CASE REPORT

A Case of Ectrodactyly–Ectodermal Dysplasia–Cleft Syndrome: An Anesthetic Challenge

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Abstract:
Ectrodactyly–ectodermal dysplasia–cleft syndrome, or EEC syndrome (2) is a rare genetic disorder characterized by the triad of ectrodactyly, ectodermal dysplasia, and facial clefts. Affected patients frequently require repeated surgeries, especially in childhood. These abnormalities often complicate the peri-opertaive course but very little has been published about the anesthetic management. In this case report we present the anesthetic management of this rare syndrome in a 2 year old child posted for cleft palate repair.

Keywords: congenital syndrome, cleft palate, periop management.

Introduction
Pediatric airway management is always a challenging task and it becomes more challenging in defects like cleft lip and palate. Cleft lip/palate can occur either alone but it is often associated with other congenital defects (1). One of these complex syndromes is EEC Syndrome. Ectrodactyly–ectodermal dysplasia–cleft syndrome, or EEC syndrome (2) is a rare genetic disorder characterized by the triad of ectrodactyly, ectodermal dysplasia, and facial clefts. Affected patients frequently require repeated surgeries, especially in childhood. These abnormalities often complicate the peri-opertaive course but very little has been published about the anesthesia management. In this case report we present the anesthetic management of this rare syndrome in a 2 year old child posted for cleft palate repair.

Case Report
A 2yr old female weighing 10 kg was brought to our hospital with complaint of defect in palate and fused digits of all four limbs since birth. On examination she had a cleft palate, syndactyly of all four limbs, dry skin, sparse scalp hair, brittle nails and grade 2 malnutrition. Her mouth opening was adequate, and the modified Mallampati classification could not be assessed as child was uncooperative. Rest examination was within normal limits. She was diagnosed as a case of EEC syndrome and was posted for cleft palate repair under general anesthesia. Patient was crying and uncooperative hence 50 mg ketamine with 0.2 mg glycopyrrolate intramuscularly was given in the preoperative area in mother’s lap. Once sedated the child was carried into operation the later, and standard monitors including pulse oximetry, non-invasive blood pressure, electrocardiogram and precordial stethoscope were attached. Intravenous line was secured with 22 G intracath inside OT and baby was kept on mask. Patient was medicated with Midazolam 0.03 mg/kg and Fentanyl 2 mcg/kg. Preoxygenation with 100% oxygen for 3 min and then was induced with sevoflurane 8% in 60:40 N2O:O2. Tracheal intubation was done with 5 uncuffed Endotracheal tube using Airtaq. Air entry on both sides confirmed, then tube was fixed with proper sticking and throat packing was done. Maintained with 60% N2O, 40% O2 and 2 % sevoflurane and intermittent injection atracurium. Temperature monitoring was done throughout the procedure via axillary temperature probe. Cleft palate defect was closed by correcting position of muscles of soft palate and reconstruction of muscle sling. Hemodynamic parameters were stable intraoperatively. Surgery lasted for one and a half hours and after confirming haemostasis, throat packs removed. At the end of surgery neuromuscular blockade was reversed with Neostigmine 0.04 mg/kg and
Glycopyrolate 0.01 mg/kg. After adequate reversal, patient was extubated, oxygenation continued and patient observed for 15 minutes in operation theatre. Post operative nebulisation with 0.9% NS was done.

Discussion
Ectrodactyly–ectodermal dysplasia–cleft syndrome, or EEC, and also referred to as EEC syndrome\(^2\) (also known as "Split hand–split foot–ectodermal dysplasia–cleft syndrome")\(^3\) is a rare congenital anomaly complex characterized by three cardinal signs of ectrodactyly and syndactyly of the hands and feet, cleft lip/palate and abnormalities in several ectodermal structures including skin, hair, nails and exocrine glands (reduction/absence of sweat, sebaceous and salivary glands)\(^4\). Autosomal dominant inheritance has been suggested. Exact incidence of this syndrome is not known but about 200 cases have been described in literature\(^5\). Affected patients frequently require repeated surgery, especially in childhood. These abnormalities often complicate the peri-operative course. The main anesthetic concerns in these patients are potential difficult airway, difficult intravenous access, malnutrition, difficulty with control of body temperature related to hypohidrosis and persistent infection of the respiratory tract\(^5\).

The following problems were encountered in our patient:
1. Syndactyly with dry and brittle skin leading to difficulty in securing intra venous access.
2. Cleft palate causing a potential difficult airway. Intubation was facilitated in this patient with the help of an airtraq and bridging the cleft with a gauge pack.
3. Difficult intraoperative temperature control due to impaired heat loss with hypohidrosis secondary to hypoplasia of the sweat glands. Continuous temperature monitoring was done intraoperatively and adequate measures were taken to prevent hypothermia.
4. Combination of malnutrition and a thin, delicate skin makes them prone to pressure ulcers. Extreme care was given in the form of proper cotton padding at pressure points.
5. Eyes were protected with ointment and then eye padding due to decrease tear production in these patients.
6. Post operative nebulisation with 0.9% NS done to improve mucocilliary clearance.

Syndromes which are associated with ectodermal dysplasia have multiple complications and manifest themselves in many different ways. We successfully managed the intraoperative problems in this rare genetic disorder.

Conclusion
Patients with EEC syndrome may present for various types of surgeries like cleft lip/palate repair or syndactyly correction. Intraoperative management of these patients is often a challenging task for an anesthesiologist. It is of utmost importance for the anaesthesiologist to understand the common complications which one can encounter in these patients and method to manage them.

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References