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CASE REPORT

Laryngotracheal clefts (LC) as an Uncommon cause of Stridor in an infant: Case report

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ABSTRACT

Stridor is conventionally thought to be of inspiratory causes in nature and usually due to airway pathology which leads upper airway obstruction in infants. The present case report illustrates that stridor would be caused by Laryngotracheal clefts (LC) which can be resulted in respiratory distress, stridor, choking episodes, coughing and recurrent croup as well as recurrent aspiration and respiratory tract infections.

***Keywords**: LC, Stridor, SALs*

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INTRODUCTION

Stridor in children is usually due to airway pathology of which would be of the main symptom of upper airway obstruction in infants [1]. This lead to a high-pitched sound caused by turbulent air flowing through a narrowed airway [2], Which is considered to be of congenital or acquired causes in terms of acute or chronic phases in neonates [3]. Laryngomalacia and secondary airway lesions (SALs) are the most common cause of Stridor which are followed by other causes, namely vocal cord palsy, tracheal anomaly, subglottic stenosis, vascular and lymphatic malformation, craniofacial abnormalities, laryngeal papillomas, laryngeal cleft and even head and neck tumors [2, 4]. We particularly more focused on the Laryngotracheal clefts (LC) as the main cause of the consequent stridor in children.

CASE REPORT

2-month and 13-day old female infant was referred to Dr. Sheikh children's hospital, in Mashhad complaining ofcough, cyanosis attack and wheezing occurred during breast feeding. She was the first child, born via normal vaginal delivery (NVD), with birth weight of 3200g and a good Apgar score

fallowing aphonia at birth, cough, wheezing and cyanosis attack after breast feeding was hospitalized at Birjand hospital after which without definitive diagnosis discharged one week later.
Infant's mother had genital warts and preeclampsia (PE) bythe last week of pregnancy ofwhich no history of drug usage during pregnancy were reported; however it is worth saying that she had appropriate care during pregnancy and no problem during pregnancy and childbirth was mentioned.

CLINICAL EXAMINATION

Infant was not ill, feverous without toxic shock syndrome's (TSS) signs. She had stridor respiration during sleep position of which got worsened with prone position. Results showed in the table 1below.Physical exam were assessed as below; normal Junction Plakoglobin (Jup), Icterous status, enlargement of the thyroid, although neck mass and specially freckles were negative. Auscultation of lungs revealed bilateral coarse crackle without wheezing and late inspiratory rales. Auscultation of heart sounds showed no heart murmurs at S_1 and S_2 sounds. Abdomen examination was normal with not having any abdominal distension and organomegaly. Genital organs were normal. Chest x-ray demonstrated no abnormal signs, cardio thoracic ratio (CTR) was of normal value. Haziness showed in the upper and middle sides of right lung.The other bone structures and soft thoracictissues were normal. Diagnostic fiber optic bronchoscopy demonstrated discontinuity of the posterior of vocal cards additionally posterior trachea had cleft with the length of 1-1.5 cm with tracheomalacia. Carina, right and left bronchial and subsegmental bronchial were showed natural.
Therapeutic interventions for this case included:

1) Serum therapy
2) Methyl prednisol AP (2.5 mg/BD/IV)
3) Ranitidine AP (12-35 mg/Q8h/IV)
4) Drop Nasal prednisolone (1-2qtt/ Q8h for 3 days/IN)
5) Drop Methoclopramid (2-3qtt/po/ Q8h)
6) Apclindarycin (50mg/IV/ Q8h)
7) SuspErythroy (1.5 ml/po/ Q8h)
8) Cap omeprazole (30granele/po/Qday)
9) Nebulizes Epinephrin (2 ml/Q6h)
10) Head elevation

