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SCIENTIFIC ARTICLE - CASE REPORT

MANAGEMENT OF A DIFFICULT AIRWAY IN A CHILD WITH GOLDENHAR SYNDROME

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ABSTRACT

Goldenhar syndrome (GS) is part of the oculoauriculovertebral spectrum (OAVS) characterized by the classic triad of ocular, auricular, and vertebral abnormalities. A 6-month-old, 8.5 kg patient with GS underwent general anesthesia for removal of a cerebral teratoma. Pre-anesthetic evaluation revealed retrognathia, asymmetrical hard palate, and facial asymmetry. Anesthetic induction was by inhalation with 8% sevoflurane combined with 10 mcg of fentanyl, 30 mg of intravenous propofol, and 1.5 mg of cisatracurium. Three attempts at orotracheal intubation were made, the first two via direct laryngoscopy, without glottic visualization, and the third using a videolaryngoscope. In the literature, few cases of successful airway management in GS using a Magill blade, blind intubation, and laryngeal mask airway have been mentioned. Thus, the objective of this article is to report on the management of a difficult airway in a patient with SG.

Keywords: Goldenhar syndrome, General anesthesia, Airway management, Brain neoplasms, balanced anesthesia.

INTRODUCTION

GS is part of the OAVS and is recognized by the presence of the classic triad of ocular, auricular, and vertebral anomalies. It is mainly characterized by preauricular appendages, epibulbar dermoid cysts, and hemifacial microsomia, in addition to cardiac, genital, renal, and pulmonary abnormalities.[1]

It has a rare incidence of approximately 1 in 56,000 births, with a higher prevalence in males (3:2)[1]. Its origin is multifactorial and includes genetic and environmental factors, with the presence of familial cases possibly explaining the genetic basis of the syndrome[2]. Its pathophysiology involves abnormal development of the first pharyngeal arches, which develop during the fourth week of gestation.[3]

Diagnosis is based on clinical data, including medical history, physical examination, and results of complementary tests, and may be established either during pregnancy or after birth. Regarding treatment, it is carried out within each specialty, aiming to provide the most appropriate management based on the literature and the experience of the professionals involved.[4]

In this context, patients with GS, due to the presence of facial malformations in their phenotype, are potentially at risk for a difficult airway. Therefore, this article aims to describe the management

of a difficult airway in a patient with GS undergoing general anesthesia.

CASE REPORT

A six-month-old male patient, weighing 8.5 kg, diagnosed with GS, underwent general anesthesia for resection of a cerebral teratoma.

During the pre-anesthetic evaluation, the patient presented with retrognathia, posterior displacement of the soft palate, asymmetric hard palate, congenital torticollis, microtia, and facial and mandibular asymmetry. Additionally, he had a recent nasofibrolaryngoscopy showing an incomplete and open midline cleft palate and mild glossoptosis. Following this examination, the patient developed apnea during feeding.

Other complementary tests revealed mild right ventricular dilation and hypertrophy on echocardiography, and a normal cervical spine on radiography.

Cranial magnetic resonance imaging demonstrated an expansive cystic lesion located in the suprasellar cistern, with calcifications measuring approximately 2.4 × 3.3 × 2.3 cm, suggestive of a cerebral teratoma. This lesion caused a compressive mass effect on the pontine base, right cerebral hemisphere, and fourth ventricle.

Anesthetic induction was performed using inhalational sevoflurane (8%), associated with intravenous administration of fentanyl 10 micrograms (mcg) and propofol 30 mg. After induction, the patient tolerated mask ventilation without difficulty. Two attempts at orotracheal intubation (OTI) using direct laryngoscopy (conventional laryngoscope) were performed, without visualization of the glottis, even with external neck pressure and use of a bougie.

A new dose of 30 mg propofol and 1.5 mg cisatracurium was administered, and a third laryngoscopy attempt was performed using a videolaryngoscope and bougie, revealing a Cormack-Lehane grade 3B view, with successful OTI using a cuffed endotracheal tube size 4.5 (Figure 1). Anesthesia was then maintained with sevoflurane 2%, without the need for additional intravenous anesthetics or further neuromuscular blockade. Adjunct medications were also administered, including dipyrone 250 mg, dexamethasone 1.5 mg, and ondansetron 1.5 mg.

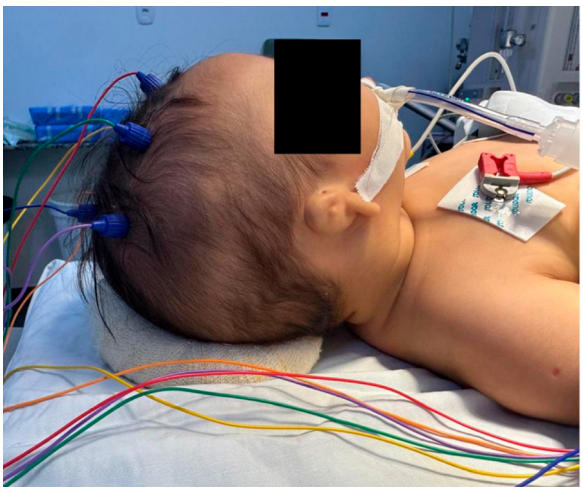

Figure 1: Patient from the clinical case under general anesthesia.

During the surgical procedure, the patient remained hemodynamically stable and eupneic, under mechanical ventilation. At the end, the patient was extubated without complications and transferred to the Intensive Care Unit (ICU)

DISCUSSION

The craniofacial abnormalities in GS may result in a difficult airway in up to 40% of cases, mainly due to the combination of micrognathia, limited cervical mobility, and tracheal deviation. This represents a challenge for the anesthesiologist and may require the use of alternative methods for laryngoscopy in order to ensure airway patency.[5,6]

Clinical airway examination should include the assessment and identification of structural malformations. Therefore, features such as palate and mandibular shape, mouth opening, and range of neck movement should be evaluated.[6]

Regarding complementary evaluation, mandibular and cranio-cervical radiological assessment is recommended prior to scheduled surgical procedures. The presence of mandibular hypoplasia identified on imaging, for example, may be considered an individual predictor of a difficult airway, as studies indicate a significant association between the severity of mandibular hypoplasia and the degree of intubation difficulty.[7]

Obstruction of the upper and lower airways during sleep is common in patients with significant craniofacial anomalies, such as those with GS. This finding is important, as it may predict difficult mask ventilation, which was not observed in the patient described, since he tolerated mask ventilation without difficulty after anesthetic induction.

Anticipation of a difficult airway is of paramount importance in patients with GS. In the literature, only a few cases of successful airway management in GS have been reported using a Magill blade, fiberoptic intubation, blind intubation, and laryngeal mask airway. In such situations, it is preferable to use a videolaryngoscope rather than more traditional techniques from the outset.[8] Thus, a point of criticism in the management of this case is that the videolaryngoscope was not used as the first attempt, but rather the conventional laryngoscope.

Regarding pharmacological effects, the combination of volatile anesthetics, muscle relaxants, and opioids places patients with GS at high risk of postoperative hypoxic events. Therefore, complete recovery of airway reflexes should be confirmed before extubation, as well as postoperative monitoring in an intensive care unit setting.[9]

CONCLUSION

Patients with GS represent a challenge in airway management and in establishing a definitive airway for general anesthesia. Craniofacial anatomical abnormalities may predispose these individuals to a difficult airway, both for mask ventilation and for orotracheal intubation. In this context, it is essential that the anesthesiologist and the team are well trained, and the use of videolaryngoscopy should be considered from the first attempt.

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EDITORIAL AND REVIEW

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