



Anesthesiology

GOLDENHAR SYNDROME-NO MORE A CHALLENGE TO ANAESTHESIOLOGIST

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ABSTRACT A patient, known case of Goldenhar syndrome came for right external ear reconstruction. Patient had typical features of the syndrome with hypoplastic mandible and maxillae, malocclusion of teeth, malaligned teeth, retrognathia and hypoplastic hyoid bone. Anticipating a difficult intubation it was decided to go ahead with awake intubation in this patient. Blind nasal intubation was planned because of unavailability of fiberoptic in our institution and it was achieved with 7 mm ivory white north pole tube with dexmedetomidine infusion at rate of 0.5mcg/kg/hr and topical anaesthesia.

KEYWORDS :

INTRODUCTION

Goldenhar syndrome or oculo – auriculo – vertebral syndrome is a rare congenital defect associated with anomalous development of first and second branchial arch. Abnormal embryonic vascular supply, disrupted mesodermal migration or some other factor leads to defective formation of the branchial and vertebral systems[2,3]. Ingestion of drugs such as thalidomide, retinoic acid, tamoxifen, and cocaine by the pregnant mother may be related to the development of this syndrome. Maternal diabetes, rubella, and influenza have also been suggested as etiologic factors[4]. Autosomal dominant, autosomal recessive, and multifactorial modes of inheritance have also been suggested[2]. It is characterized by incomplete development of ear, nose, lip, soft palate and mandible usually on one side of body. Some patients may also have developmental defects in internal organs specially heart, lungs and kidneys. Though this defect is characterized by unilateral presentation, 10% of the patients can have bilateral presentation too. Patients of goldenhar syndrome have to undergo multiple settings of surgical procedures for correction and reconstruction of their bodily defects which poses a great challenge to the anaesthesiologist. Proper airway maintenance is one of the concerns during anesthesia in such cases. Therefore, the anaesthesiologist should have several resources available in order to guarantee opened airways. The difficult airway algorithm of the American Society of Anesthesiologists (ASA) recommends the use of several devices, such as: laryngeal mask, ML-Fastrack®, retrograde intubation, lighted guide-wire and fiberoptic endoscopy in such cases of difficult airway [5]. Here we are reporting a similar case of goldenhar syndrome who was posted for right ear reconstruction with excision of preauricular skin tags bilaterally. CASE REPORT 23 years male patient Weighing 36 kg, classified under ASA 1 of American society of anaesthesia, K/C/O Goldenhar syndrome came for right ear reconstruction and removal of preauricular skin tags bilaterally. Our patient had typical features of the syndrome which included

1. Maxillary hypoplasia of right side
2. Mandibular hypoplasia
3. Hypoplastic Temporo mandibular joints
4. Preauricular skin tags bilaterally
5. High arched palate
6. Palatal teeth along with Malocclusion
7. Absent right external auditory meatus with right conductive hearing loss
8. Hyoid bone hypoplasia

Maxillary hypoplasia



Figure 1

Buck teeth

Preauricular skin tags

Mandibular hypoplasia



Figure 2

Preoperative assessment : A detailed history of patient focussing on any medical or surgical interventions in the past was taken. There was no significant medical or surgical past history. Airway assessment formed the core of preoperative examination of this patient. Patient had a mouth opening of 2 fingers, mallampati grading was MPC III. There was gross misalignment of teeth with some palatal teeth. Due to the defective development of mandible & hyoid thyrohyoid and thyromental distance could not be ascertained but they were shorter than normal. Temporo mandibular joint mobility was adequate. Nasal patency was checked by passing nasopharyngeal airway lubricated with 2% lignocaine. Nasopharyngeal airway no 7 could be passed easily through the patients right nares.

All laboratory investigations of the patients were within normal limits. An informed consent of the patient was taken for anaesthesia and patient was counselled for awake intubation. Nasal packing was done with 2% lignocaine 30 minutes before taking the patient on table and was given nebulisation of 2% lignocaine – adrenaline solution. 2-3 drops of Otrivin containing xylometazoline were put in the right nostril following the nebulisation. 0.2 mg of glycopyrrolate was given intramuscularly at the same time. Patient was wheeled inside the operation theatre. On table monitors were attached which included pulse oximeter, 3 leads Electrocardiogram, Non invasive blood pressure monitoring, End tidal CO₂. Oxygen was given through Hudson mask at 6L/m. Baseline readings were within normal limits. Anticipating a difficult airway, difficult airway trolley was already prepared which included laryngoscopes with blades of all types and sizes, endotracheal tubes and oral and nasopharyngeal airways of appropriate sizes, bougie, Airtraq, laryngeal mask airway of appropriate size and cricothyrotomy equipments. A suction device was kept ready. ENT surgeon was also kept standby. IV line was secured on left hand with 18G cannula. Patient was started on

intravenous dexmedetomidine infusion at rate of 0.5 mcg/kg/hr . Patient received intravenous midazolam 0.05mg/kg and fentanyl 2 mcg/kg. Airway was then anaesthetized with bilateral glossopharyngeal, superior laryngeal and transtracheal injections . Glossopharyngeal was blocked inferior and posterior to the mastoid while superior laryngeal was blocked at the paratracheal level because of hypoplasia of mandible and hyoid bone respectively . Two sequential puffs of 10% lignocaine were given at posterior pharyngeal wall .

Patient end of the no 7 ivory white north pole tube was well lubricated with 2% lignocaine jelly .Angle connector with the EtCO₂ sample line was attached to the machine end of the tube and the tube advanced slowly through the right nare keeping an eye on the capnogram and simultaneously listening for the breath sounds . Because of superiorly placed glottis tube insertion was difficult, so tube length was cut and tube rotation maneuvers were performed to facilitate its entry through glottis.

After confirming capnogram patient was given intravenous ketamine 1mg/kg and intravenous propofol 2mg/kg for induction of anaesthesia .5 point chest auscultation was done to confirm the position of tube and fixation was done .A check laryngoscopy was done to visualize the position of tube beyond glottis , but only tip of epiglottis could be seen, so check airtraq was done which confirmed the position of tube . Then Sevoflurane and nitous oxide were started . When patient got into deeper plane of anaesthesia intravenous rocuronium 1mg/kg was given . Patient was then maintained on O₂ + N₂O + Isoflurane and intravenous dexmedetomidine intraoperatively with intavenous vecuronium 1 mg top ups as and when required . Intraoperative period was uneventful . after completion of the surgery patient was allowed to come out of the anaesthetic effect completely. Dexmedetomidine was stopped and neuromuscular reversal done .When extubation criteria were fulfilled tube removed after thorough oropharyngeal suctioning. A smooth recovery with no complications: no nasal bleeding, no decrease in oxygen saturation, no sore throat, no voice changes were noted. I.V.paracetamol 1gm was given post-operatively

Post procedure patient was given oxygen by hudson mask and observed for 30 minutes for post anaesthetic effects. During shifting patients vitals were stable.

DISCUSSION

Patients with goldenhar syndrome pose great challenge to the anaesthetist .Facial deformities make mask ventilation difficult , reduced mouth opening , limited oropharyngeal space make direct laryngoscopy difficult. Whenever one comes across such a case a thorough examination of the patients airway should be done one day prior to the surgery . Nasal patency should also be checked so that tubes of adequate size can be kept ready at ones disposal . Fibreoptic is the gold standard in cases of difficult airway . However, since very few institutions have it , other components of a difficult airway trolley should be kept ready .

Whenever awake intubation is planned , patient has to be counselled properly as success of this procedure depends on the patients cooperation. Patient has to be explained the procedure in detail so that we can get maximum participation from him .

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