DISCUSSION

Having considered that the Stridor would be of the inspiratory sources in nature regarding the conventional thought it is now believed that expiratory or biphasic ones in both phases of the respiratory cycle could be emerged in the same consequences. Stridor separated into acute and chronic processes(5).The former is most likely caused byinfectious agents in addition to other causes of which can be presented as viral croup,for instance, or epiglottitis, retropharyngeal abscess and foreignbody (6). Croup occurs specially in children aged 6 months to 2 years old whose stridor can be mild, occurring only with crying, or in severe cases, can occur at rest with severe respiratory distress(5).
The latter especially in neonate and infancy often represents as congenital laryngeal anomalies including subglottic stenosis, subglottic hemangioma, laryngomalacia and vocal cord paralysis [6]. Laryngomalacia can prevail at birth but usually presents at 2 to 4 weeks of age. Stridor of laryngomalacia is exacerbated when the infant lays supine, cries, or when being fedwhereas it is alleviated when the infant is prone [5]. Congenital laryngeal anomaliesare less frequent but can be accompanied simultaneously with airway lesions and various comorbidities, and these may be important factors for management including surgical intervention and the outcome. Therefore, the presence of synchronous airway lesions and status of comorbidities should be considered as the best possible outcome for each patient eventually [6].
Laryngotracheal clefts (LC) are congenital malformations of the upper aerodigestive tract, resulting from the unusual communication of posterior fusion of the larynx and trachea, which can extend into the esophagus [7].This malformation creates an unsuitable communication between the larynx and hypopharynx, esophagus which can cause aspiration, dysphagia, and pulmonary complications [8]. LC can cause respiratory distress, stridor, choking episodes, coughing and recurrent croup as well as recurrent aspiration and respiratory tract infections [9]. The severity of the symptoms correlates with the severity of the cleft. Clefts with Lower grade can be asymptomatic, but when symptomatic, chronic cough, wheeze, present most frequently with feeding difficulties, recurrent chest infections and stridor [10].
Benjamin and In glis classified LC into four types in 1989 [11] and modified by Sandu in 2006 [12]. Indeed, they differentiate partial and total cleft of the cricoid cartilage. Those elements are of prodigious importancefor making a decision on therapeutic strategy.

Type 0: submucosal cleft
Type I: supraglottic, interarytenoid cleft, above the vocal fold level
Type II: cleft extending below the vocal folds into the cricoid cartilage
Type III a: cleft extending through the cricoid cartilage but not into the trachea
Type III b: cleft extending through the cricoid cartilage and into the cervical trachea
Type IV: cleft extending into the thoracic trachea, potentially down to the carina (figure 1) (9).
It would be of though matter as it comes to diagnosis this diseasesince its symptoms are unspecific and obscure(9, 13).Type 0 LC were diagnosed at an average age of 6 months and may display mild to no obvious symptoms. Their diagnosisparticularly is difficult as ifmaybe their clinical appearance would be assessed coincidently during other purposes including endoscopy or an external procedure initiated for other reasons of which could be overshadowed by being not aware of their symptoms by the surgeon [14, 15].
Type I clefts were diagnosed before the age of 6 months and usually present with mild to moderate symptomsincludingswallowing disorders, stridor and a toneless or hoarse cry. During feeding there is possibilities regarding the occurrenceof cough, aspiration, cyanosis and dyspnoea [16]. They often go misdiagnosed for long periods and usually surface at tertiary care institutions. Direct laryngoscopies have incidence rate ranges from 0.2 to 7.6% for patients with recurrent respiratory symptoms [17]. Medical management for type I clefts comprises of feeding therapy bymaximizing liquids concentrate and food consistency or swallowing evaluation [18]. Treating comorbid medical conditions that overlap with or contribute to swallowing dysfunction is of a secondary medical management for these patients. Specifically, treatment for reactive airway disease with proton pump inhibitor therapy in addition to treatment of food allergy which can improve aerodigestive edema and irritation and, consequently, swallow function thus, many authors do agree that it is most likely that surgical repair could be avoided because of such currentmedical management in type I clefts [10, 19].
Type II and III clefts usually display more swallowing disorders (aspiration mostly) and pulmonary tract infections. Type II clefts were diagnosed before the age of 2 months [16].Watters et al. in 2007 [20] and Ojha et al. in 2014 [21] reported that 50–88% of children with laryngotracheal cleft undertaken endoscopy with pediatric otolaryngology who were also under surveillance of pulmonaryand gastroenterology system examinations including laryngoscopy, bronchoscopy, and esophagoscopy tests. There is increased risk of aspiration in some cases of type III so that parenteral nutrition may temporarily be needed [9]. Endoscopic closure of type III clefts successfully had performedby 1996 and2009 in some casesof survey conducted by Pinlong *et al*. [22] and Rahbar *et al.* [10] respectively.

