Anaesthetic management of a patient with Crouzon syndrome

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Abstract

Crouzon syndrome is a rare hereditary disorder, characterised by marked craniofacial dysostosis from birth or early childhood. Typically, patients present in early childhood for craniofacial reconstruction surgery. Presentation in early adulthood is unusual. The most challenging aspect, for an attending anaesthesiologist, is the management of the difficult airway that is usually present in these patients, due to various craniofacial abnormalities of the neck region. Regional anaesthesia may be complicated in these patients, because of vertebral abnormalities. Here, we describe the successful neuraxial anaesthetic management of a 17-year-old male with Crouzon syndrome, who presented to us in the orthopaedic emergency outpatient department with a fracture of the left tibia, sustained during an accident.

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Introduction

Childhood syndromes pose unique challenges to the attending anaesthesiologist, whether they present during early childhood, or adulthood. The most difficult among these syndromes are associated with craniofacial deformities because they make airway management extremely challenging and difficult. An example is Crouzon syndrome, which is characterised by marked craniofacial dysostosis from birth.[1] Such patients may present to the hospital for reconstructive surgery of the craniofacial bones, or for surgical procedures for unrelated issues. This case study describes a patient with Crouzon syndrome who was brought to the emergency trauma ward with a fractured tibia, and the challenges relating to neuraxial anaesthesia.

Case study

A 17-year-old male, accompanied by his parents, presented to the orthopaedic emergency trauma ward with a fracture of the left tibia, sustained during a roadside accident. Initial examination revealed abnormal facies and the diagnosis of Crouzon syndrome, based on the clinical features, was made by the attending physician. A closed fracture of the left tibia was diagnosed clinically and with the appropriate radiological investigations.

According to the patient's parents, he had exhibited the characteristic features at birth, but these had progressively become less marked as the patient grew. There was no significant family history. Surgical correction of the facial deformities had been advised, but because of economic constraints and other issues that the family did not disclose, the patient had never had this carried out. The patient had delayed milestones and mild mental retardation. A history of mild intermittent headaches was noted, but the patient did not have any visual disturbances. Hydrocephalus was ruled out by the neurosurgeon. An ear, nose and throat examination indicated mild conductive deafness. Airway examination showed adequate mouth opening, despite a narrow oral aperture and high arched palate. A class 3 Mallampati score was calculated. The neck was noted to be short, and neck movement was grossly restricted.

Craniofacial examination revealed a malformed skull with flat occiput and microcephaly, a prominent forehead with frontal bossing, mild exophthalmos, mild divergent strabismus, a slightly beaked nose, a compressed nasal bridge, mild mandibular prognathism, a V-shaped maxillary dental arch, a narrow and high palate, malformed, widely spaced upper teeth, and anomalies of the spine, such as scoliosis (Figures 1, 2 and 3).

