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BINDER'S SYNDROME AND ANAESTHESIA.

Anaesthesiology

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ABSTRACT

Binder's syndrome is a rare congenital facial deformity. It is a rare form of maxillofacial dysplasia with several nasal and mid-face deformities. The etiology of this condition, as suggested by Binder, is a disturbance of prosencephalic induction center during embryonic life. There have been very little literature about anaesthetic concerns and intraoperative management regarding this uncommon disorder. As this syndrome is associated with a difficult bag-mask ventilation, difficult oral and nasal intubation, all the next plans must be kept ready, both during intubation and extubation. These patients may also be associated with mental retardation, cervical spine abnormalities and dental problems. Preoperatively anaesthetists must be aware of this anomaly and its peri-operative implications. In the present case, we are describing the anaesthetic considerations in a case of Binder's syndrome in a young male patient presented for elective nasal reconstruction surgery for cosmetic purpose and have difficulty in intubation. To the best of our knowledge, there had been very few previous reported articles on anaesthetic implications of Binder's syndrome.

KEYWORDS

Binder's Syndrome; Difficult Airway; Maxillofacial dysplasia; Reconstructive surgery.

INTRODUCTION

Binder's syndrome is a rare, congenital, maxillofacial dysplasia with several nasal and midface deformities requiring planned plastic and reconstructive surgery. Since it is an uncommon condition, peri-operative physicians may be unaware of its anaesthetic considerations. Along with the possibility of a difficult airway, there can be other co-existing conditions like mental retardation, cervical spine abnormalities and dental problems. Difficult airway cart, including fibre-optic bronchoscope, must be kept ready both during intubation and extubation. This article describes the anaesthetic management, in a case of Binder's syndrome posted for elective nasal reconstruction surgery. Except for difficulty in intubation, no other problems associated with Binder's syndrome were encountered in our case. For a successful perioperative outcome, adequate preoperative preparation should be done.

CASE REPORT

A 26-year-old male patient presented for elective nasal deformity correction. He was diagnosed as a case of Binder's syndrome after visiting the hospital for plastic reconstructive surgery by the Plastic surgery OPD team. On pre-anaesthetic evaluation, all routine investigations were within normal limits. He had complaint of occasional nasal obstruction. There was no recent history of upper respiratory infection. On airway examination, mouth opening 3 fingers, Mallampatti grade was class 2 . Other remarkable facial features included a depressed nasal bridge, prominent upper lips, slight mandibular prognathism. Neck movements and airway distances were within normal limits. The systemic organ system evaluation was also normal. Oral intubation was planned after discussion with the surgical team to avoid involvement of surgical field. All standard ASA monitors were attached. General anaesthesia was administered after preoxygenation, considering all the precautions of difficult airway. Due to the unavailability of FOB it was planned to proceed with video laryngoscope. The modified Cormack and Lehane grading was 2b. An oral south pole cuffed endotracheal (size 7.5mm) was tried with external laryngeal manipulation initially which was failed. Second attempt was given with the stellate which was also unsuccessful . Third attempt was made with a gum elastic bougie which was successful, and hence airway was secured with a cuffed endotracheal tube of size 8.0 mm ID .The patient was ventilated for few minutes and it was made sure that the oxygen saturation and other hemodynamic parameters were within normal limits. Nasal prongs were applied at 4 lit /min flow rate . The circuit was disconnected for a while and south pole tube was railroaded in the place of normal endotracheal tube using gum elastic bougie and 2% lignocaine gelly to lubricate and straighten the south pole. Bilateral equal air entry was confirmed with the help of chest rise, auscultation and capnography. The tube was fixed at the centre of the lower lip and mechanical ventilation was resumed. Oral packing was done and labelling was done and adequate care was taken for proper positioning. The surgery lasted for around six hours and was uneventful. After completion of surgery, throat pack was removed and thorough oral suctioning was done under vision to rule out any secretions or blood clots. Some blood clots were visualized in the pharynx which were coming out on applying pressure over soft palate , which were removed. The patient was extubated after adequate and spontaneous breathing efforts. A difficult airway cart was kept handy at the time of extubation also. The patient was observed in the post anaesthesia care unit before shifting to the ward.

