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Case report

Management of supraglottic dysgenesis presenting as laryngomalacia

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ABSTRACT

Laryngomalacia is a common source of stridor and can lead to significant upper airway obstruction and feeding disturbances in infants. We describe a unique case of supraglottic dysgenesis presenting as laryngomalacia featuring a prominent "s-shaped" epiglottis with both posterior edges fused to the right aryepiglottic fold/arytenoid complex. Although this anomaly is not accounted for in any of the current laryngomalacia classification schemes, modified laser supraglottoplasty was a satisfactory approach leading to successful decannulation. Laryngeal embryology and possible timing of the pathogenesis of this rare occurrence are reviewed as well.

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1. Introduction

Laryngomalacia is the most common congenital malformation of the larynx. It is also the most common cause of stridor in neonates and infants [1]. Manifestations include an inspiratory high-pitched stridor associated with abnomally flaccid supraglottic laryngeal tissue, which collapses inward on inspiration. The diagnosis is established with laryngoscopy. The majority of children present with mild symptoms that resolve between the ages of 2 and 5 years [1]. Surgical intervention may be indicated however for more severe disease associated with obstructive sleep apnea, failure to thrive, severe reflux, cor pulmonale, or lack of anticipated spontaneous resolution [2]. While laryngomalacia is generally classified with respect to the degree and/or location of supraglottic flaccidity, this schema may be limited owing to the multitude of underlying pathophysiologies in this disease process [3]. Here we present a unique form of laryngomalacia related to supraglottic dysgenesis which is not accounted for by any of the current classification systems. Successful management with modified carbon dioxide (CO_2) laser supraglottoplasty and the embryologic origins of this anomaly are described as well. This report was granted exemption from institutional review board approval.

2. Case report

A 13-month-old girl with a tracheostomy was referred to our outpatient pediatric otolaryngology clinic from an outside institution for evaluation of her airway. The patient had a past medical history significant for premature birth at 24 weeks of gestation complicated by bronchopulmonary dysplasia and reactive airway disease. Management in the neonatal intensive care unit included brief endotracheal intubation at birth for 18 h to rule out sepsis. Beginning at 2 months of age, the patient began experiencing multiple incidences of self-resolving stridor. These symptoms were investigated with two rigid bronchoscopies at an outside facility with no definitive diagnosis. The stridorous episodes continued and eventually culminated in an episode of respiratory decompensation managed with emergent tracheostomy at age 6 months. Progressive deficits in swallowing without aspiration on modified barium swallow were attributed to underlying cognitive deficits combined with her airway anomaly. A gastrostomy tube was placed at 11 months of age. Accurate assessment of phonation was limited due to the presence of the tracheostomy.

Initial fiberoptic laryngoscopy in our clinic demonstrated an ill-defined anomaly of the supraglottic structures that was further investigated in the operating room (OR) with a direct laryngoscopy and rigid bronchoscopy. Close inspection revealed an abnormality of the epiglottis and aryepiglottic (AE) folds (Fig. 1). The left side of the epiglottis appeared to be rotated over to join the right side of the epiglottis, pulling the left aryepiglottic and left pharyngoepiglottic folds with it. This also resulted in an asymmetry with shortened AE folds on the right side. The arytenoid cartilages were

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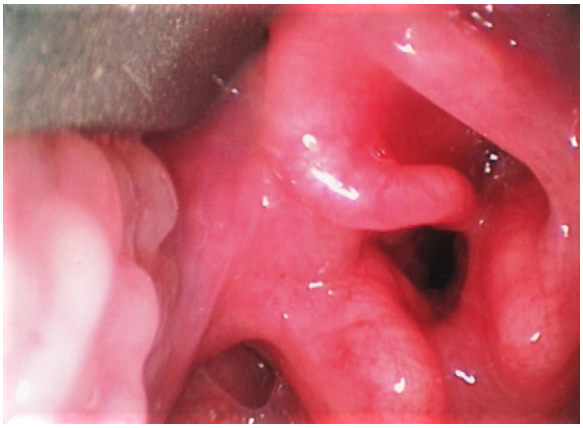

Fig. 1. Supraglottic dysgenesis visualized on direct laryngoscopy with both posterior edges of the epiglottis fused to the right arytenoid complex.

noted to be in good position with spontaneous movement, indicating at least some degree of vocal fold mobility on both sides. The pyriform sinuses and postcricoid region appeared to be normal as well. Acquired subglottic stenosis with subglottic narrowing and suprastomal granulation tissue without tracheomalacia was also noted. Correction of the supraglottic abnormality took place with a carbon dioxide (CO_2) laser set to 3 W. The redundant epiglottic tissue was removed from its posterior attachment, allowing it to swing forward and heal in its intended position. Both AE folds were also lysed to further facilitate epiglottic repositioning. Six weeks following this initial procedure, the patient was returned to the OR for a repeat direct laryngoscopy, rigid bronchoscopy, and revision supraglottoplasty (Fig. 2). The CO_2 laser set to 3 W was this time used to trim down redundant epiglottic tissue in an effort to prevent collapse during inspiration. The suprastomal granuloma was addressed with a microdebrider set to 1000 rpm. The subglottic lumen was then dilated with a 10 mm balloon. Successful decannulation took place shortly afterwards with no subsequent stridor or respiratory difficulty at 7 months follow-up. Voicing was satisfactory as well. Although swallowing did improve, the gastrostomy remained due to cognitive limitations.

