Anesthetic management of craniosynostosis repair in patient with Apert syndrome

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ABSTRACT

Apert syndrome is an autosomal dominant disease characterized by craniosynostosis, midface hypoplasia and syndactyly. In general, patients present in early childhood for craniofacial reconstruction surgery. Anesthetic implications include difficult airway, airway hyper-reactivity; however, possibility of raised intracranial pressure especially when operating for craniosynostosis and associated congenital heart disease should not be ignored. Most of the cases described in literature talk of management of syndactyly. We describe the successful anesthetic management of a patient of Aperts syndrome with craniosynostosis posted for bicornual strip craniotomy and fronto-orbital advancement in a 5-year-old child.

Key words: *Acrocephalosyndactyly, anesthesia, Apert syndrome, craniosynostosis*

INTRODUCTION

The incidence of Apert syndrome is about 15/1,000,000 live births[1] and with rare reports from India.[2] It is an autosomal dominant syndrome that presents with a combination of craniosynostosis, syndactyly and midfacial hypoplasia. It may present a number of challenges to the anesthesiologist such as; a difficult airway, difficult intravenous (IV) access, airway hyper-reactivity, temperature dysregulation etc. Varying degrees of mental deficiency have been associated with Apert syndrome. Individuals who have undergone craniectomy in early life may have improved intelligence. These children may come for craniofacial remodeling, which are high blood loss surgeries or relatively minor procedures such as syndactyly release. In this case report, we present a 5-year-old boy with Apert syndrome who presented to our institute for bicornual strip craniotomy and fronto-orbital advancement for craniosynostosis.

CASE REPORT

A 5-year-old child with a history of enlarging head since birth presented to our facility. He was born by term normal vaginal delivery and no perinatal complications. He also complained of fused digits in both upper and lower limbs. He had delayed milestones. He had no history of seizures, weakness of any limbs or bowel and bladder incontinence. He had a history of two episodes of pneumonias at 2 years of age none of which required any intubation or mechanical ventilation. He had no history of cyanosis, jaundice or known cardiac disease. He had never been operated on in the past. There was no history of upper respiratory tract infection. Patient did give a history of snoring, but there was no history suggestive of obstructed sleep apnea.

On examination, he was found to have brachycephally with open anterior fontanelle. The child also had proptosis and xerophthalmia [Figure 1]. The pupils were normal and reactive to both light and accommodation. He had fusion of all the digits in all the four limbs [Figure 2]. Airway was modified mallampati Grade 2 with adequate mouth opening and neck movement. The child had high arched palate; teeth were intact, but mottled and discolored. On running blood investigations, he was found to have hemoglobin of 9.9 g/dl and the rest of the investigations were within the normal limit.

Patient was premedicated with 200 μg of oral atropine ½ h before shifting to the operating room. When the patient was brought to the operating room, he already had an IV line *in situ*. The patient was induced with fentanyl 40 μg, propofol 50 mg and on establishing adequate mask ventilation 20 mg of rocuronium was given. The patient was intubated with 5.0 sized cuffed endotracheal tubes,

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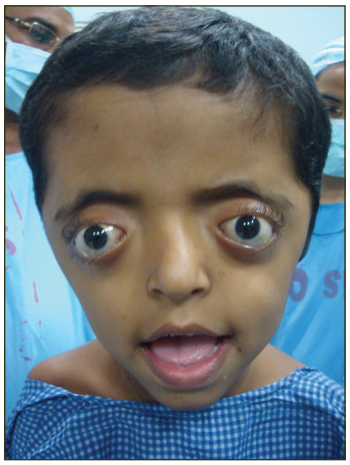

Figure 1: Craniosynostosis, midfacial hypoplasia and proptosis

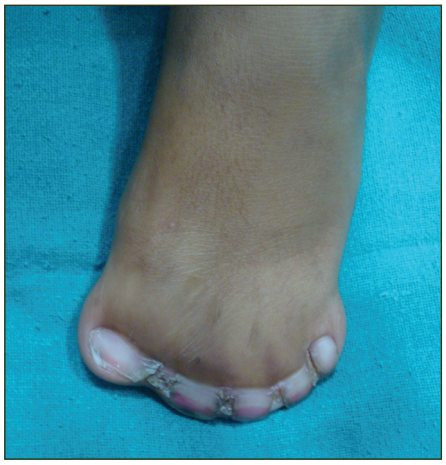

Figure 2: Syndactyly

which was fixed at 14 cm. The Cormack-Lehane view was Grade 1. Right internal jugular vein was cannulated with 5Fr triple lumen. Right radial was also cannulated with 20G cannula for invasive blood pressure monitoring. The anesthesia was maintained with oxygen, nitrous oxide, sevoflurane, fentanyl and vecuronium. A total volume of 100 ml of 20% mannitol was also administered. The patient maintained an intraoperative temperature of 36-36.2°. The patient suffered a blood loss of hardly 200 ml which was replaced with packed red blood cells. The patient was hemodynamically stable maintaining a systolic blood pressure of 90-110 mmHg. After the completion of surgery, the patient was assessed and on ensuring that the patient was fully awake trachea was extubated.

