft[8]. It has also been reported that unresolved feeding issues may continue for a short period postoperatively despite successful surgical repair of the cleft, due to neurological disturbance of the swallowing reflex[12].

The prognosis for laryngeal cleft has improved significantly with the advances in medical care and surgery. However, the success and survival largely depend on the type of cleft. In 1983, Roth et al. reported a mortality rate of 43% for types I and II, 42% for type II, and 93% for type IV[14]. Myer et al. found that 67% of the patients who died had another congenital malformation. A combination of anomalies and associated diseases drastically worsens the prognosis[11]. Most recent series found a mortality rate from 6% to 25%[1]. The improvement in terms of survival was most tremendous in cleft type IV, for which the mortality decreased from 93% in 1983[14] to 50% in 1996%[15]. It is known that early diagnosis, airway protection, treatment of reflux and multidisciplinary care are the principal elements in improving outcomes.

CONCLUSIONS

In summary, laryngeal cleft is a rare disease that is exceptional in the paediatric age group. The manifestations and the management of the disease vary depending on the severity of the cleft. Laryngeal cleft should be considered as one of differential diagnoses, especially in neonates presenting with stridor and recurrent aspiration. The outcome of patients with laryngeal cleft has improved significantly over the years due to increased awareness of the disease, early detection, good intensive care and advancement in surgical techniques. As documented in the present case, the patient managed to survive up to 6 weeks of life. Regrettably, the child had succumbed to other comorbidities.

Conflict of interest

The authors do not declare any financial or personal links with other persons or organisations that might adversely affect the content of the publication or claim any right to the publication.

References

1. Pezzettigotta SM, Leboulanger N, Roger G et al.: Laryngeal cleft. Otolaryngol Clin North Am 2008; 41: 913–933.
2. Watters K, Ferrari L, Rahbar R: Laryngeal cleft. Adv Otorhinolaryngol 2012; 73: 95–100.
3. Strychowsky JE, Rahbar R: Laryngotracheoesophageal clefts. Semin Pediatr Surg 2016; 25: 128–131.
4. Blumberg JB, Stevenson JK, Lemire RJ et al.: Laryngotracheoesophageal cleft, the embryologic implications: review of the literature. Surgery 1965; 57: 559–566.
5. Lim TA, Spanier SS, Kohut RI: Laryngeal clefts: a histopathologic study and review. Ann Otol Rhinol Laryngol 1979; 88: 837–845.
6. Benjamin B, Inglis A: Minor congenital laryngeal clefts: diagnosis and classification. Ann Otol Rhinol Laryngol 1989; 98: 417–420.
7. Glossop LP, Smith RJ, Evans JN: Posterior laryngeal cleft: an analysis of ten cases. Int J Pediatr Otorhinolaryngol 1984; 7: 133–143.
8. Chien W, Ashland J, Haver K et al.: Type 1 laryngeal cleft: establishing a functional diagnostic and management algorithm. Int J Pediatr Otorhinolaryngol 2006; 70: 2073–2079.
9. Slonimsky G, Carmel E, Drendel M et al.: Type I–II laryngeal cleft: clinical course and outcome. Isr Med Assoc J 2015; 17: 231–233.
10. Ojha S, Ashland JE, Hersh C et al.: Type 1 laryngeal cleft: a multidimensional management algorithm. JAMA Otolaryngol Head Neck Surg 2014; 140: 34–40.
11. Myer CM 3rd, Cotton RT, Holmes DK et al.: Laryngeal and laryngotracheoesophageal clefts: role of early surgical repair. Ann Otol Rhinol Laryngol 1990; 99: 98–104.
12. Rahbar R, Rouillon I, Roger G et al.: The presentation and management of laryngeal cleft: a 10-year experience. Arch Otolaryngol Head Neck Surg 2006; 132: 1335–1341.
13. Johnston DR, Watters K, Ferrari LR et al.: Laryngeal cleft: evaluation and management. Int J Pediatr Otorhinolaryngol 2014; 78: 905–911.
14. Roth B, Rose KG, Benz-Bohm G et al.: Laryngo-tracheo-oesophageal cleft. Clinical features, diagnosis and therapy. Eur J Pediatr 1983; 140: 41–46.
15. Simpson BB, Ryan DP, Donahoe PK et al.: Type IV laryngotracheoesophageal clefts: surgical management for long-term survival. J Pediatr Surg 1996; 31: 1128–1133.

DOI: 10.15557/PiMR.2019.0014