Case Report

Lamellar Ichthyosis; An Uncommon Acquaintance with Anesthesiologists: Overcoming Barriers

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Abstract

Lamellar ichthyosis constitutes a distinct group of inherited ichthyosis characterized by excessive scaling and dryness. Abnormalities in epidermal differentiation and metabolism contribute to excessive keratinization, which is a hallmark of this disorder. We present here the successful anesthetic management of a 13-year-old boy with lamellar ichthyosis for the correction of Genu Valgum deformity.

Key words: Hyperkeratosis, lamellar ichthyosis, skin scaling

Introduction

Ichthyosis refers to a relatively uncommon group of skin disorders characterized by the presence of excessive amounts of dry surface scales. It is regarded as a disorder of keratinization or cornification, caused due to abnormal epidermal differentiation or metabolism.

Five distinct types of inherited ichthyosis are noted, namely;

- Ichthyosis vulgaris,
- Lamellar ichthyosis,
- Epidermolytic hyperkeratosis,
- HarlequinIchthyosis, and
- X-linked ichthyosis.

The most common cause of lamellar icthyosis is inactivating mutations in the TGM1 gene, which encodes transglutaminase-1 (TGase1), a key enzyme that cross-links proteins in the cornified cell envelope of the dermis. It presents significant challenges to anesthesiologists in view of difficult intravenous access, airway problems, monitoring, malnutrition etc. Only negligible number of cases of lamellar icthyosis with genuvalgum for deformity correction has been reported for anaesthetic management Hence we like to share our experience and views about it.

Case History

A 13-year-old boy presented to the Orthopaedic department with bilateral knee deformity, which was diagnosed as Genu Valgum, and was advised to undergo bilateral "8 Plate" application.

Patient had complaints of skin peeling since birth, and on further probing, it was noted that the child was born out of a second degree consanguineous marriage. Family history of a similar disorder was seen to be present in a distant relative of the patient.

On examination of the skin, patient had fish-like ichthyotic scales on the face, trunk, and limbs with more scaling present in flexural areas [Figure 1].

Ectropion was present in the left upper eyelid for which patient was applying topical lubricant eye drops [Figure 2]. Nails and teeth were normal. Dermatologist opinion was sought and he was diagnosed to have congenital ichthysioform erythroderma/ lamellar ichthyosis. An ophthalmologist's opinion was sought for the ectropion, and a trial of oral retinoid was planned on being given after the correction of bony deformities. Patient was put on liquid paraffin and petroleum jelly topical application for the skin. Dental opinion was also taken to rule out tooth problems.

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Pre-anesthetic evaluation

Birth history revealed that child was born full term and was delivered by lower segment caesarean section. Developmental history was normal for age. Airway examination showed mouth opening of more than two fingers. Neck extension was restricted to some extent due to extensive scaling and contractures. Modified Mallampatti Scale was 2. Routine blood tests were found to be within normal range. Child weighed 25 kg.

Anesthetic management

Patient's baseline heart rate was 95/min, SpO_2 was 98%, noninvasive blood pressure was 110/66 mmHg. Electrocardiogram (ECG) electrodes were placed at places of minimal scaling. Intravenous access was established with 22-gauge cannula and was secured to the skin with tegaderm [Figure 3]. Patient was induced with Propofol 50 mg and Fentanyl 50 µg. Eyes were covered with microporeplaster after applying his regular eye drops. Scales over the lower jaw were covered with gauze during bag and mask ventilation [Figure 4].

After confirming ability to ventilate with bag and mask, non-depolarizing muscle relaxant injection Atracurium 12.5 mg was given. Patient was intubated with 6.0 mm cuffed oral endotracheal tube. Endotracheal tube was fixed with a roller bandage at 18 cm [Figure 5].

Anesthesia was maintained using Isoflurane 1–1.5%; dial reading and nitrous oxide–Oxygen mixture was 50–50%. Patient was covered with Gamzee rolls and warmer was kept at 38°C to prevent hypothermia. In addition to opiods, the child also received intravenous Paracetamol for analgesia. Intraoperative period was uneventful. At the end of procedure, meuromuscular blockade reversed with Neostigmine 1.25 mg along with Glycopyrolate 0.25 mg. Patient was extubated and shifted to the recovery room. The postoperative period was uneventful.

