Case Report

Anaesthetic management of a case of Haberland's syndrome (encephalocraniocutaneous lipomatosis)

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ABSTRACT

While the Haberland syndrome was first reported in 1970, the anaesthetic management of this rare cohort of patients has not been described. With only 54 such cases reported, describing primarily the unilateral cutaneous, ocular, and neurologic malformations associated with this syndrome without focussing on the anaesthetic management. We describe the case of a 7 year old case of Harberland syndrome with special focus on the difficulties faced by us in mask ventilation, as well as intubation and the need for elective ventilation and planned extubation. We also discuss the precautions to be taken while undertaking such a case like preparations for emergency tracheostomy and possibility of re-intubation after extubation. A meticulous preoperative workup along with neurological and airway examination along with preparation for elective ventilation and tracheostomy is a prerequisite for the successful management of this case.

Key words: Difficult airway, Haberlands syndrome, Mallampatti grade, nasal intubation, odontogenic cyst

INTRODUCTION

Encephalocraniocutaneous Lipomatosis (ECCL) or Haberland syndromeis an uncommon neurocutaneous syndrome. It is characterised by unilateral cutaneous, ocular, and neurologic malformations. Clinically, this syndrome is characterised by unilateral lipomatous hamartomas on the scalp and face, ocular damage and ipsilateral malformations of the central nervous system. Certain abnormalities associated with this syndrome may pose a challenge to the anaesthesiologist when such cases are posted for therapeutic, corrective or cosmetic surgeries. To the best of our knowledge, anaesthetic management of such a case has not been reported yet. Informed written consent was taken from the parents prior to the write-up and publication of this case report.

CASE REPORT

A 7-year-old boy, born by a normal vaginal delivery at term to a non-consanguineous parentage was diagnosed to have Haberland's Syndrome in the neonatal period. The child had delayed milestones and seizure disorder, for which he was on anticonvulsants. Presently the child had a 3-day history of fever, swelling in the jaw, and difficulty in breathing and swallowing, and was posted for incision and drainage of odontogenic cyst.

The child weighed 18 kgs, was well-nourished and had a large swelling in the left side of the mandible and cheek, with signs of airway narrowing [Figure 1a]. He was tachycardic and tachypnoeic. His mouth opening was limited to 1-finger breadth [Figure 1b] and neck movements were restricted. Computed Tomography (CT) head and neck showed narrowing of the oropharynx on the leftside due to the mandibular

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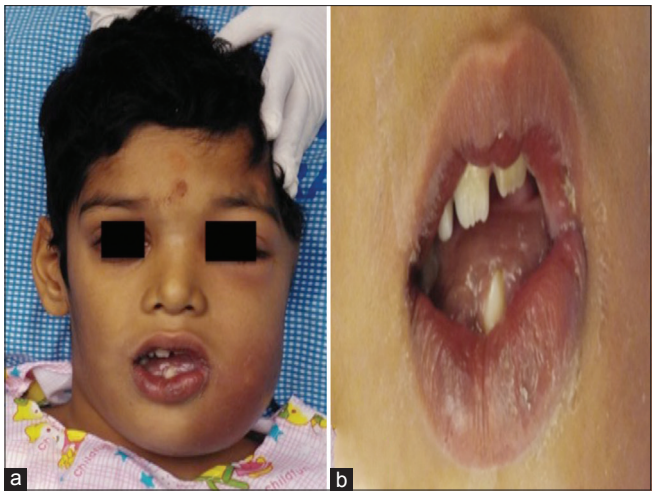

Figure 1: (a): Clinical Photograph showing the large swelling involving the left face, mandible and cheek causing the airway narrowing and difficulty in intubation; (b): Clinical Photograph showing the mouth opening of the patient (1 finger)

mass [Figure 2]. Difficulty in mask ventilation, as well as intubation, was anticipated, and difficult airway cart was kept ready.

The child was premedicated with intravenous (IV) midazolam (0.02 mg/kg) and glycopyrrolate (0.005 mg/kg) in the preoperative area. In the operating room, after placement of monitors (electrocardiogram, pulse oximeter, non-invasive blood pressure, skin temperature probe), intravenous ketamine (0.4 mg/kg) and midazolam (0.02 mg/kg) was administered, to allow airway manipulation while preserving his spontaneous ventilation. The upper airway was sprayed with 10% lignocaine and xylometazoline was instilled in the nostrils. A nasopharyngeal airway (size 3.5) in the left nostril, was used to induce anaesthesia with oxygen (O_2) and sevoflurane, via the circle system, and monitor end-tidal carbon dioxide ($ETCO_2$). Fibreoptic bronchoscopy (FOB) was done through the other nostril, using a size 3.5 bronchoscope, and a size 5 cuffed endotracheal tube (ETT). Airway narrowing and oedema were noted. Airway collapse was prevented by a gentle jaw thrust by an assistant. ETT was secured in place and was confirmed with auscultation and capnograph. Rocuronium (0.5 mg/kg) IV was used to achieve neuromuscular blockade and the ventilation was controlled. Anaesthesia was maintained with O_2, air and sevoflurane. Intravenous paracetamol in the dose of 250 mg was given for analgesia and dexamethasone IV in the dose of 4 mg was given for airway oedema. Incision and drainage of the cyst was carried out and the total procedure lasted for 1 hour. In view of airway oedema and difficult intubation,

