CASE REPORT

Successful management of difficult airway in an adult patient of Goldenhar syndrome

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ABSTRACT

The present case emphasizes on the increasing difficulty in management of patients with Goldenhar syndrome as they age. Fiberoptic intubation using high dose dexmedetomidine along with local anesthetics is a safe technique for securing the airway in a difficult situation, even if the patient dose not cooperates for awake procedure. A16-year-old Goldenhar patient with multiple facial deformities was scheduled for correction of hollowness of right cheek and was successfully managed with high dose dexmedetomidine infusion along with local anesthetics to facilitate fiberoptic intubation with no untoward side-effects.

Key words: *Dexmedetomidine, difficult airway, fiberoptic intubation, Goldenhar syndrome*

INTRODUCTION

Goldenhar syndrome is a rare congenital hemicraniofacial dysmorphology, which results from developmental defects of the first and second branchial arches.[1] Airway management is challenging due to the presence of various facial and neck deformities. A number of pediatric cases of Goldenhar syndrome have been described in the literature.[2] The characteristic feature of this syndrome is that airway abnormalities worsen with age.[3] We report a case of 16-year-old patient with Goldenhar syndrome presenting for cosmetic correction of hollowness of right cheek, with multiple facial deformities, resulting in difficulties in securing the airway. Written informed consent was obtained from the patient's parents.

CASE REPORT

A 16-year-old, 60 kg, female patient presented to our plastic surgery department for cosmetic correction of hollowness of right cheek. Past history revealed the presence of hemifacial dysmorphism with speech and swallowing difficulty since birth with decreased neck movements as the child grew. As a child, the patient had undergone seven corrective surgeries elsewhere. During the last surgery, done 8 years back for correction of nasal deformity, patient was mechanically ventilated postoperatively for 48 h in view of aspiration of blood, leaving a young child traumatized mentally. Past medical records revealed the absence of any cardiac anomaly. The detail of techniques followed for securing the airway during previous surgeries was not mentioned in discharge summaries.

Physical examination revealed several facial deformities, prominently on the right side of face with unilateral mandibular hypoplasia. Mandibular protrusion was restricted, micrognathia was significant with an inter-incisor gap of about 3 cm. Unilateral ankylogossia present on the right side prevented protrusion of the bulky tongue, attached to the floor of the mouth with a fleshy septum giving rise to speech and swallowing difficulty [Figure 1]. The neck was short with a fixed flexion deformity restricting neck movement [Figure 2]. Hematological and biochemical investigations were unremarkable, along with normal 12-lead electrocardiogram.

We anticipated difficulties in securing airway and intubation through the nasal route, retrograde intubation and surgical route were all contraindicated. Despite the presence of a difficult airway, awake procedures were not possible as our patient refused to cooperate.

An 8 h of fasting were advised and patient received tablet ranitidine 50 mg and tablet metoclopromaide 10 mg 1 h

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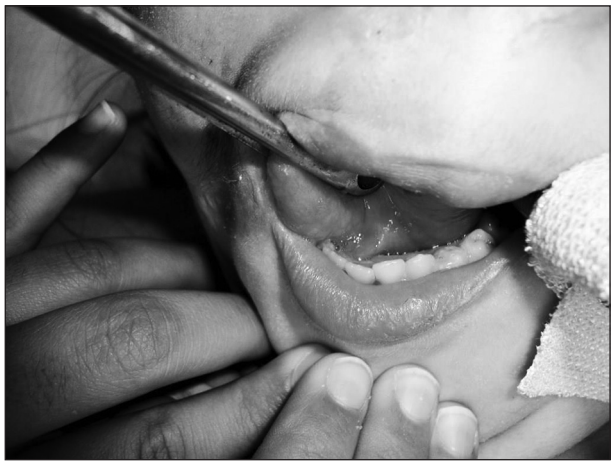

