Postoperative Airway Emergency In A Patient With An Unknown Tracheal Cartilaginous Sleeve

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Introduction: Tracheal cartilaginous sleeve (TCS) is a rare airway malformation where a continuous tracheal cartilaginous structure envelops the airway. Tracheal rings are not formed and the cartilage is vertically fused in a C- or O-shaped manner[1]. At least 20% of patients presenting with craniosynostosis have concomitant syndromic conditions may not be known to the anesthesiologist and/or surgeon[2]. We describe a case of an unknown tracheal cartilaginous sleeve in a pediatric patient requiring emergent tracheostomy postoperatively.

Case Presentation: A 12 month old, 10-kg male presented for elective craniosynostosis repair due to trigoncephaly. He was born premature at 28+6 weeks by primary cesarean section due to twinning and breech presentation and required NCPAP for 37 days in NICU. He had a history of chronic sinusitis and asthma. For the surgical procedure a general anesthetic was performed. A grade 3 airway was viewed and ETT was placed after two atraumatic attempts. Operation was completed in four hours and was uneventful. The patient had an estimated blood loss of 100 mL and received 80 mL in packed red blood cells and 440 mL of lactated ringer's solution. The patient was given 0.5 mg/kg dexamethasone and 65 mcg of fentanyl total for the procedure. The patient was extubated and then transported to the PICU at 1530.

Morphine dose was increased in PICU from 0.1 to 0.2 mg/kg/dose Q2 due to increased pain. The patient also received Tylenol and a 20cc/kg bolus of fluid. 12 hours postoperatively the patient developed inspiratory stridor and ultimately respiratory distress despite racemic epinephrine and naloxone being given. Patient desaturated and developed bradycardia despite bag mask ventilation. CPR was started and bradycardia resolved in 5min. Anesthesia arrived and a glidescope was used to perform intubation and showed extensive supraglottic edema and no visualization of the glottis. After 9 unsuccessful intubation attempts, ENT performed an emergent bedside tracheostomy successfully (3.5 uncuffed Bivona).

The following day a direct laryngoscopy/ bronchoscopy was performed by ENT and findings included a Grade III view and a tight omega shape epiglottis, subglottic persistent swelling and complete dynamic collapse was reported. Trachea with cartilaginous sleeve was diagnosed. In addition, ENT was unable to pass flexible scope retrograde to subglottis from tracheostomy site due to severe swelling (Fig. 1).

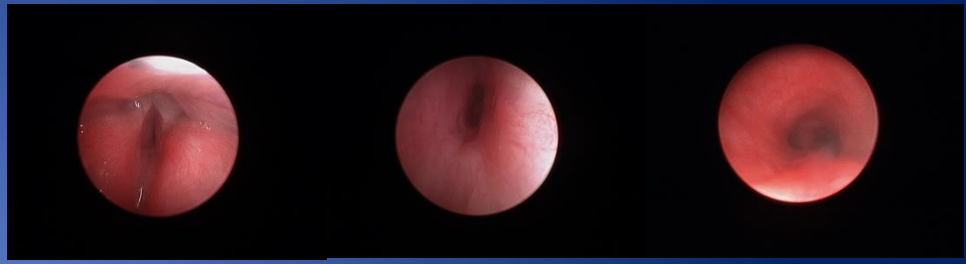

Fig. 1: From L to R, Views of edematous vocal cords, subglottis, and trachea without cartilaginous rings.

Discussion: Syndromic craniosynostosis has a strong correlation with TCS in over 150 described syndromes[2]. The most common syndromes associated with craniosynostosis and TCS are Crouzon, Apert, and Pfeiffer. The presence of TCS has been associated with premature death in almost all cases with a mean age of death of 3 years[1]. Davis et al proposed that the tracheal dispensability is altered leading to turbulent air flow and obstruction[2]. These changes are thought to lead to impairment in clearing secretions and increased number of respiratory infections[3]. There is a described benefit to early tracheostomy which bypasses the upper airway that can become obstructed in patients with CTS. Additionally, a survival advantage has been described in children with TCS that have an elective tracheostomy[1].

Conclusion: Patients presenting for craniosynostosis repair with a history of frequent respiratory infections, post-partum stridor and airway reactivity, could have an undiagnosed syndrome associated with tracheal cartilaginous sleeve especially in the setting of other syndromic features. Early ENT evaluation could be warranted.

References:

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