3. Discussion

While the etiology of laryngomalacia remains largely unknown, four primary causes have been suggested: cartilage immaturity, anatomic abnormalities, neuromuscular immaturity, and the presence of gastroesophageal reflux disease (GERD) [2].

Proposed classification schemes reflect the most frequently encountered sites of supraglottic collapse (anterior, posterior, lateral, or any combination of the three). Holinger and Konior [4] defined laryngomalacia based on the direction of supraglottic collapse: posterolateral (type A), complete (type B), or anterior (type C). In an attempt to account for severity in addition to location of collapse, Lee et al. [3] created an expansion of the Holinger system as group I (type A only), group II (type B or B + A), or group III (type C, C + A, or C + B + A). Kay and Goldsmith [5] further classified laryngomalacia anatomically and functionally as: type 1 (foreshortened or tight AE folds), type 2 (presence of soft tissue in the supraglottis), or type 3 (other functional etiologies, such as neuromuscular disorders). Features on our patients intraoperative and awake fiberoptic laryngoscopic examinations of excessive/obstructive epiglottic tissue and foreshortened AE folds are most consistent with a Holinger type A and C, Lee group III, or Kay type 1 and type 2. The unilateral fusion of the epiglottis to the right arytenoid/AE fold complex observed in our case (Fig. 3) however, is not accounted for any of the known classification systems.

A brief review of laryngeal embryology may shed light on the most likely timing of our encountered anomaly (Table 1) [6]. We hypothesize that the supraglottic dysgenesis observed in our patient took place during days 30–32 of gestation, where the hypobranchial eminence and primitive arytenoid swellings gave rise to the epiglottis and aryepiglottic folds, respectively.

Surgical techniques for laryngomalacia generally involve supraglottoplasty for posterior and lateral supraglottis, and epiglottopexy with or without supraglottoplasty for obstruction anteriorly [2]. In terms of supraglottoplasty, satisfactory results have been achieved with both cold knife or laser approaches [7,8], although these techniques have never been compared prospectively. In this matter, success may lay in the surgeon's judgment, personal preference, and technical skills rather than the specific surgical tools. Despite the unique presentation as a supraglottic dysgenesis, we were pleased with the positive response to

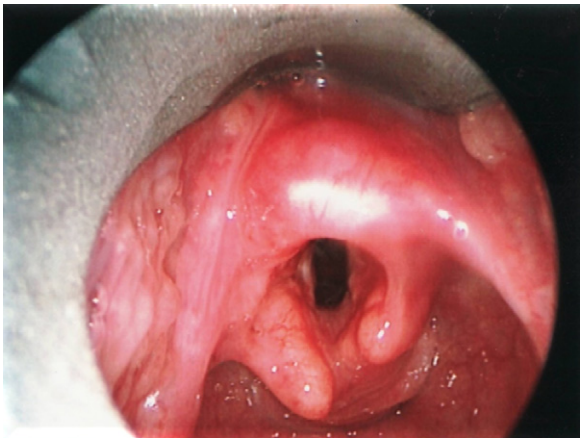

Fig. 2. Direct laryngoscopic view of the supraglottic structures 6 weeks after initial supraglottoplasty. The epiglottis is returned to an anterior location, no longer obstructing the glottis.

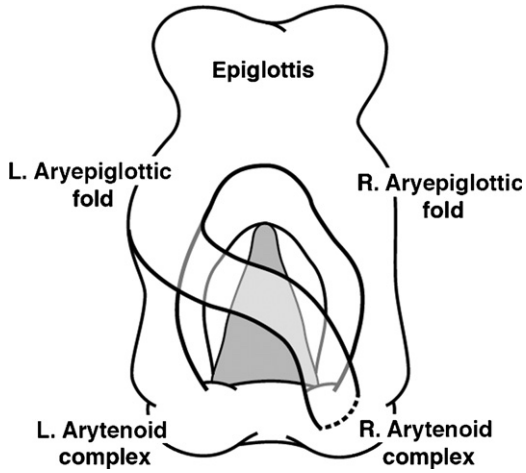

Fig. 3. A schematic drawing depicting the aberrant fusion of the left edge of the epiglottis fused to the right arytenoid complex.

Table 1
Laryngeal structures formed during the embryonic phase of development.

Embryonic phase of prenatal development: weeks 1–8	
Day	Structure formed
20	Foregut first identifiable with a ventral laryngotracheal groove into the endoderm and adjacent mesenchyme
22	Laryngotracheal groove differentiates into the primitive laryngeal sulcus and the respiratory primordium
24	Right and left lung buds appear
26	Lateral fusion of the laryngotracheal groove separates the trachea from the esophagus as the tracheoesophageal septum
30	Hypobranchial eminence gives rise to epiglottic and cuneiform cartilages
32	Primitive arytenoid swellings form the arytenoids, corniculate cartilages, and aryepiglottic folds
33	Superior laryngeal nerve emanates from fourth branchial arch
37	Recurrent laryngeal nerve derived from sixth branchial arch
40	Larynx, cartilages, and intrinsic muscles clearly evident
48	Epiglottis achieves its concave shape
55	Epithelial lamina dissolves leaving a patent trachea

modified CO_2 laser supraglottoplasty in addressing both the anterior and posterolateral obstruction. Similar surgical success rates have been observed in studies where approaches were based on the pattern of supraglottic collapse [1].

4. Conclusion

We conclude that consideration of the site of obstruction and degree of disease severity will dictate therapeutic decisions, regardless of the classification. Additionally, effective management of our patient's acquired subglottic stenosis with balloon catheter dilation and microdebrider further illustrates the surgeon's armamentarium in managing pediatric airway challenges.

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