Controlled CHAOS: Undiagnosed Congenital High Airway Obstruction Syndrome due to a Laryngeal Cyst

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INTRODUCTION

There are few scenarios more gut-wrenching than an unanticipated difficult airway. While there are many case reports of pediatric difficult airways, there are scant reports of neonatal difficult airways. In both the literature and the difficult airway algorithm, a major junction is the ability or inability to ventilate.

CASE REPORT

- Patient IS was born via NSVD at 39weeks gestational age to a 29 year-old G1P0 mother with gestational diabetes.
- Third trimester ultrasound was unremarkable and showed no evidence of polyhydramnios.
- Immediately after birth, the patient noted to be in severe respiratory distress.
- Intubation was attempted unsuccessfully by multiple providers, revealing a grade 4 view with presence of a posterior oropharyngeal mass.
- Positive pressure ventilation was however possible.
- Venous blood gas revealed profound mixed metabolic and respiratory acidosis.
- The patient was taken emergently to the operating room for tracheostomy.
- Hypothermic protocol was initiated based on concern for hypoxia during events following delivery and the patient was transported to Ronald Reagan UCLA for higher level of care.
- The patient was brought to the OR on day 6 of life for direct laryngoscopy (DL), bronchoscopy, and mass excision.

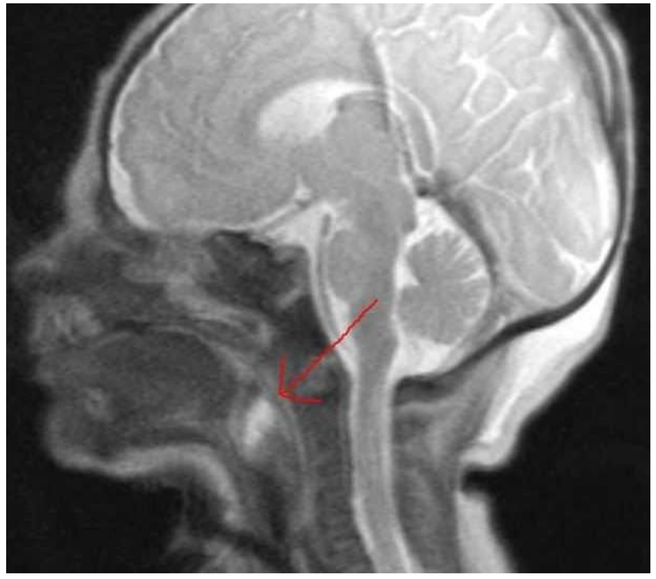

Figure 1: Pre-excision MRI: Note the near-total obstruction of the airway by the mass. Note the in situ tracheostomy tube.

- Initial DL by the surgeon revealed a large non-vascular appearing mass fully obstructing the airway (fig 2a). The mass was lateralized and epiglottis visualized. (fig 2b & 2c).
- Cyst excision resulted in extravasation of clear fluid, and the mass was partially excised. Subsequent DL revealed a grade 1 view (fig. 2d).
- Pathology was consistent with a saccular cyst. He was ultimately decannulated. Follow-up MRI brain was negative for hypoxic ischemic encephalopathy (HIE).

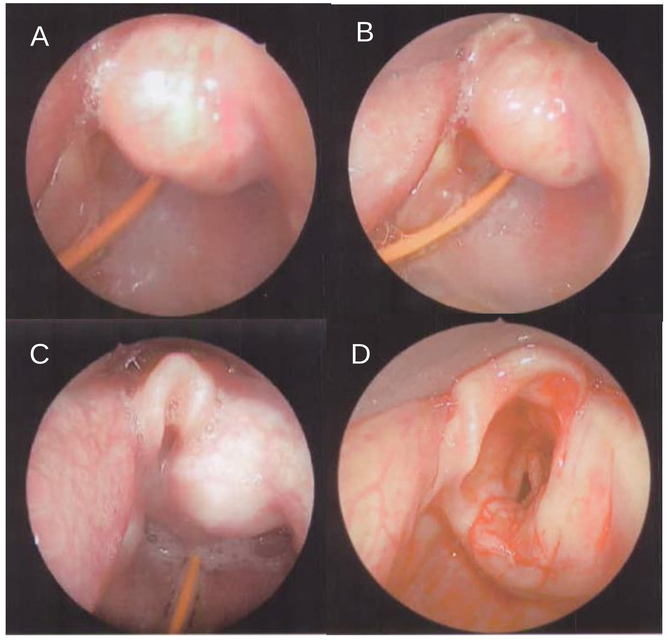

Figure 2: Direct laryngoscopy images: Initial DL (a). Note bubbling left of the mass and nasogastric tube visible posteriorly. With manipulation the view was improved (b and c). Laryngoscopy post-excision (d).

DISCUSSION

Vocal fold paralysis and laryngomalacia account for the majority of congenital laryngeal malformations. Less common lesions include congenital subglottic stenosis, subglottic hemangioma, laryngeal atresia, and cysts. A brief review of congenital airway malformations can be found in Table 1. As seen in this case, laryngeal cysts can cause marked upper airway obstruction, particularly in neonates. Saccular cysts, as seen in our patient, are fluid-filled submucosal cysts covered in mucous membrane and can cause near total airway obstruction. Conversely, laryngoceles are air-filled cysts that communicate with the laryngeal cavity and are less likely to cause complete airway obstruction.

SUMMARY

In our case, maintenance of spontaneous ventilation with assisted positive pressure ventilation was lifesaving. If ventilation had not been possible, the outcome could have been devastating. In hindsight proceeding more quickly to tracheostomy may have prevented the progressive acidosis and concern for HIE that developed. Ultimately, in cases such as this, the best means of avoiding disaster remains the availability of equipment and staff to perform a tracheostomy.

Table 1. Congenital airway malformations

Malformation	% of All Cases	Etiology and Clinical Notes
Laryngomalacia	60%	Caused by collapse of the supraglottic structures during inspiration. Presents with inspiratory stridor worse with supine position, feeding and agitation
Vocal Cord Paralysis	15-20%	Multiple etiologies. Bilateral paralysis presents with inspiratory stridor with progressive airway obstruction. Unilateral paralysis presents with hoarse cry or feeding difficulty or may go unnoticed
Congenital Subglottic Stenosis	15%	Membranous type is caused by circumferential submucosal hypertrophy. Cartilaginous type results from abnormal shape of the cricoid cartilage. Often presents with biphasic stridor with or without respiratory distress.
Subglottic Hemangioma	1.5%	Caused by a vascular malformation. Usually asymptomatic until age 2-12 months, however can grow rapidly resulting in progressive respiratory distress. Can be associated with PHACES syndrome.
Laryngeal Atresia	< 1%	Caused by failure of epithelial growth and canalization in the vestibule and subglottic regions. Acute airway obstruction after cord clamping. Without concurrent tracheoesophageal fistula, tracheostomy is required.
Laryngeal Cyst	< 1%	Obstruction of the laryngeal saccule in the ventricle leads to mucus retention and formation of saccular cysts. Laryngoceles are air-filled outpouchings of saccular mucosa. Mostly asymptomatic when diagnosed in adulthood but can cause airway obstruction in pediatric patients (especially neonates).

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