Case Report

Anaesthesia Management of a Patient with Saethre-Chotzen Syndrome – A case report

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Abstract

Saethre-Chotzen syndrome (SCS) is a craniosynostosis syndrome characterized by premature fusion of sutures. Patients with SCS have midface hypoplasia, a high-arched palate, obstructive sleep apnea, increased intracranial pressure, congenital heart malformations and ophthalmic disorders. Patients with SCS may be required general anaesthesia due to ophthalmic disorders. These patients may have difficult face mask ventilation, laryngoscopy or intubation. The anaesthesia management of SCS may be similar to other craniosynostosis syndromes. Due to upper limb defects, difficult intravenous cannulation may be a problem. We suggest an appropriate preoperative evaluation, physical examination, and preoperative preparation for difficult airway management and intravenous cannulation. Here, we report an uneventful general anaesthesia management of a child with SCS diagnosed, who underwent strabismus surgery.

Keywords : Craniosynostosis syndrome, Saethre-Chotzen syndrome, Strabismus surgery, Airway management

Introduction

Saethre-Chotzen syndrome (SCS), or acrocephalosyndactyly type III, is a rare craniosynostosis syndrome characterized by premature fusion of sutures with an autosomal dominant inheritance pattern. The prevalence of SCS is between 1:25,000 and 1:50,0001. This syndrome is caused by mutations of the TWIST1 gene. It is characterized by coronal synostosis, brachycephaly, a low frontal hairline, low-set ears, clinodactyly, syndactyly, mild-to-moderate intelligence disabilities, increased intracranial pressure, and congenital heart malformations. Ophthalmic involvement includes hypertelorism, ptosis, strabismus, amblyopia, loss of vision,

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Article History
Received: 1st Oct 2022
Revision: 15th Oct 2022
Accepted: 22nd Nov 2022
Published: 28th Jan 2023

How to cite this article: Sibel Catalca, Ozlem Ozmete, Ozlem Ozkan Kuscu, Cigdem Simsek, Meltem Kipri, Oya Yalcin Cok, Nesrin Bozdogan Ozyilkan. Anaesthesia Management of a Patient with Saethre-Chotzen Syndrome – A case report. Ind J Ophthal Anaesth 2022;3(1):16-9

downward-slanting palpebral fissures, epicanthal folds and blepharophimosis. The clinical features which lead to possible airway management problems are midfacial hypoplasia, high arched palate and obstructive sleep apnea (OSA).[1,2] Patients with pediatric syndromes may need medical or surgical treatment for ophthalmic disorders. These patients may require sedation or general anaesthesia to provide ideal conditions for surgery and repeated examination and treatment. Anaesthetic management of SCS can be challenging for anesthesiologists because of these deformities which can lead to difficult airway management and intravenous cannulation as well as intraoperative challenges such as interaction between drugs used for general anaesthesia and ophthalmic drugs, increased intraocular pressure and oculocardiac reflex.[3] There is limited information about the anaesthetic management of SCS in the literature. Here, we report general anaesthesia management of a child with diagnosed SCS for strabismus surgery.

Case report

A 3-year-old, 18 kg male patient was evaluated for elective strabismus surgery. In the preoperative evaluation, the patient was diagnosed with SCS with clinical features of the syndrome such as mental retardation, micrognathia, macrocephaly, strabisumus and with craniofacial abnormality, Mallampati score was III. See Figure 1 His laboratory tests, electrocardiography and echocardiogram were within the normal range.

