Case Report

Anesthetic management of craniosynostosis repair in a 13-month-old boy with Apert syndrome

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ABSTRACT

Apert syndrome is a congenital disorder characterized by malformations of the skull, face, hands, and feet. It is a form of acrocephalosyndactyly. It is classified as a branchial arch syndrome, affecting the first branchial (or pharyngeal) arch, the precursor of the maxilla and mandible. Presently described is a case of 13-month-old boy diagnosed with apert syndrome. French pediatrician Eugène Apert described Apert syndrome in 1906. The purpose of this report is anesthetic management of craniosynostosis repair in this child under general anesthesia. Craniosynostosis, the premature fusion of skull sutures, results in failure of normal bone growth perpendicular to the suture and the compensatory growth at other suture sites, which results in an abnormally shaped head. Anesthetic challenges include management of a difficult airway, airway hyperreactivity, possibility of raised intracranial pressure, blood loss, blood transfusion, and its associated complications.

Keywords: Acrocephalosyndactyly, Apert syndrome, Anesthetic challenges, Craniosynostosis, Syndactyly

Apert syndrome is caused by a mutation in the FGFR2 gene which has a role in tissue repair and embryonic development [1]. Mutation in this gene leads to an increase in the signals that promote bone formation. The key feature of this syndrome is premature closure of the bones of the skull known as craniosynostosis. This early fusion prevents the skull from growing normally leading to an abnormal shape of the head and face. It has been associated with varying degrees of mental insufficiency. The patients can have webbed or fused fingers and toes (Syndactyly) [2]. Apert syndrome is a rare disease and is estimated to occur in 1 in 65,000–200,000 births depending on the study cited, and males and females are equally affected [3]. The rationale of this case report is to describe a 13-month-old boy penned for cranioplasty and anesthetic challenges associated with it.

CASE REPORT

A 13-month-old boy presented with symmetrical syndactyly of both hands and feet and abnormal head shape. Family history was inquired and both the parents were normal. The mother had a full-term normal vaginal delivery. She had no history of any infection or drug use during pregnancy. Developmental milestones were according to his age. Vaccination was complete as per schedule. Congenital heart diseases were ruled out. The patient had a history of upper respiratory tract infection (URTI) 7 days before surgery and at that time, the total leukocyte count (TLC) was raised to 16,000/ml. The child was never operated in the past.

General examination revealed abnormal skull shape and fused fingers of the hands and feet (Fig. 1). Pupils were normal on examination. Systemic examination was normal and the body weight was recorded to be 10 kg. Mallampati grading could not be assessed. The mouth opening was adequate. Few teeth were present. The patient had a high-arched palate. Investigations were within normal limits and the TLC had come to normal on the day of surgery. The patient was posted for cranioplasty for craniosynostosis.

Corrective surgery presented with many anesthetic challenges that included the management of a difficult airway, airway hyperreactivity, possibility of raised intracranial pressure, hypothermia, blood loss, transfusion, and its associated complications. A difficult airway cart containing Guedel oropharyngeal airway, endotracheal tubes cuffed and uncuffed 2.5, 3.0, 3.5, 4.0, and 4.5 ID, stylets, conventional laryngoscope with straight Miller's blades #1 and 2, manual resuscitation bag, suction catheters, Magill's forceps for pediatric age group, etc.

The patient was received in the operation theater with an intravenous line *in situ*. Premedication was done with glycopyrrolate and midazolam. Intravenous fentanyl and

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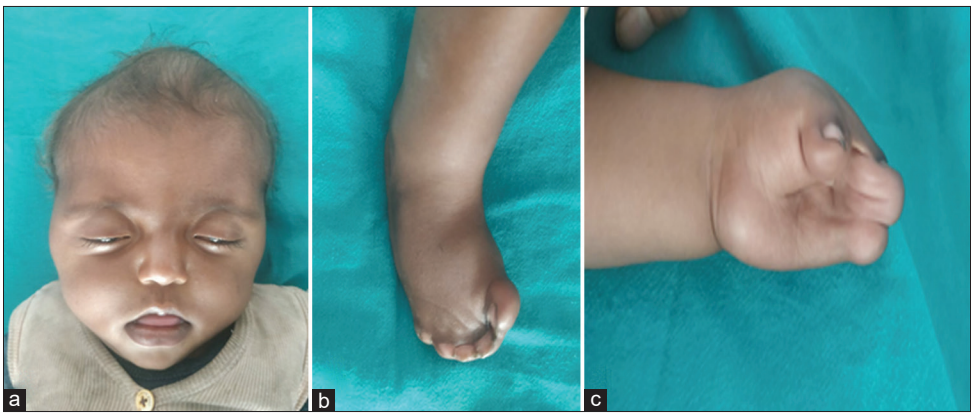

Figure 1: (a) A 13-month-old boy with Apert syndrome showing syndactyly of (b) toe and (c) hand

paracetamol suppository were used as analgesics. Induction was done with inhalational sevoflurane combined with intravenous thiopentone. After assuring adequate bag and mask ventilation, the patient was given succinylcholine and was intubated with a 4 mm microcuff endotracheal tube. Video laryngoscopy-assisted intubation was done with a styleted endotracheal tube. An injection atracurium loading dose was given. Oxygen, nitrous oxide, sevoflurane, atracurium, and fentanyl were used for maintenance.

