Abstract: Saethre-Chotzen syndrome (SCS) is a type of acro-cephalo-syndactyly (ACS) syndrome, characterized by premature fusion of the coronal sutures, facial dysmorphism, syndactyly, skeletal deformity, and congenital heart malformations. We here describe a child with diagnosed SCS, who underwent squint surgery under general anesthesia, and review the anesthetic concerns thereof.

Key words: Saethre-Chotzen syndrome (SCS); general anesthesia; squint surgery; airway; mandibular hypoplasia.

Saethre-Chotzen syndrome (SCS) is a type of acro-cephalo-syndactyly (ACS) syndrome. ACS syndromes are a group of autosomal dominant syndromes, in which craniosynostosis is associated with acrocephaly and syndactyly. Main features characterizing SCS, also known as ACS III, are premature fusion of the coronal sutures leading to skull deformation, facial dysmorphism, syndactyly, skeletal deformity and congenital heart malformations. Presence of mid-facial hypoplasia is expected to predispose the patient to a difficult airway (Table 1).

These patients can present with multiple surgical and medical complaints. Most of them will undergo surgery and anesthesia at least once in their life time for correction of craniofacial, orthopedic, ophthalmic, or cardiac lesions apart from incidental surgical conditions. We here describe a child with diagnosed SCS, who underwent squint surgery under general anesthesia, and also discuss the associated anesthetic concerns thereof.

Case report

A four year old female child weighing 15 kg presented to the ophthalmic outpatient department with esotropia and bilateral superior oblique muscle palsy. She had dysmorphic facial features and hence was sent for genetic counseling, where SCS was diagnosed. Her birth and developmental history were otherwise normal. She had normal learning abilities. There was no history suggestive of raised intracranial pressure, congenital heart disease, or obstructive sleep apnea (OSA). On examination, the child’s skull was brachycephalic with hypertelorism and ptosis of the left eye (Fig. 1). Cutaneous syndactyly was present in both upper limbs. There was no other limb deformity. She had mid facial hypoplasia and airway examination revealed a high arched palate (Fig. 2). No other airway abnormality was noticed. Cervical spine X-ray did not show any vertebral anomaly. All biochemical investigations were within normal limits. The systemic examination was unremarkable. During pre-anesthetic evaluation, the child was cooperative but unwilling for intravenous access. She was explained about steps of inhalational induction. Informed written consent was obtained from child’s parents during the preoperative period. The plan for airway management included the use of a supra-glottis device, and, if required, fiber-optic guided intubation of the trachea.

In the operative room (OR), all routine monitors (ECG, pulse oximetry and NIBP) were installed. Inhalational induction was performed using incremental doses of sevoflurane in nitrous oxide and oxygen. Inhalation fraction of sevoflurane ranged between 5 and 8%. After induction, a 24G cannula was inserted into a vein of the right hand. Once adequate mask ventilation was established, atracurium 5 mg and fentanyl 40 µg were given. The airway was then secured using a 1.5 size air-Q™ intubating laryngeal airway (ILA) (Cookgas LLC, Mercury Medical, Clearwater, FL, USA). Throughout the airway management, the head of the patient was kept in neutral position. Anesthesia was...
Discussion

SCS was named after two physicians who independently reported it in the early 1930s, Haakon Saethre, a Norwegian psychiatrist, and F. Chotzen, a German psychiatrist (1, 2). Closely related to Crouzon syndrome, SCS results from an early fusion of the coronal or lambdoidal suture. It occurs with a frequency of one in 25,000-50,000 live births.

Table 1

The various organ systems involvement and their anesthetic concerns in a patient with Saethre-Chotzen Syndrome (4, 5, 6, 7)

<table>
<thead>
<tr>
<th>Organ involvement</th>
<th>Characteristic findings</th>
<th>Anesthetic concerns and their management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Airway</td>
<td>Progressive cervical spine fusion, cleft palate, high arch palate</td>
<td>Limited neck extension, difficult intubation, cervical spine instability</td>
</tr>
<tr>
<td>Face</td>
<td>Mid facial hypoplasia, small maxilla, relative mandibular prognathia and asymmetry, underdeveloped cheekbones, deviated nasal septum, narrow palate, cleft palate</td>
<td>Difficult bag and mask ventilation, difficult oral and nasal intubation</td>
</tr>
<tr>
<td>Skeletal deformity</td>
<td>Short stature, cutaneous syndactyly, small distal phalanges, clinodactyly of fifth finger, digitalization of thumb, limited elbow extension, contracture of elbow and knee</td>
<td>Difficult positioning on OT table, difficult IV access</td>
</tr>
<tr>
<td>Chest</td>
<td>Short clavicles</td>
<td>Difficult subclavian vein cannulation.</td>
</tr>
<tr>
<td>Neurological disorders and skull</td>
<td>Brachycephaly, plagiocephaly, late closure of fontanelles, ossification defects, hyperostosis of skull</td>
<td>Elevated ICP, seizure disorders</td>
</tr>
<tr>
<td>Eyes and Ears</td>
<td>Eyes- Shallow orbits with orbital asymmetry, orbital hypertelorism, ptosis, strabismus, blepharophimosis, down-slaning palpebral fissures, sparse eyebrows medially, epicanthal folds, optic atrophy, and corneal opacity. Ears- small, low-set, unusually shaped ears, SNHL (sensorineural hearing loss)</td>
<td>Increased incidence of oculocardiac reflex (OCR), and postoperative nausea and vomiting during eye surgery. Difficulty in communication with child</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Atrial septal defect, ventricular septal defect, pulmonary stenosis, patent ductus arteriosus, and tetralogy of Fallot</td>
<td>Anesthetic implications as per the cardiac lesion</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>Renal anomalies, cryptorchidism, ano-rectal malformations</td>
<td>Deranged renal parameters, electrolyte imbalance</td>
</tr>
</tbody>
</table>

