Noma Neonatorum: An Airway Challenge In Paediatrics

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Abstract

Noma Neonatorum (NN) is a devastating gangrenous disease that leads to severe tissue destruction in the face. Patients who survive Noma suffer from its sequelae- facial disfigurement, trismus and speech impediments which often necessitate extensive reconstructive surgery. These facial anatomic distortions present a challenge to attending anaesthesiologist. Reports on anaesthetic techniques for Noma sequelae are limited. We describe the anaesthetic management of a 13-month-old child with sequelae of NN. The child underwent reconstructive surgery of nose and upper lip with orotracheal intubation. We discuss the characteristic features of NN and review techniques of intubation in difficult paediatric airway. To the best of our knowledge, this is the first report of anaesthetic management of NN sequelae in paediatric age group in India.

INTRODUCTION

Cancrum oris (Noma) is a devastating gangrenous disease involving mucocutaneous junctions of oral, nasal and anal areas and occasionally, the eyelids and scrotum.[1, 2] Noma Neonatorum (NN) is a completely different disease than Noma and is commonly seen in pre-term, low birth weight neonates. It is rarely seen in full term neonates.[3 4.] The anatomic deformities of Noma sequelae in those surviving children present formidable challenges in perioperative airway control. We describe the anaesthetic management of a 13-month-old child with sequelae of NN. The child underwent reconstructive surgery of nose and upper lip with orotracheal intubation. We describe the features of NN and review techniques of intubation in difficult paediatric airway.

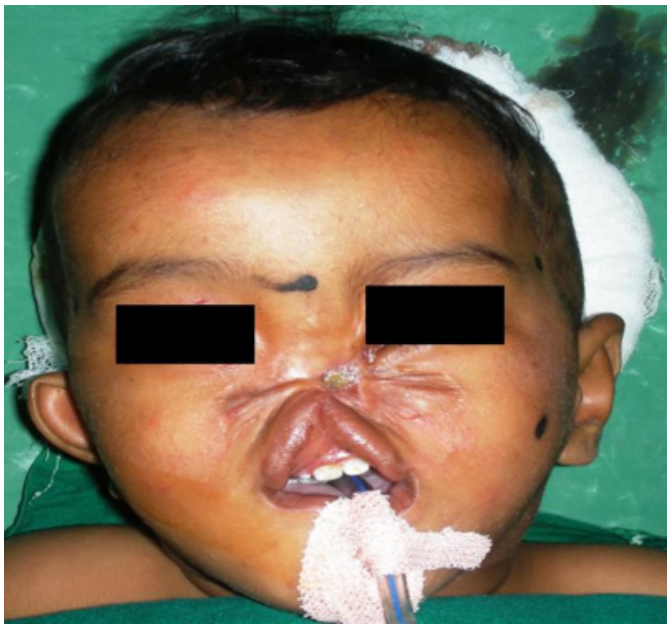
CASE REPORT

A 13-month-old male child, born preterm to a primigravida, with uneventful antenatal period had a past history of NN on the 10th day of life. The child was scheduled to undergo correction of the orofacial deformity by with reconstruction of upper lip and incomplete cleft alveolus.

On pre-operative evaluation, the child was poorly built and weighed only 8kg. His vital parameters were within normal limits. Airway examination revealed stenosed nostrils, hypoplastic maxillae, left incomplete cleft alveolus, deformed right ear lobe and adequate range of motion of the neck [Figure-1]. Systemic examination revealed no abnormalities. Laboratory investigations revealed haemoglobin=9.5g%, bleeding time=3min 30sec, clotting time=4min, platelet count=225 000 /mm^3, blood sugar=96mg/dl, blood urea=35mg/dl, serum creatinine=1.2mg/dl, serum sodium=138meq/L, serum potassium=4.2 meq/L and serum calcium=8.4 mg/dl. The chest x-ray was normal.

Figure 1

Figure 1



Standard fasting guidelines were followed. Under topical anaesthesia with Eutectic Mixture of Local Anaesthetics (EMLA) (Prilox, Neon) cream, a 24 gauge intravenous cannula was secured. A difficult airway cart was kept ready.

In the operating room, standard monitoring with electrocardiogram, non-invasive blood pressure, pulse oximeter and a precordial stethoscope were initiated. Intravenous atropine 0.16mg, midazolam 0.4 mg and Fentanyl 16µg were administered. The child breathed oxygen for After 3 minutes of preoxygenation, anaesthesia was induced with 3 to 5% Sevoflurane in oxygen. Adequate mask ventilation was achieved with a Rendell-Baker-Saucek mask size 2 with padding over the maxillae as the other masks were ill-fitting. A gentle check laryngoscopy was done to rule out anatomical deformities of the oropharynx and larynx. As vocal cords could be readily visualized, succinylcholine 15mg was administered and an orotracheal intubation was done with a 4.5mm internal diameter PVC South Pole tube. A throat pack was inserted after securing the endotracheal tube. Anaesthesia was maintained with sevoflurane in O_2:N_2O (50:50) and atracurium for skeletal muscle relaxation. The procedure lasted for 3 hours and the intra- and post-operative course were uneventful.

