Anesthetic and Airway Management in a Child with Goldenhar Syndrome

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Introduction: Goldenhar (facioauriculovertebral syndrome, oculoauriculovertebral syndrome) syndrome was first described by Dr. Goldenhar in 1952. The major features that characterized this newly recognized entity were preauricular appendages, fistulas and epibulbar dermoids (1). Since then, the clinical presentation has evolved to include vertebral abnormalities, upper eyelid colobomas, subconjunctival lipomas, ear anomalies, hearing loss, unilateral facial hypoplasia, micrognathia, cleft or high-arched palate, congenital heart disease, and renal anomalies (2). The etiology of this syndrome remains unclear, however it has been suggested that it may result from a vascular accident occurring during fetal development, leading to defects of the first and second branchial arches (3). We describe the anesthetic management of a child with Goldenhar syndrome scheduled for elective surgery.

Case Report

The patient was a 21-month-old (weight 14.5 kg) with diagnosed Goldenhar syndrome and scheduled for dental restoration and extractions. The preoperative assessment revealed right-sided facial hypoplasia and microtia, deviation of the mandible to the affected side, micrognathia, and high-arched palate. No vertebral anomalies were noted.

We discussed our concerns and plan of airway management with the child's parent. In view of the anticipated difficult airway, we planned an awake fiberoptic intubation with intravenous sedation.

Premedication was with midazolam 0.5 mg/kg orally. On arrival into the operating room, pulse oximetry, ECG, and noninvasive blood pressure monitoring were placed. An intravenous catheter was inserted, and atropine (0.01 mg/kg) was administered. A remifentanil (0.1 mcg/kg/min) infusion was begun. Topical anesthesia with phenylephrine was instilled into both nostrils. The patient was maintained spontaneously breathing. Subsequently, a nasal fiberoptic intubation was attempted. However, two anesthesiologists were unable to advance a 3.7 mm fiberscope (Karl Storz Endoscopy, Culver City, CA) through either nare. Oral fiberoptic intubation was also unsuccessful. The vocal cords were visualized, but difficulty was encountered with attempts to advance the fiberscope beyond thickened supraglottic structures. On direct laryngoscopy only the epiglottis was visualized (Cormack and Lehane grade 3). A blind oral intubation was performed on the second attempt following a change of laryngoscope blade and endotracheal tube size. The patient's airway was secured with an uncuffed 3.5 mm ID tube. Anesthesia was maintained with 0.5% endtidal concentration of isoflurane, 60% nitrous oxide in oxygen and a remifentanil infusion. The dental surgeon noted a communication between the gingiva and nasal cavity, for which an otorhinolaryngologist was consulted.

The operation lasted for 55 minutes, and extubation and postoperative recovery of the patient were uneventful.

Discussion: Goldenhar syndrome is a rare multisystem syndrome with a wide spectrum of clinical features (1,2). The anesthetic management may be complicated by a difficult airway because of micrognathia or vertebral anomalies. Although the LMA has been described for use in Goldenhar syndrome (4), its use here was not appropriate for dental surgery. However, it was available to us as a rescue airway device. Of primary importance are a thorough preoperative evaluation and preparation for the anesthetic management of patients with Goldenhar syndrome.

References

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