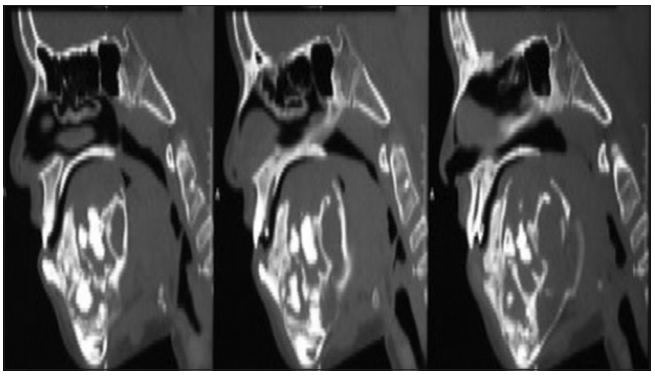

Figure 2: Mid-Saggital section of non-contrast Computed tomography image of the mouth and oropharynx showing the odontogenic cyst belwo and mandible above with severly compromised airway

the child was shifted to paediatric intensive care unit (PICU) for elective ventilation and planned extubation.

After ventilator support under sedation for 24 h, adequate preparations for reintubation and emergency surgical airway placement, an extubation trial was attempted. The child was given injection midazolam (0.5 mg IV) and adequate spontaneous respiratory rate and tidal volume were ensured. Fibreoptic bronchoscopy was done through the opposite nostril, and reduction in airway oedema was noted. The upper airway was suctioned thoroughly. Then the bronchoscope was reintroduced through the nasal ETT, and extubation was done over the bronchoscope along with suctioning. Child maintained the airway well and vitals were stable.

The child recovered well and was discharged on postoperative day 2.

DISCUSSION

ECCL is a unique, non-progressive congenital syndrome involving mainly the cutaneous, ocular, and neurologic systems. The first case was described by Haberland and Perou in 1970.[1] It is a rare syndrome and only 54 cases have been delineated since 1970.[2-5] Clinically, ECCL is characterised by unilateral abnormalities of the brain, eyes, and skin. The most common neurological findings are hemiatrophy, dilated ventricles, porencephalic cysts, abnormal calcifications, intracranial lipoma, and cranial asymmetry. Most patients present with seizures and mental retardation.[5-12] The hairless lesion of the scalp ("nevuspsiloliparus") is pathognomonic, and the papular lesions of the eyelid, consistent with lipomas, are the most frequent feature in all cases reported.

However, other lesions have also been reported, such as lipomas of the vertebral spine, odontoma, and "café-au-lait" spots.[5,12]

Skeletal anomalies like macrocephaly, jaw tumours, non-ossifying fibromas, and osteolyticbone lesions have been described. These lesions are progressive unlike other features of the disorder. Coarctation of the aorta is the prominent feature in the cardiovascular system and conduction abnormalities may be seen due to fibrosis of the atrial walls.[2]

Our patient had delayed developmental milestones, seizures, jaw tumor, cranial asymmetry, which were significant for the anaesthesia management, along with a hairless scalp lesion and abnormalities of the eye.

Management of anaesthesia is largely influenced by the mental status, underlying seizure disorder, status of cardiac function and conduction as well as the airway characteristics such as abnormal calcifications, jaw, and neck mobility. Postoperative airway management is also of the utmost importance in such cases.

Adequate antiseizure prophylaxis, perioperative maintenance of cardiac rhythm, was important in our case. Also, a detailed preparation for difficult ventilation and intubation as well as a plan for emergency surgical airway was needed. We preferred to preserve spontaneous ventilation till a definitive airway was established, and opioids were used sparingly intra and postoperatively, to prevent undue respiratory depression. A planned postoperative airway assessment and controlled extubation was required.

CONCLUSION

The patients with Haberland's syndrome pose a significant challenge to the anaesthesiologist owing to a difficult airway, problems with cardiac conduction, risks of perioperative seizures, the tendency for difficulty in positioning due to abnormal ossification and difficulty in patient cooperation due to mental status. A thorough preoperative workup by the anaesthesiologist, neurological and radiological evaluation, planning for anaesthesia and postoperative airway management is of utmost importance in these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate consent from the parent(s) of the patient. In the form the parent(s) has/have given his/her/their consent for his/her/their child's images and other clinical information to be reported in the journal. The parent(s) understand that their child's names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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