Failed Awake Intubation in Fibrodysplasia Ossificans Progressiva
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Background

- Fibrodysplasia ossificans progressiva (FOP) is an extremely rare disease resulting in ossification of skeletal muscle, fascia, ligaments and tendons.
- FOP involves a pathologic response to tissue trauma such as venipuncture or intramuscular injection.
- Largest series on anesthetic management described 30 patients in whom no significant airway difficulties or postoperative complications were encountered, concluding that general anesthesia can be safely performed.1
- Here we describe airway management difficulties that may be seen in the disease.

Patient

- 19-year-old male presented for dental extractions. He weighed 34 kilograms and was 4 feet 6 inches tall.
- Bed bound and room air oxygen saturation was 88%. Used supplemental oxygen while sleeping. Hypertension, asthma, scoliosis, and obstructive sleep apnea requiring CPAP while sleeping.
- Kyphotic cervical spine and the left humeral head approximated the left mandibular angle. Maximal interincisor distance was 7mm, and the mouth could close 2mm. Chest wall ossification with obvious thoracic insufficiency.
- Normal cognition and cooperative.

Anesthetic Management

- Multiple cotton swabs were dipped in 4% cocaine and left in both nasal passages. Two lubricated nasal trumpets were serially placed.
- Video bronchoscope loaded with a lubricated, warmed endotracheal tube was advanced through the nose, and a small amount of blood was visualized in the nasopharynx.
- It was difficult to manipulate the bronchoscope for suctioning or advancement inferiorly. A 10 French suction catheter was helped little.
- Very weak coughing due to blood began. He eventually fatigued.
- Oxygen saturation decreased and mask ventilation was initiated. Ventilation was particularly difficult but accomplished with a two-handed, two-person technique.
- Case was cancelled since, if nasal intubation were successful with further attempts, thoracic insufficiency would likely lead to severe post-extubation pulmonary complications due to additional bleeding from the dental extractions.

Discussion

- Advanced FOP such that a laryngeal mask and oral flexible bronchoscopic intubation were not possibilities.
- Santoro’s account also describes mask ventilation was extremely difficult due to both upper airway anatomy distortion and restrictive pulmonary physiology.2
- Tracheostomy would be almost impossible as the mandible mostly covers the trachea.
- This case highlights three fundamental challenges in airway management: severe restrictive lung disease, a fixed mandible due to masseter ossification (causing airway obstruction and preventing use of an oral airway), and lack of access to the trachea.

References