Fig. 1. — Saethre chotzen syndrome: esotropia, bilateral superior oblique muscle palsy with brachycephaly, hypertelorism, ptosis of the left eye.

Fig. 2. — Airway examination may reveal high arched palate.

maintained using isoflurane in 50% nitrous oxide/oxygen. For postoperative nausea and vomiting prophylaxis, 1.5 mg ondansetron was administered intravenously. At the end of surgery, neuromuscular blockade was reversed using 750 µg of neostigmine and 150 µg of glycopyrrolate intravenously. Air-Q was removed once the child was fully awake. The patient had uneventful recovery and post-operative period.

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SCS is an autosomal dominant inherited syndrome, and results from a mutation of the TWIST1 gene on Chromosome 7, which plays a key role in the early development of the skull, face and limbs (3). The signs and symptoms of SCS vary widely, even among affected individuals in the same family. However, because symptoms may be mild, it can go undetected in some family members. Although the literature teems with reports and reviews about anesthetic management of patients with other forms of craniosynostosis such as Apert syndrome, the literature regarding this syndrome is scarce. This is surprising, insofar as it is the most common form of cranio-synostosis syndromes. One reason may be lack of very marked features, which may lead to under recognition and under reporting.

The main pathological feature of this syndrome is an early fusion of cranial sutures, especially the coronal suture. It may lead to brachycephaly and plagiocephaly, late closure of fontanelles, and reduced ICP. Our patient had brachycephaly, but did not have raised ICP.

Mid facial hypoplasia leads to small maxilla and relative mandibular prognathia, as well as high arched palate (5). These patients can have beaked nose, deviated nasal septum, narrow palate, cleft palate, super numerary teeth, small, low-set, unusually shaped ears, and enamel hypoplasia. Facial appearance tends to improve with age throughout childhood. High-arched palate is a known indicator of difficult airway. It happens when mandibular hypoplasia shifts the tongue superiorly, right into the space that would typically be occupied by the developing palate. High arched palates make placement of the tube difficult, because of limited lateral space availability. All these facial features can lead to difficulty in bag and mask ventilation, intubation, and LMA insertion. Since our patient had high arched palate, we chose air-Q® for airway management. Indeed, this supra glottic airway device allows blind and fiber-optic guided intubation. This option would have been helpful if ventilation through the air-Q® had been compromised. Detailed airway examination using indirect laryngoscopy (IDL) is recommended prior to anesthesia, whenever possible, to prepare airway management and map any upper airway deformity (6, 7).

Vertebral fusion is also a known association with this syndrome. This is progressive and hence may present at a more advanced stage, leading to cervical instability. The ideal age recommended for screening eventual vertebral fusion is two to three years. At this age, the vertebral bodies are ossified, and cervical instability can be seen on standard neck X-ray. Despite normal cervical spine x-ray in our patient, we took all precautions to prevent cervical spine movement during airway management at the time of induction and recovery from anesthesia.

These patients may have shallow orbits with orbital asymmetry. It may predispose to exacerbated oculo-cardiac reflexes (OCR) during extra-ocular muscles handling.

SCS may concern multiple organs. The predominantly involved systems are the cardiac system, the skeletal system, as well as the sensory and motor systems. These and their associated anesthetic concerns are tabulated in Table 1 for easy referral.

We did not encounter any difficulty during mask ventilation or LMA placement in our patient. Netke et al. have reported a series of 10 patients with this syndrome, who underwent GA uneventfully (6). They hypothesized that this syndrome is associated with fewer chances of airway compromise as compared to other acro-cephalo-syndactyly syndromes, such as Apert syndrome, Crouzon syndrome, Pfeier syndrome, and Carpenter syndrome. Other authors have also reported uneventful anesthesia courses in children with SCS. However, it is imperative for successful anesthetic management that the variability of its features be taken into account (Table 1) (5, 7).

CONCLUSION

SCS is a type of ACS with variable presentation but may be associated with airway difficulty and other anesthetically important challenges. Being aware of these co-morbidities is important for proper anesthetic management of those patients. If undiagnosed, co-morbidities may lead to un-anticipated problems during anesthesia.

References
