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 laryngotracheal 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.

INTRODUCTION

•Laryngomalacia is the most common congenital malformation of the larynx and most common cause of stridor in neonates and infants1. Manifested by inspiratory high-pitched stridor associated with abnormally flaccid supraglottic laryngeal tissue.
•Diagnosis established with laryngoscopy.
•Majority of cases resolve between the ages of 2 and 5 years1.
•Surgical intervention may be indicated for more severe disease associated with obstructive sleep apnea, failure to thrive, severe reflux, cor pulmonale, or lack of anticipated spontaneous resolution2.

CASE REPORT

•13-month-old girl with a tracheostomy referred to our outpatient pediatric otolaryngology clinic for evaluation of her airway.
•Premature birth at 24 weeks of gestation complicated by bronchopulmonary dysplasia and reactive airway disease.
•Management in the neonatal intensive care unit included endotracheal intubation for two weeks followed by successful extubation prior to being discharged home at one month of age.
•Beginning at two months of age, multiple incidences of self-resolving stridor, which culminated in an episode of respiratory decompensation managed with emergent tracheostomy at age six months.
•Close inspection under direct laryngoscopy revealed an abnormality of the epiglottis and aryepiglottic (AE) folds.
•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 (Figure 1).
•This also resulted in an asymmetry with shortened AE folds on the right side.
•Acquired subglottic stenosis with subglottic narrowing and suprastomal granulation tissue without tracheomalacia was also noted.
•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 (Figure 2 & 3).
•During repeat procedure six weeks later, the CO2 laser set to 3 watts was this time used to trim down redundant epiglottic tissue (Figure 4).
•Successful decannulation took place shortly afterwards with no subsequent stridor or respiratory difficulty at eight months follow-up.

DISCUSSION

•Proposed classification schemes reflect the most frequently encountered sites of suprastomal collapse:
•Features on our patients intraoperative and awake fiberotic laryngoscopic examinations most consistent with a Holinger type A and C, Lee group III, or Kay type 1 and type 23,4.
•Unilateral fusion of the epiglottis to the right arytenoid/AE fold complex is not accounted for any of the known classification systems.
•Supraglottic dysgenesis likely took place during days 30 through 32 of gestation, where the hypobranchial eminence and primitive arytenoid swellings gave rise to the epiglottis and aryepiglottic folds, respectively5.
•Surgical techniques for laryngomalacia generally involve supraglottoplasty for posterior and lateral glottis, and epiglottopexy with or without supraglottoplasty for obstruction anteriorly6.
•Despite the unique presentation as a supraglottic dysgenesis, we were pleased with the positive response to modified CO2 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 collapse1.
•We conclude that a correlation between the site of obstruction and degree of disease severity will dictate therapeutic decisions, regardless of the classification.

REFERENCES