Discussion

Lamellar ichthyosis and congenital ichthyosiform erythroderma are the most common types of autosomal recessively inherited ichthyosis.[1]

Both forms present at or shortly after birth. Most infants with these forms of ichthyosis present with erythroderma and scaling. Six genes have been identified that cause non-Harlequin ichthyosis.

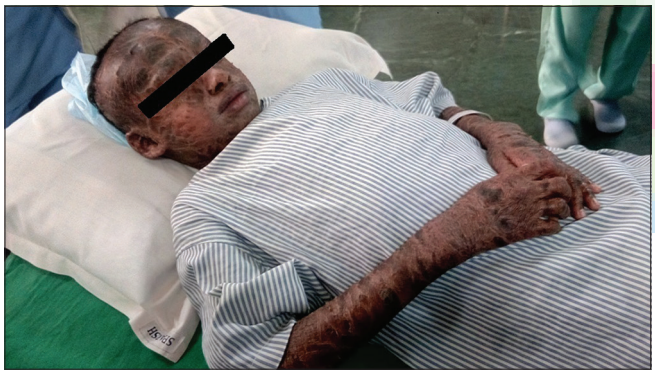

Figure 1: Extensive scaling and keratinization

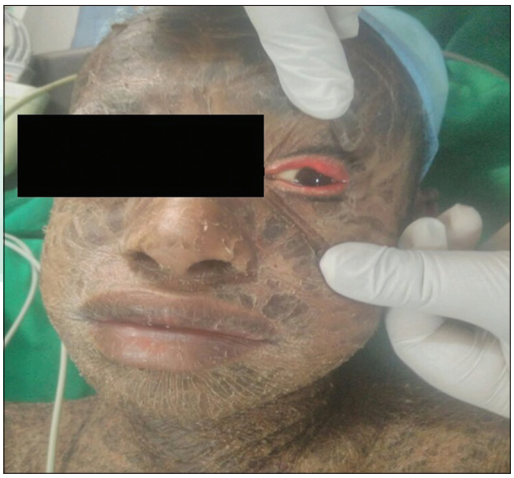

Figure 2: Visible ectropion

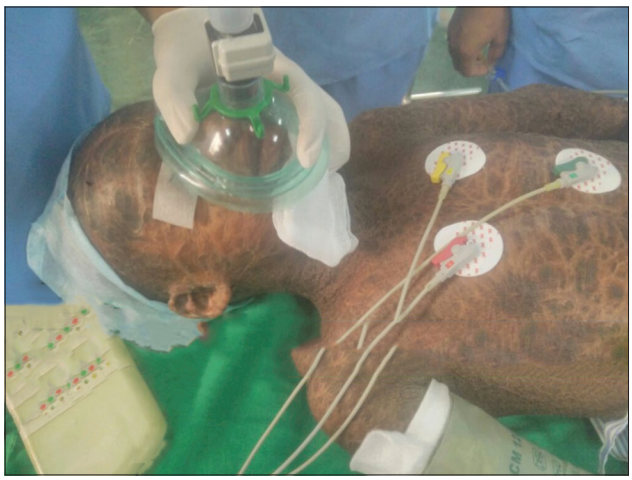

Figure 3: Protective gauze over arm

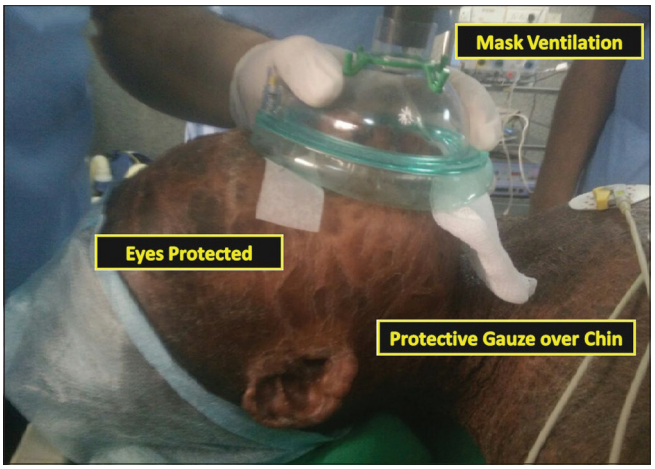

Figure 4: Mask, protected eyes, and chin

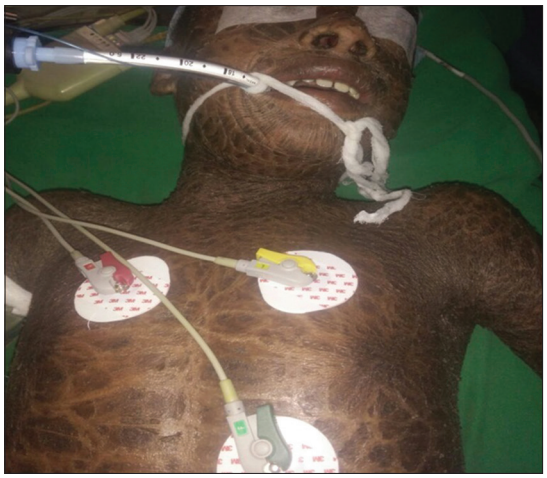

Figure 5: Secured endotrachel tube

TGM (The gene encoding transglutaminase) mutations[2] lead to abnormalities in the cornified envelope, whereas defects in *ABCA12* cause abnormal lipid transport and those in *CYP4F22* produce abnormal lamellar granules. The lipoxygenases are likely to play a role in epidermal barrier formation by affecting lipid metabolism.

The estimated incidence of lamellar ichthyosis is 1:200,000 ± 300,000.[3] Scaling is pronounced and involves the entire body surface, including flexural surfaces. The face is often markedly involved, including ectropion and small crumpled ears. The palms and soles are generally hyperkeratotic. The hair may be sparse and fine, however, the teeth and mucosal surfaces are normal. There is little erythema, and hyperkeratosis is particularly noticeable around the knees, elbows, and ankles. Palms and soles are uniformly hyperkeratotic. Patients have sparse hair, cicatricial alopecia, and nail dystrophy.[4] Pruritus may be severe at times and responds minimally to antipruritic therapy.

The unattractive appearance of the child and bad odour from bacterial colonization of macerated scales may create serious psychological problems. A high-humidity environment in winter and air conditioning in summer reduce discomfort. Generous and frequent applications of emollients and keratolytic agents such as lactic or glycolic acid (5–12%), retinoic acid (0.1% cream) may lessen the scaling to some extent. Oral retinoids (1 mg/kg/day) have a beneficial effect in these conditions but do not alter the underlying defect.[5]

The long-term risks of these compounds (teratogenic effects and toxicity to bone) may limit their usefulness. Ectropion requires ophthalmologic care, and at times, plastic surgery procedures.