DISCUSSION

Apert syndrome is a rare autosomal dominant disorder also known as acrocephalosyndactyly caused by the mutation of the fibroblast growth factor 2 gene. It consists of 4.5% of all cases of craniosynostosis with equal male to female ratio. Because of the premature fusion of the skull bone, characteristic facial features arises such as craniosynostosis, midfacial hypoplasia, hypertelorism and decreased nasopharyngeal diameter. Intellectual development can be disrupted because of early fusion of the skull bones, which affects the development of the brain. A number of visceral anomalies are associated with this syndrome, most common of which are the cardiovascular anomalies, which are present in 10% of the cases. Genitourinary anomalies such as hydronephrosis and cryptorchidism occur in 9.6% of the cases.[3] Since our patient did not give any history of cyanosis, fatigue or recurrent chest infections and had a normal electrocardiogram, hence further cardiac investigation in the form of echocardiography was not warranted in this case. A multidisciplinary care is required for perioperative management of these patients.

The perioperative concern in such patients include potentially difficult airway, difficult IV access, risk of bronchospasm, eyes related, hypothermia, blood loss, venous air embolism and long duration of surgery. One of the major concerns of a patient of Apert syndrome is potential difficult airway. Obstructive sleep apnea syndrome is common in patients of Apert syndrome. It may be attributed to the nasopharyngeal malformation found in these patients.[4,5] Although in infants lower airway pathology is primarily responsible for it, the upper airway especially the pharynx takes the blame for the same in adults. Cervical spine anomalies occur in 71% of these patients and mainly consist of complex fusion of the spine at C5-C6 level.[6] This however, is of little consequence in intubation of these patients as positioning for intubation involves the movement at upper cervical spine levels. Because of abnormal facial features achieving a good mask seal may be difficult therefore different sizes and types of masks should be available. Our patient though had a high arched palate, did not have any difficulty in masks ventilation or intubation. Barnett *et al.*, in a series of 509 anesthetics in patients with Apert syndrome reported that although the incidence of respiratory complications was low in this group of patients (6.1%), a significant proportion of this was constituted by supraglottic airway obstruction at the time of induction and the emergence from anesthesia.[7] Bronchospasm is also more common in these patients as compared to rest of the population. In a study by Elwood *et al.*, they could not demonstrate any benefit from pre-operative administration of nebulized albuterol. As compared to normal patients more attention is to be needed during tracheal suctioning and deeper level of anesthesia is required during airway stimulation.[8] The higher incidence of airway complications in this population could be related to the fact that these children suffer from tracheal anomalies in the form of complete or partial cartilage sleeve abnormalities which may lead to accumulation of secretions

leading to bronchospasm, lack of tracheal distensibility and tracheal injury due to increased suctioning.[9,10] In fact there is a case report where obstruction of an endotracheal tube due to secretions resulted in a sudden build-up of airway pressure requiring a replacement of the tube.[10] We had premedicated our patient with atropine prior to the surgery in order to avoid such a complication. Furthermore, we kept bronchodilators ready and kept a constant watch on the airway pressure, in order to catch any rise in airway pressure. Since this syndrome is associated with syndactyly, IV access is expected to be difficult. However, in our patient we found no such difficulties.

There is always a risk of hypothermia in these patients as they are known to have excessive sweating. In addition to that in craniosynostosis surgery a large surface area is exposed to the atmosphere, which may result in excessive heat loss. We maintained the temperature of our patient by using warm fluids and warming blankets. Lubrication and proper care of eyes should be done as there is inadequate lid closure in these patients. Craniosynostosis repair challenges include severe blood loss, risk of venous air embolism. Blood loss was anticipated and hence an invasive arterial blood pressure monitor and central venous pressure monitor were included in the monitoring.

CONCLUSION

We were able to successfully manage this case of Apert syndrome because of background knowledge of associated anomaly and anticipatory precautions for expected neurosurgical as well as non-neurosurgical complications. We suggest that Anesthesiologist should be aware of the clinical features of Apert syndrome and all involved organs ensuring appropriate monitoring for complication. A thorough assessment of the individual is crucial for developing a safe anesthetic plan for such patients.

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