The child was undergoing bronchospasm intermittently leading to difficult ventilation which was managed with dexamethasone and hydrocortisone plus secretion removal (suspected to be a result of reactive airway since there was a history of URTI a week ago). The patient suffered a blood loss of approximately 300 ml which was replaced with packed red blood cells. Fresh frozen plasma was also transfused intraoperatively. The patient remained hemodynamically stable intraoperatively. Hyperventilation was done intermittently to prevent raised intracranial pressure. Near completion of the surgery, injection phenytoin intravenously was given to prevent post-operative convulsions. After the completion of the surgery, the patient was assessed and on ensuring that the patient was fully awake and the trachea was extubated (Fig. 2).

Follow-up of the patient was done and there was a slight swelling in the initial post-operative period which subsided on its own. The laboratory tests were normal and the patient was fine when discharged. Further follow-up showed healed wound after suture removal.

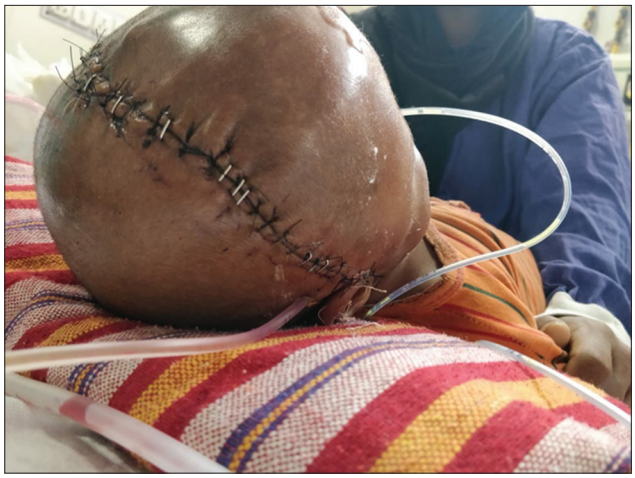

Figure 2: Corrected craniosynostosis

DISCUSSION

Apert syndrome is an inherited craniosynostosis syndrome and autosomal dominant in nature. Both the genders are equally affected. The incidence of the disease significantly increases with increasing paternal age [4].

People born with this syndrome may have problems with their vision and teeth because of the abnormal shape of the facial and skull bones. The head is usually long, with a high forehead. Eyes are often wide-set and bulging with poorly closing eyelids with a sunken middle face. Life expectancy is normal; however, some of the health problems associated with the syndrome can lead to complicated disease and premature death. The facial bones can be affected by craniosynostosis. This can lead to characteristic facial abnormalities. Palate abnormalities such as cleft palate may be found. The right and left sides of the face may not be symmetrical. They can have moderate-to-severe acne. Individuals can have URTI, sleep apnea, and malnutrition.

Apert syndrome has several hand and foot malformations characterized by short fingers and broad thumbs and great toes that deviate outward, partial to complete fusion (syndactyly) of certain fingers and toes or complete fusion of the bones of the second to the fourth fingers, and one single, continuous nail known as characteristic mitten-like syndactyly. A multidisciplinary approach is usually required to deal with these patients.

Anesthetic challenges include management of a difficult airway, potentially difficult IV access, airway hyperreactivity (risk of bronchospasm), possibility of raised intracranial pressure, hypothermia, blood loss, blood transfusion, and its associated complications, and the long duration of surgery [5]. Intravenous access could have been a difficulty due to syndactyly. However, this was not the case in this child. Barnett *et al*. found there to be a low incidence of major perioperative major complications in this group of patients. Nevertheless, a significant proportion of these children has obstructive sleep apnea and may develop supraglottic airway obstruction on induction and emergence from anesthesia due to the associated mid-face anatomical abnormalities [6].

Kumar *et al*. successfully managed a case of a 5-year-old child with Apert syndrome posted for craniosynostosis and paid special attention to the airway as well as heat loss during the surgery using warm blankets as a large area was exposed during surgery [7]. These patients are vulnerable to develop hypothermia as significant heat loss occurs through the exposed head and blood loss. Moreover, patients with Apert syndrome are known to have excessive sweating. Covering of the extremities raised operation theater temperature, warm air blankets, and fluid warmers should be employed throughout the procedure [8].

Atalay *et al*. reported two cases of Apert syndrome and the anesthetic challenges related to that. Anesthetists must be ready for airway problems, intubation difficulties, and even visceral anomalies. In particular, Apert syndrome with visceral anomalies may cause significant problems for anesthesia and surgery [9]. Intraoperative airway complications such as bronchospasm and wheezing are present to a greater degree than the normal population as complete or partial tracheal cartilage sleeve abnormalities lead to accumulation of secretions, decreased tracheal distensibility, and increased injury during suctioning. Thus, suctioning is to be done carefully, and greater depth of anesthesia is to be maintained during airway stimulation [10]. Eyes should be well lubricated as these patients have improperly closing eyelids.

There are chances of hypovolemia due to blood loss and, hence, the availability of adequate volumes of typed and cross-matched blood products must be ensured beforehand [11]. A multidisciplinary approach with the help of a neurosurgeon, plastic surgeon, ophthalmologist, geneticist, and psychiatrist is generally required for these cases.

CONCLUSION

Craniosynostosis is a complex condition presenting a challenge for anesthesiologists as well as surgeons complicated by the young age of patients, associated syndromes, high incidences of intraoperative complications, and demands strict intraoperative and post-operative care. We were able to successfully manage this case of Apert syndrome because of the background knowledge of associated anomaly and anticipatory precautions for expected neurosurgical as well as non-neurosurgical complications.

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