Anesthetic concerns and airway management of a 12-year-old child born with craniofacial cleft: A case report

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ABSTRACT

Children born with craniofacial anomalies are usually associated with syndromes such as Treacher Collins, Pierre Robin, Klippel–Feil, craniosynostosis, and its related disorders. These children present with difficult challenges for the anesthesiologist, one being the difficult mask ventilation and intubation. With the recent advances in modern craniofacial surgeries, these facial defects can be corrected to a large extent and thus, a person can lead a normal life. It is important to understand the development and characteristics of the more common anomalies and the challenges associated with them so that we can plan safe anesthesia for these patients. Here, we describe the successful anesthetic management of a 12-year-old child with left Tessier type 12 naso-orbital cleft with a cleft lip and high arched palate. The child was posted for corrective surgery of the facial cleft.

Key words: *Airway management, Anesthesia, Craniofacial surgery, Tessier N.12*

Craniofacial clefts are one of the rare anomalies present in the children associated with syndromes. These anomalies affect the behavioral, social, and psychological development of the children. Although the exact incidence is unclear, it is estimated between 1.4 and 4.9/100,000 live births. These clefts are mainly due to the maldevelopment at an embryonic stage [1-3]. These clefts can further be divided into predominantly cranial, predominantly facial, or a combination of both, i.e., craniofacial with the palpebral tissue as the reference line [4,5]. With the help of recent developments in surgical interventions, these anomalies can be repaired to a major extent.

Tessier, the father of craniofacial surgery, classified the facial clefts on the basis of the anatomical position of these clefts. These clefts were numbered from 0 to 14. The 15 clefts were put in four groups, based on their position – midline, paramedian, orbital, and lateral clefts [4]. Tessier suggested various surgical techniques which involve a multidisciplinary team approach involving specialties – plastic surgery, pediatric surgery, ophthalmology, orthodontics, otolaryngologist, and speech specialist.

The role of an anesthesiologist is to maintain perfect coordination among these specialties [4,6,7]. Proper pre-operative workup, discussion with surgeons, proper and stepwise planning of anesthesia for airway management, vigilant intraoperative monitoring, and adequate post-operative care and intensive care units (ICU) backup lead to the successful outcome of these surgeries [8]. The major concerns for an anesthetist are difficult airway management, hypothermia due to prolonged duration of surgery, and intraoperative blood loss, leading to major fluid shifts. Here, we will discuss the anesthetic management of craniofacial cleft with difficult airway associated with intracranial extensions which are rare so needs to be documented.

CASE REPORT

A 12-year-old male child, weighing 46 kg, in functional ASAII was posted for reconstructive surgery of facial cleft, left heminasal reconstruction with alar transposition, and glabellar flap with frontal bone defect reconstruction with split-rib graft (Fig. 1). The child had been diagnosed with Tessier 12 anomaly with a history of developmental delay, decreased vision left eye, and episodes of tonic-clonic seizures associated with trauma during early childhood. At present, the child was on syrup carbamazepine and seizures were controlled, the last episode was 2 years back.

On general examination, the child was conscious, oriented, following commands and vitals were stable. On local examination, a cleft lip with the high-arched palate and tongue tie was present, so modified Mallampati grading cannot be assessed. The left craniofacial cleft was present with the left deformed alae, displaced left eye and orbit, and hypertrophied inferior turbinate associated with the absent medial wall of the left orbit with soft-tissue enlargement. His systemic examination was normal.

All his blood investigations (hemoglobin – 12.5 g/dl, white blood cell count – 7500/mm^3, and platelets – 125,000/mm^3) were normal. Coagulation profile, including international normalized

Figure 1: A 12-year-old child with craniofacial cleft Tessier 12

ratio – 0.9, prothrombin time, and activated partial thromboplastin time, was normal. Serum electrolytes (sodium – 142 meq/L and potassium – 3.7 meq/L) were within normal limits. His non-contrast computed tomography brain revealed focal bony defect of the left frontal bone with herniation of the brain parenchyma (meningoencephalocele), gliosis, and medial orbital wall defect. His 2-D echocardiography was normal.