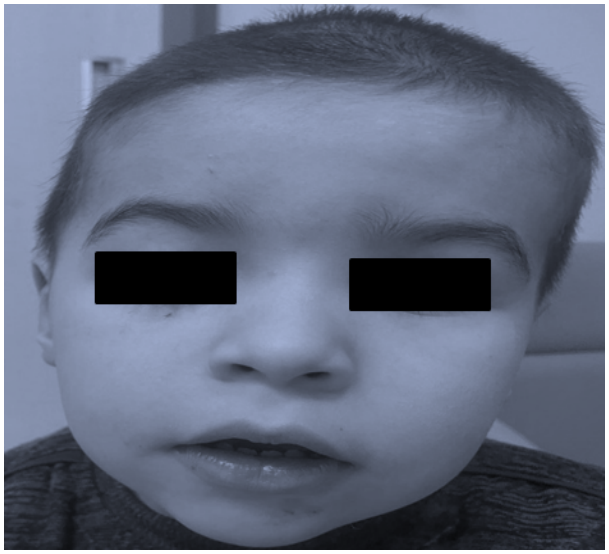

Figure 1. Facial features of the patient with Saethre-Chotzen syndrome

After obtaining written informed consent from his parents, the patient underwent standard anaesthesia monitoring. The patient's initial vital signs in the operating room were as follows: heart rate, noninvasive blood pressure and SpO_2 were 113 beats/min, 84/55 mmHg, and 98%, respectively. All alternative difficult airway devices according to pediatric difficult airway algorithm (oral and nasal airways, different-sized endotracheal tubes, laryngeal mask airway (LMA), video laryngoscope, fiberoptic bronchoscopy) and ultrasonography (USG) were kept available in the operating room due to a possible requirement for difficult airway management and difficult intravenous cannulation. Intravenous cannulation was achieved by an experienced anesthesiologist without USG. Anaesthesia was induced by lidocaine (1 $mg.kg^{-1}$), propofol (3 $mg.kg^{-1}$), and fentanyl (1 $\mu g.kg^{-1}$). Face mask ventilation was proved to be sufficient. The airway was successfully secured with a size 2.0 LMA.

Anaesthesia was maintained with 2% sevoflurane, and 1:1 O_2/N_2O. The operation was completed after 75 minutes without any hemodynamic instability. Regarding the risk of an immediate respiratory depression due to OSA, the patient was extubated in the operating room when fully awake, still with the precautions for a failed postoperative airway management in place. Then he was transferred to postoperative care unit. The patient was followed closely in the postoperative care unit and then transferred to the ward after the observation duration.

Discussion

Patients with special genetic syndromes may have surgery for ophthalmic issues and require attentive anaesthetic management. While patients sometimes have diagnosed syndrome beforehand, genetic syndromes should be highly suspected in the patients with specific clinical features during preoperative evaluation and consultation with a paediatrician or geneticist is required to confirm the accurate diagnosis.

Patients with SCS have many prominent clinical features which may, in turn, affect anaesthetists' care and preferences for the anaesthesia plan. Although there are many reviews about such craniosynostosis syndromes, there is limited literature describing the perioperative management of SCS. The management plan should include a preoperative visit to figure out possible perioperative risk factors and issues to deal with.

Ideally, anaesthesia management of these cases should be safe, practical, and without adverse effects. Appropriate perioperative management will provide a stable hemodynamic, minimal effect on intraocular pressure, and prevent potential procedure-specific adverse events3. Patients with midface hypoplasia, fused cervical vertebrae, cleft palate or proptosis can have difficult face mask ventilation, laryngoscopy or intubation. Due to upper limb defects, difficult intravenous cannulation might be expected in patients with SCS. Intubation and extubation are the most important stages of anaesthesia management that affect the patient's hemodynamics. The use of endotracheal tube may increase intraocular and intracranial pressure more than LMA. It prolongs the anaesthesia duration and recovery time. Extubation and emergence periods may be complicated with OSA.[1,4] Increased intracranial pressure and congenital heart anomalies may be a big problem during intraoperative management because of hemodynamic instability risk.

In our case, we used a LMA to avoid hemodynamic instability and the risk of increase in intracranial and intraocular pressure because LMA was an appropriate airway device for strabismus surgery. Therefore, we had no problem with airway management and hemodynamic stability.[5,6] The anaesthetic agents may affect intraocular and intracranial pressures.[3] We preferred propofol, lidocaine and fentanyl to maintain intraocular and intracranial pressures' stabilities.

In conclusion, when general anaesthesia is required for ophthalmic procedures in pediatric patients with SCS, perioperative evaluation and management of pediatric patients with SCS should be given attention to not only critical clinical features but also nonphysiological conditions such as hypoxia, desaturation or bradycardia which compromise patients' tolerance to anaesthetics' effects. We suggest an attentive preoperative evaluation, physical examination, and preoperative preparation for difficult airway management such as alternative airway devices and difficult intravenous cannulation in patients with SCS. Anaesthetists should know the factors and anaesthetic agents that influence physiological stability of this vulnerable population.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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