DISCUSSION

NOMA

Cancrum oris (Noma) is a devastating gangrenous disease that leads to severe tissue destruction in the face. The term Noma was first described by Tourdes in 1848 which originates from the Greek word Numein meaning 'devour'.[1] The peak incidence of this acute infection is around 1-4 years, coinciding with the period of linear growth retardation in deprived children.[2] The disease is mainly seen in Africa and occasionally in Latin America and Asia.[3] Predisposing factors include malnutrition, infectious diseases, HIV infection, measles and immune compromised conditions. Poverty is the most important risk factor and Noma is a good biological parameter of extreme poverty. It has been eloquently named the 'face of poverty'.[4] Noma in children is caused by corynebacterium, fusobacteria and bacteroids. [1]

NN is a completely different disease than Noma and is exclusively seen in neonates. Its name reflects the similarity of the facial lesions. However, the differences in age group, clinical course, microbiology and prognosis make them two distinct disease entities. The term NN was coined by Ghosal et al in 1977 from Calcutta, India, when they described forty eight cases of NN.[5] NN affects mostly preterm, low birth weight neonates, though three NN cases in full term newborns have been reported. NN occurs most commonly after third day, usually within the first two weeks of life. The onset of NN is followed by a rapid spread of sclerema followed by death within next 1-3 days. Most cases of NN have been described with Pseudomonas aeruginosa sepsis. NN needs early diagnosis and specific antimicrobial therapy. Despite aggressive treatment, most cases of NN die. Very few cases have reported survival.[6, 7] Extensive surgical debridement is contraindicated in these cases and reconstructive surgery is advocated after the first year of life.[7] Patients who survive NN suffer from its sequelae-facial disfigurement, trismus, and oral incontinence and speech problems.[8] Surgery is targeted at restoring normal speech, oral competence and acceptable cosmesis.

Anaesthesia described for Noma sequelae are limited. Various techniques for the management of difficult paediatric airway include awake intubation through a Laryngeal Mask Airway (LMA) [9] fiberoptic intubation [10] and tracheostomy.[11] Trismus may necessitate a fiberoptic nasal intubation. Ankylosis of the mandible has also been frequently reported and this may also affect the anaesthesia management.

Our patient had suffered from NN in the neonatal period which was treated with antibiotics, the details of which were not available. Nasal intubation was not possible as the nostrils were stenosed due to scarring. We anticipated a difficult mask ventilation rather than difficult intubation. We decided to preserve spontaneous respiration with Sevoflurane induction. A diagnostic laryngoscopy in deep plane of anaesthesia was performed to rule out any abnormal upper airway anatomy due to Noma sequelae. Appropriate size paediatric fiberoptic bronchoscope (2.2 mm) was not available at our institution. Even though the airway management of our patient was relatively uneventful, due caution should be exercised as difficulty may be encountered depending on the deformities due to the Noma sequelae. Securing airway in such patients may require novel techniques.

PAEDIATRIC DIFFICULT AIRWAY

Provision of a secure airway may prove challenging at times in paediatric or adult anaesthesia. Difficult paediatric airway could be because of various congenital and acquired causes. Management of paediatric airway requires expertise due to its peculiar anatomical variations. Generally, inhalation induction is the technique of choice as it preserves spontaneous ventilation and can be carried out with 100% oxygen to increase the margin of safety from hypoxemia. Sevoflurane is preferred over halothane.[12]

Various techniques of intubation have been described to manage difficult paediatric airway. Eipe et al have reviewed

the technique of submental intubation for a patient with Noma sequelae.[13] Fiberoptic intubation is the most versatile technique for securing the difficult airway which may be accomplished either awake or after inhalation induction. Ultra thin fiberoptic bronchoscopes are available with an external diameter of 2.2mm which can accommodate a 3mm endotracheal tube. It can be particularly frustrating using the oral approach due to lack of plastic airway guides for paediatric patients.

LMAs have a critically important role in the management of difficult paediatric airway. They are useful as ventilation adjuncts and conduits for tracheal intubation. Lastly, LMAs may be useful as rescue airway in a "cannot intubate/cannot ventilate" situation. A limitation of many infant sized LMAs when used as conduits to fiberoptic intubation is the inability to pass the pilot balloon of a cuffed endotracheal tube through the airway tube of LMA.

With recent advances in video technology have come many new tools for paediatric airway management. Optical stylets are useful in the management of difficult airway. Shikani optical stylet is a j-shaped stylet with central optical channel which can be used as a useful adjunct in the management of routine and difficult paediatric airway management. Bonfils Endoscope, a rigid stylet with 40 ^{O} anterior curve is recently modified for paediatric use. Currently, video/optical laryngoscopes like the Glidescope, Storz video laryngoscope; Airtraq and Truview EVO_2 are available in sizes appropriate for paediatric patients of all ages.[14]

CONCLUSION

In conclusion, airway management in children with Noma sequelae is challenging. Securing airway in such patients may require novel techniques and the success lies in adequate preparation, optimal use of the available airway equipments and availability of anaesthesiologists skilled in paediatric airway management.

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