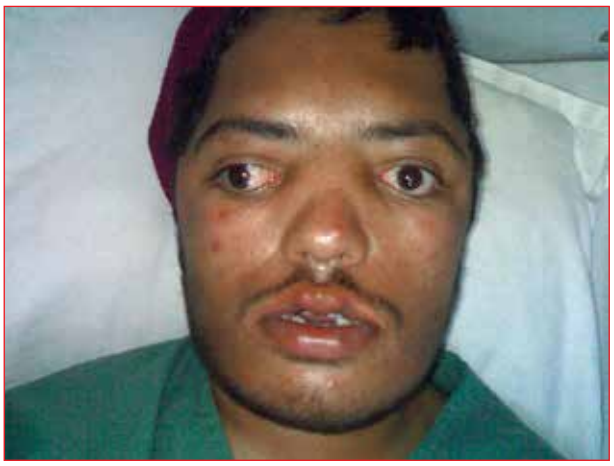

Figure 1: The face with typical features of Crouzon syndrome

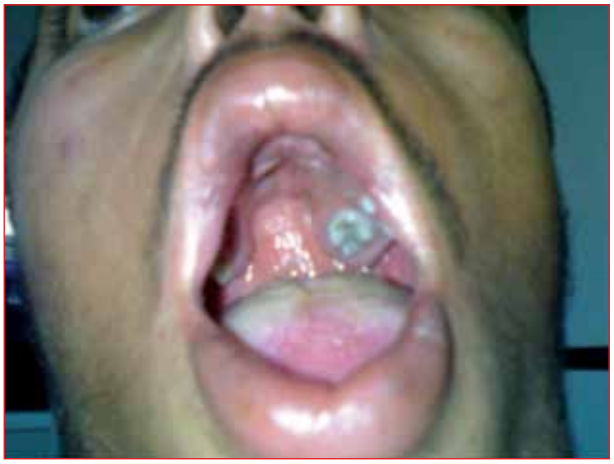

Figure 2: The intra-oral view, showing the high, arched palate and other features of Crouzon syndrome

On clinical examination, the pulse rate was observed to be 78/minute and blood pressure to be 116/82 mmHg. There was bilateral equal air entry of the chest with vesicular breathing. No abnormality of the heart was detected by auscultation. Special investigations were within normal limits, and haemoglobin was estimated at 11 gm/dl.

The patient was booked for placement of an interlocking nail of the tibia by the orthopaedic surgeon. The immaturity and the mental condition of the patient prompted a thorough discussion with the parents regarding the various implications, and complications, of general and regional anaesthesia, and the merits of neuraxial anaesthesia. After this counselling session, the parents agreed to neuraxial anaesthesia, and gave their consent to the administration of general anaesthesia too, in case the need arose.

The choice of neuraxial over general anaesthesia was mainly driven by the possibility of difficult airway management. The patient exhibited a good level of cooperation during the counselling session. He was not given any sedative premedication. Ranitidine 150 mg per os was administered overnight and on the morning of the surgery, with a sip of water. In the operating room, the procedure was explained

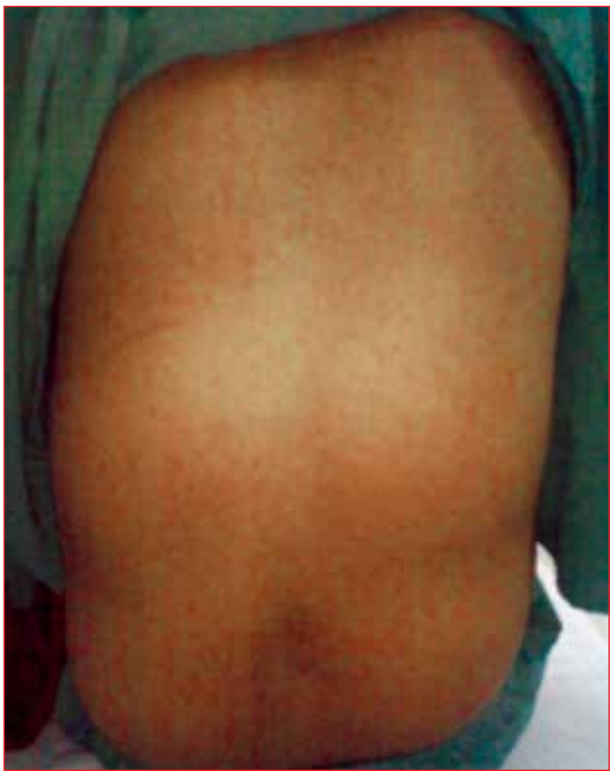

Figure 3: The posterior view of the patient, showing slight scoliosis

to the patient once more, and he was warned of mild pain during the establishment of epidural anaesthesia. The presence of the parents might have been comforting for the patient, but currently, family members are not allowed inside the operation theatre at our institution. One millilitre of lignocaine 2% was infiltrated at both the L3-L4 and the L4-L5 interspaces. Epidural anaesthesia was attempted with an 18 G Tuohy epidural needle at the L3-L4 interspace, but after three attempts, with bony resistance encountered at each attempt, this was abandoned. The epidural anaesthesia was planned with provision for postoperative pain relief through top-up doses of local anaesthetics through the epidural catheter.