Figure 1: Fleshy septum joining the tongue to the base of mouth

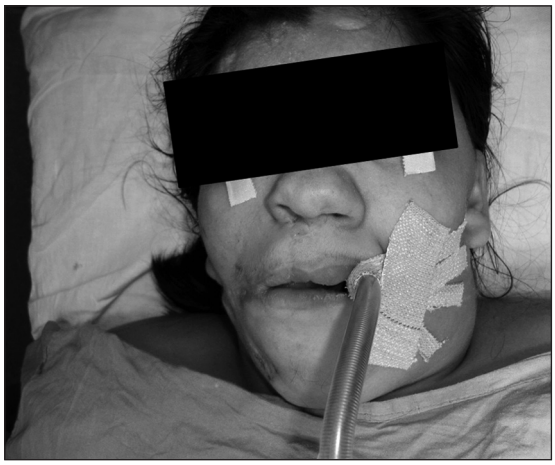

Figure 2: Facial deformities along with fixed flexion neck deformity

before shifting to the operating room. She was nebulized with 4% lignocaine, preoperatively, for 20 min. Inside the operating room; monitoring was established as per American Standards Association standards. Injection glycopyrrolate 0.2 mg was administered after securing intravenous access. Lignocaine spray (10%) was administered over the posterior pharyngeal wall, but glossopharyngeal nerve, superior laryngeal nerve, and transtracheal blocks could not be given in view of unidentifiable anatomical landmarks. Patient was given supplemental oxygen and 1 mcg/kg of injection dexmedetomidine was administered intravenously over 10 min, followed by infusion at the rate of 0.2 mcg/kg/min. Although patient had effective spontaneous ventilation, she did not allow significant airway manipulation at this dose. Thus, we gradually increased the dose of dexmedetomidine infusion up to 3 mcg/kg/h. After the patient was adequately sedated, oral fiberoptic bronchoscopy was done, 2 ml of 2% lignocaine was sprayed over the glottic opening and the patient was successfully intubated with a 7.0 mm flexometallic endotracheal tube. After securing the airway, dexmedetomidine infusion was stopped and the patient was given 100 mcg of injection fentanyl, 80 mg of injection propofol and neuromuscular blockade with intravenous atracurium 30 mg. Anesthesia was maintained with nitrous oxide (66%) and oxygen (33%) along with propofol infusion. The intraoperative course was uneventful. At the end of surgical procedure, which lasted for 1.5 h, neuromuscular blockade was reversed and patient's trachea was successfully extubated.

DISCUSSION

Goldenhar syndrome was first described by Dr. Maurice Goldenhar in 1952.[1] Airway difficulty is a known entity and difficult intubation has been reported to be as high as 39.5%.[2] Cases of airway management in children with Goldenhar syndrome have been reported earlier, but with every passing year in the child's life, airway management and administration of a general anesthetic becomes increasingly difficult. The degree of laryngoscopic difficulty increases with increasing age.[3]

In about 40% of patients with Goldenhar syndrome, there are associated vertebral anomalies in the form of Klippel-Feil anomaly and there is an added problem of a short immobile neck.[4] Fixed flexion deformity of the neck in our patient further added to the difficulty.

There are very few cases of adults with Goldenhar syndrome reported in the past. Use of airtraq optical laryngoscope has been described in a 23-year-old female Goldenhar patient presenting for tympanoplasty.[5] However, this was done as an awake procedure with patient's cooperation. Use of awake oral fiberoptic intubation has also been described in a woman with Goldenhar syndrome undergoing caesarian section.[6] Even failed intubation has been reported in a 24-year-old Nigerian female patient who presented for excision of left eye epibulbar dermoid. Surgery in this patient was done after laryngeal mask airway placement.[7] We performed successful fiberoptic intubation under high dose dexmedetomidine sedation, along with the use of local anesthetics, without any untoward side effects.

Use of dexmedetomidine has been described in monitored anesthesia care, fiberoptic bronchoscopy, dental procedures, cardiac catheterization, ophthalmic surgeries, head and neck surgeries, in the pediatric population as well in neonates.[8] In our patient, dexmedetomidine provided adequate sedation, stable hemodynamics, along with maintenance of spontaneous respiration with no episode of desaturation or requirement of assisted ventilation.

Standard dose dexmedetomidine prescribed is 1 mcg/kg over 10 min followed by infusion at the rate of 0.2-0.7 mcg/kg/min.[8] However, this dose remains ineffective wherever

airway manipulation is required. Use of high dose of dexmedetomidine has been reported even in the pediatric population.[9] In our case also, we gradually increased the infusion rate to 3 mcg/kg/h to facilitate fiberoptic intubation. In a study done using target controlled infusion of dexmedetomidine, a high dose requirement has been reported as compared to the dose used for sedation in intensive care unit. Dexedetomodine level was escalated slowly until patients developed tolerance to laryngoscopy and condition for laryngoscopy was reported excellent in all cases.[10] Higher dose of dexmedetomidine did not affect hemodynamics, probably because of high stimulation during airway manipulation in an anxious patient.

Thus, by using a high dose of dexmedetomidine infusion, along with local anesthetics, we could successfully secure the airway, fiberoptically, of a 16-year-old patient of Goldenhar syndrome with fixed flexion deformity of the neck, where bag and mask ventilation, surgical airway and retrograde intubation were not possible. Further, fiberoptic intubation was also challenging as our patient refused to cooperate, had unidentifiable anatomical landmarks for blocks, difficulty in jaw protrusion and contraindication to the use of nasal route for fiberoptic intubation.

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