DISCUSSION

Binder's syndrome [1] is an uncommon congenital condition with characteristic facial features. It was identified and defined by Von Binder in 1962 [2], though described earlier by Noyes in 1939. It is also called "maxilla-nasal dysostosis", with the following six characteristic features [3]: arhinoid face, intermaxillary hypoplasia (associated with malocclusion), abnormal position of the nasal bones, nasal mucosal atrophy, anterior nasal spine agenesis and a lack of frontal sinuses. Embryologically, there is a failure of development in the premaxillary area with associated deformities of the nasal skeleton and the overlying soft tissues (rhino-cephalic dysplasia) [4]. Patients have an unusually flat, underdeveloped midface and an abnormally short nose with the flat nasal bridge, lack of nasal tip projection, peri-alar flattening, mandibular prognathism and an acute nasolabial angle. Other associated features include mental retardation, bilateral hearing loss, prenatal vitamin K deficiency, irregularities in the cervical spine[5] (e.g. Separate odontoid process, short posterior arch, and spina bifida occulta), prominent lips, poor oro-dental hygiene and psychological problems due to cosmetic defect. According to Holmstrom, inheritance of this syndrome may be an autosomal recessive trait with incomplete penetrance [6]. These patients present for surgical correction of facial deformities and orthodontic treatment of dental problems. The main surgery performed is a nasal reconstruction with bone or cartilage grafts[7]. Others include maxillary protraction with rapid palatal expansion. Patients may come for repeated surgeries due to graft resorption or an unsatisfactory appearance. The main anaesthetic consideration is the presence of difficult airway. Nasal intubation should not be attempted in these patients due to reduced nasal passages and possibility of trauma during tube passage. Insertion of a nasogastric tube can also be challenging. Direct laryngoscopy must be gentle and cautious due to dental malocclusion and crowding. Complete difficult airway cart[8] must be prepared and checked preoperatively. Associated abnormalities with Binder's syndrome also need to be evaluated in the pre-anesthetic assessment. An otorhinolaryngologist evaluation, cervical spine X-rays, psychological counselling, dental opinion and measures to improve oral hygiene are important in the preoperative period. Awake fibreoptic

oral intubation (after adequate pre-oxygenation) can be attempted under intravenous sedation with Dexmedetomidine or Remifentanil infusion, along with upper airway blocks, nebulised lignocaine and "say go" technique during fibreoptic bronchoscopy. If airway assessment is otherwise normal, a video-laryngoscopic guided oral intubation can be done after intravenous and/or inhalational induction, with the muscle relaxant given after confirmation of correct tube placement. A wire-reinforced tracheal tube is preferable to prevent its kinking during head and neck movements during surgery. Endotracheal tube fixation requires special attention as it should not come in the way of the surgical field and at the same time do not cause a drag on the angles of the mouth. Tube fixation at the centre of the lower lip may be preferred to help the surgeon assess the symmetry of the mid-face during reconstruction. Oral packing must be done to prevent blood and secretions trickling into the lower airway during surgery. Care must be taken in documenting the use of oro-pharyngeal packs (with a radio-opaque line) and remembering to remove them at the end of surgery[9]. Since osteotomy and grafts are required for the surgery, pain management (multi-modal techniques) must be given adequate importance. Temperature maintenance (normothermia) must be ensured, especially in prolonged procedures with the help of patient and fluid warmers. Difficult airway precautions must be followed at the time of extubation as well, including tube exchanger device. There has been less anesthesia-related literature concerning Binder's syndrome. Most importantly, the attending anesthesiologist must be aware of this rare syndrome and its associated anomalies for improving the overall perioperative outcome of the affected patients. This case was reported to improve the knowledge bank of the anesthesiologists worldwide, to handle such cases effectively.

CONCLUSION

Binder's syndrome, though rare, must be kept in mind while anaesthetizing a patient for facio- maxillary reconstructive surgery. A thorough pre-operative evaluation for concurrent conditions and complete airway assessment is of paramount importance. Since these patients can present for several staged procedures, multiple anaesthetic exposures is a possibility, especially in a scenario of difficult airway. Awareness of this rare syndromic disorder enables the anesthesiologist to prepare the operation theatre adequately and to manage the associated problems successfully.

Conflicting Interests

The authors declare that except difficult intubation they have no conflicting Interests.

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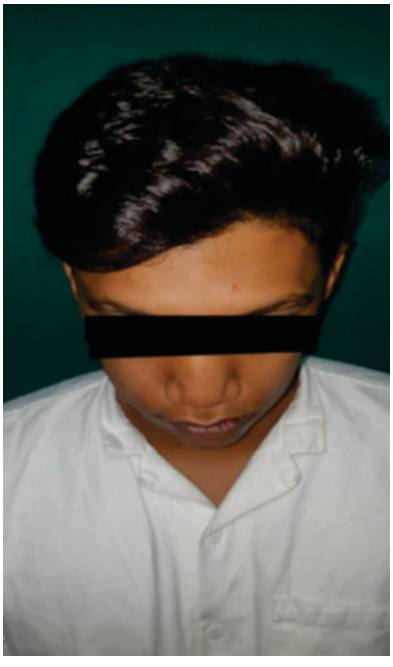

Image 1

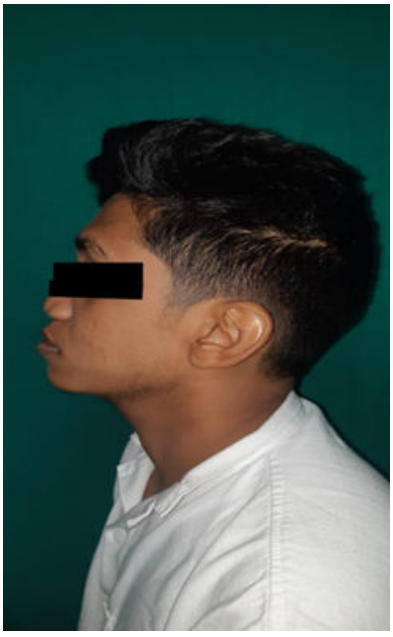

Image 2

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