Thereafter, we decided to administer subarachnoid block with a 26 G Quincke spinal needle. The spine had a little scoliosis. Hence, after two failed attempts in the midline, we persisted with a paramedian approach at the L3-L4 level. Successful lumbar puncture was achieved, and 2.8 ml of heavy bupivacaine hydrochloride was injected into the subarachnoid space, after establishing a clear flow of cerebrospinal fluid.

Surgery commenced after establishing an adequate sensory block at the T10 level, which was judged five minutes after injection by the pin-prick method. The delayed checking was deliberate, to avoid any panic because of pain during pin-prick testing. Throughout the surgical procedure, a difficult airway management trolley was kept ready besides the operation table. This trolley included an appropriately sized laryngeal mask airway and a fibre-optic bronchoscope. No

sedation was administered during the surgical procedure to avoid any chance of airway obstruction. The surgical procedure was one-and-a-half hours in duration. Both this, and the recovery period, were uneventful. No complications occurred. The patient was discharged home after eight days, with advice for follow-up after 15 days. This prolonged stay was at the request of the parents, as it was difficult for them to take care of him at home. During the hospital stay, the patient was kept in a private ward.

Discussion

Crouzon syndrome is a rare autosomal dominant disorder, with a prevalence of 1:60 000 live births. There is variable expression of the gene, leading to a variation in the phenotypic presentation.[1,2] The mutation of the FGFR2 genes, which is responsible for the condition, is also present in Apert syndrome, Pfeiffer syndrome and Jackson-Weiss syndrome.[3,4] Crouzon syndrome is characterised by craniofacial dysostosis, which includes a triad of skull deformities, facial anomalies and exophthalmos. It is responsible for approximately 4.8% of all cases of craniosynostosis,[5,6] and there is no race or sex predilection. Usually, it is detected at birth or in infancy. Characteristic dysmorphic features are detected. Craniofacial deformity occurs mainly because of different rates and timing of closure of skull sutures, resulting in either premature or delayed closure, causing various abnormalities and deformities of the face.[7,8]

These dysmorphic features either become more prominent, or may show regression in deformity with advancing age. These features include exopthalmos, hypertelorism, strabismus, hypoplastic maxilla, a beaked nose with a compressed nasal bridge, deformed lips, mandibular prognathism, optic atrophy and visual disturbances. The worsening of these features can be a big socio-cultural stigma for a child as he or she is then vulnerable to frequent teasing by his or her peer group.[1] Therefore, surgeons advise early intervention to correct these craniofacial deformities in early childhood, and to eliminate clinical and socio-cultural problems.

The most challenging aspect in a case of Crouzon syndrome is airway management during any surgery performed under general anaesthesia. Use of neuraxial anaesthesia may circumvent the problems that Crouzon syndrome presents, but may be difficult to carry out, owing to vertebral fusion and the presence of scoliosis.[9,10] Sciatic and femoral nerve blocks can be used for lower-limb surgery, but our lack of expertise was the sole reason that the use of neuraxial anaesthesia was selected. Scoliosis and spinal canal stenosis can also occur in association with acanthosis nigricans in patients with Crouzon syndrome.[9] In the present case, surgery of a lower limb was performed, so subarachnoid block was an ideal option. However, the limitations imposed by the mental retardation were a worrying feature, and hence an intubation trolley was prepared in anticipation of possible conversion to general anaesthesia.

It is always difficult to counsel patients with mental retardation on regional anaesthesia, because of the need for patient cooperation and an understanding of the implications of general anaesthesia, both on the part of the anaesthesiologist and the patient. In this case, the parents had to be involved, because they were the best persons to help this patient with mental challenges to cooperate with the planned clinical procedures. The strange atmosphere of the operation theatre, the masked faces of personnel, and the sound and sight of instruments and equipment can be disconcerting to patients. Administering sedation to assist with this is fraught with danger, as it may lead to respiratory obstruction.[11,12] It was only after a long session of counselling, and final assurance by the parents, that we were able to take the patient to the operation theatre.

Conclusion

To conclude, anaesthetic management is very challenging in patients with Crouzon syndrome, especially when airway management is considered. Neuraxial anaesthesia is not a guarantee of success. Very careful planning should be carried out before the procedure, and alternative plans should be in place for reversion to either general or regional anaesthesia.

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