Kind trail	Result
Pulse Rate (PR)	122/min
Respiratory Rate (RR)	35/min
Temperature (TP)	36.6/min
Blood Pressure (BP)	80 mm/Hg
Weight (WT)	4.80 Kg
Height (HT)	57 cm
Head Circumference (HC)	39 cm
CBC	
WBC	13100
Neutrophils	20%
Lymphocyte	78%
Eosinophil	2%
Hematocrit	33.7%
Hemoglobin	10.6
Platelet count	461000
PT	12.6"
PTT	38"
INR	1.0
Blood group (BG)	O^+
BS	73
Urea	14
Cr	0.4
Na	135
K	5.3

Table 1: Summary of the clinical findings for infant

Type IV encountered practical difficultiesin maintaining correct mechanical ventilation of which had a poor prognosis, since it is precipitated by the early respiratory distress (23, 24). In type III and IV clefts mostly tracheobronchomalaciacan be seen(9). Roth et al. reported mortality rate ranging from around 43% for types I, II and III to 93% for type IV, and thus Type IV had a high mortality rate [25]. treatment of type IV LCs were consisted of both cervical and thoracic approaches simultaneously of which may require extracorporeal membrane oxygenation or cardiopulmonary bypass [26, 27] additionally this also can be performed by the means of Gastric division with a proximal drainage tube and distal gastrostomy [28].
Surgical repair for type I clefts is useddue to medical treatment strategies failure. Cleft types II, III, and IV are usually a foregone conclusionof aspiration; therefore almost all of these cases could end up with surgical therapy [29]. Regarding type IV and paediatric surgical procedure many literatures have described an anterior sternotomy approach or transthoracic approach with possible need to use cardiopulmonary bypass [30]. Endoscopic repair should be considered for type I, II and selective type III clefts. However, Numerous publications have described the endoscopic technique for the closure of type I and type II LC [10, 14, 29, 31] whereas Most type III and all type IV clefts are treated with open thoracotomy procedures[10, 12, 19].
Laryngotracheal clefts (LC1) are congenital malformations of the larynx, whose prognosis is highly dependent on the extension of the cleft and other associated malformations, however stridor is due to laryngomalacia and exacerbated when the infant lays supine, cries, or when feeding and is alleviated when the infant is prone. Type I and, sometimes, type II clefts can have an insidious presentation with persistent vague respiratory and feeding difficulty and commonly Endoscopic repair is preferred for them. Type III and IV laryngeal clefts are of significant respiratory distress and coincidently of congenital anomalies of which surgical repair would be needed according to their management.

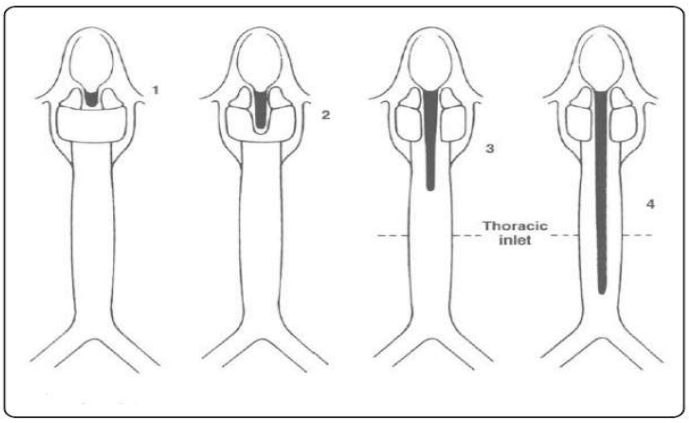

Figure 1: Benjamin and Inglis' original classification [11]

CONCLUSION

In this case report, Laryngotracheal clefts (LC)reported tobe the cause ofrespirationstridor whichsuccessfully was treatedregardless of surgery. Type I laryngeal clefts can be managed medically or surgically depending on the degree of morbidity whereas Types II, III, and IV require endoscopic or open surgery to avoid chronic respiratory and feeding complications.

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