Perioperatively, tubes and catheters need to be sewn or tied into place. Peripheral intravenous access may be difficult. In severe ichthyosis, temperature regulation is impaired. Cremers *et al*. observed early childhood deafness, congenital nonbullous ichthyosiform erythroderma, corneal involvement, photophobia, hypotrichosis, anhidrosis, hyperkeratosis of the nails and dental dysplasia.[6]

Management of ichthyosis is multidisciplinary involving genetic counselling at early stage along with coordinated support from pediatrician, dermatologist, ophthalmologist, otorhinolaryngologist, orthopedic surgeon and a psychiatrist.[7]

In this patient, Intravenous access on the dorsal part of the hand was secured with Tegaderm instead of normal adhesive plasters, which do not stick well on hyperkeratinized skin, and any attempt to increase the adhesiveness may be met with difficulty during removal. ECG electrodes were placed at places of minimal scaling as electrodes of an ECG may not stick well and there may be poor quality of signal because of reduced conductivity.[8] Endotracheal tube was secured using a gauze roll tied around the face. Ideally a tube holder should be used to secure the endotracheal tube.

Warming measures such as use of warm intravenous fluids, use of bair hugger, adjustment of operating room temperature, and covering the child with warm blankets were done. These patients are susceptible to develop perioperative hypo as well as hyperthermia because of the abnormal keratinization. Various eye protective measures were instituted such as application of lubricant eye drops and covering the eyes with micropore plaster. Patients are susceptible to eye injuries due to direct trauma as well as exposure because of the ectropion and inability to close the eyes completely.

Caudal analgesia was avoided due to the presence of excessive skin flakes and skin excoriation at the site of sacral hiatus. Extensive literature search revealed only few articles and case reports pertaining to anaesthetic management of lamellar icthyosis for the correction of Genu Valgum deformity.

Conclusion

Successful perioperative management of case of lamellar ichthyosis for various surgeries calls for meticulous planning and skilled anesthetic management particularly with reference to airway management, venous access, monitoring, and maintenance of normothermia. Complete management of case of lamellar ichthyosis is often multidisciplinary task involving close coordination and communication among various disciplines of caregivers.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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