The child was fasted as per standard guidelines. Operation theater (OT) was warmed and prepared (difficult airway cart along with videolaryngoscope [C-MAC]) was kept ready. The patient was taken into OT, monitors were attached and a 20 G intravenous (IV) cannula was placed and the child was premedicated with IV fentanyl 90 ug. The facial cleft and herniated brain tissue were covered with saline-soaked sterile gauze and anesthesia was induced with IV thiopentone 225 mg. Due to difficult mask ventilation, after achieving adequate depth with mask ventilation (large-sized mask used) oxygen, air, and isoflurane, ProSeal laryngeal mask airway (LMA) was inserted for ventilation to avoid undue pressure on soft tissues and to prevent any rise in intracranial pressure (ICP).

After confirming adequate ventilation, IV vecuronium bromide was given and after 3 min of intermittent positive pressure ventilation, ProSeal LMA was removed and the trachea was intubated with C-MAC using cuffed right angle endotracheal tube of size 6. We preferred using C-MAC because of the tongue tie which could have made conventional laryngoscopy difficult. Laryngoscopy revealed a good view of vocal cords (Cormack-Lehane I). Oral packing was done. Anesthesia was maintained with air, oxygen, isoflurane, IV morphine 3 mg, IV vecuronium bromide, IV fentanyl 50 mcg, and IV paracetamol 675 mg. End-tidal carbon dioxide concentration ($etCO_2$) was kept within normal limits (30–40 mmHg). The total duration of surgery was 3 h. The child had an uneventful intraoperative course and the trachea was extubated when the patient was fully awake and following commands. The post-operative period was uneventful and the patient was shifted to ICU for close observation. The patient was discharged on the 5th post-operative day and was kept on 15 days follow-up by the plastic surgery team.

DISCUSSION

The etiology of the craniofacial cleft is multifactorial and includes maternal infections such as toxoplasmosis, alterations in phenylalanine, and intake of thalidomide drugs. Folic acid intake is known to lower the risk of a child being born with facial clefts [6]. These clefts are mainly due to the maldevelopment at an embryonic stage. Tessier N 12 is described as a rare anomaly of the bony tissue positioned between the nose and frontal bone. It is characterized by the medial orbital wall defect with soft-tissue enlargement, herniation of brain tissue, and displacement of eyes and orbit, as was observed in our case [1,6].

Challenges associated with such cases include airway management, prevention of rise in ICP, and to prevent venous air embolism due to the opening of various non-collapsible venous channels. A careful pre-operative assessment is, therefore, essential to prepare and execute a safe and proper anesthetic plan for the patient.

Airway management in these patients is difficult using conventional facemasks because of the large frontonasal cleft with displaced orbit, soft-tissue enlargement, and herniation of the brain tissue. Our patient had a large frontal cleft with an absent left medial wall of the orbit, displaced left orbit and eyeball, hypertrophy of inferior turbinates, soft-tissue enlargement, and meningoencephalocele. This could compromise the mask ventilation, so to deal with these problems; we used sterile saline gauzes to fill the defect, which avoided any undue pressure and injury, followed by the use of large size mask after induction of anesthesia for supporting ventilation. After attaining adequate depth, we inserted ProSeal LMA appropriate for weight size and maintained adequate ventilation until the onset of action of the muscle relaxant used. We used a videolaryngoscope to intubate the patient because of anticipated difficult intubation associated with these anomalies and also the presence of tongue tie would have made conventional laryngoscopy difficult. The difficult airway cart was checked and kept standby if required [9].

To prevent injury and any rise in ICP, large-sized mask and ProSeal LMA were used. Induction was done with IV thiopentone as it is neuroprotective in nature and $etCO_2$ monitoring was done. Air was preferred in place of nitrous oxide for the maintenance of anesthesia, as N_2O leads to significant changes in cerebral blood flow and cerebral oxygen demand, thus leading to a rise in ICP. The precautions undertaken were covering the defect with sterile-saline soaked gauzes, use of the large-sized masks or alternative use of supraglottic devices for mask ventilation, and the use of videolaryngoscope for successful intubation.

CONCLUSION

Thus, the major concerns in our surgery were difficult airway; ICP control, hypothermia, and blood loss were well dealt. Although airway management is difficult, a proper anesthetic plan leads to the successful administration of the general anesthesia and a successful on table extubation. ICP control was achieved by the use of neuroprotective agents, maintaining normal $etCO_2$, and

by avoiding pressure on herniated brain tissue. The use of warm fluids, replacing losses prevented major fluid shifts. Thus, with proper evaluation, coordinated plans, and vigilant monitoring, such cases can be successfully managed.

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