Anaesthetic management of a rare case of paediatric epidermolysis bullosa

Sir,

Epidermolysis bullosa (EDB) is a rare hereditary disorder characterised by abnormal fragility of skin and mucosal surface which leads to a life-threatening medical condition by blister formation.[1] The three common types of this condition are EDB simplex, junctional EDB and dystrophic EDB, with EDB simplex being the most common type that has intraepidermal blistering with mucosal involvement. Junctional EDB presents with severe blistering at birth and most of patients die during infancy. Dystrophic EDB involves the dermis. Perioperative care of EDB has to be addressed uniquely as it requires special attention toward monitoring, careful positioning, scrupulous asepsis, perioperative pain relief and averting skin and mucosal trauma. Case of penetrating keratoplasty (PK) in EDB simplex patient as young as 6 months of age has not been reported.

A 6-month-old female patient, a known case of EDB simplex since birth, of The American Society of Anesthesiologists (ASA) physical status II was posted for PK and lensectomy of the right eye. She was malnourished, weighed 3 kg and had growth retardation due to the poor oral intake resulting from oral ulceration. Multiple skin erosions, numerous bullae, widespread blisters and healed scars were present on the body [Figure 1]. Airway examination revealed tongue attachment to the floor of the mouth with involvement of the cheek and perioral area. However, the involvement of the pharynx or trachea could not be visually confirmed. The results of all laboratory investigations were within normal limits.

Anaesthesia was induced with sevoflurane 4–8%. Face mask was held gently and care was taken to prevent injury to the contralateral eye, including application of moisturizing ophthalmic ointment and moistened protective gauze and lubricating the fingers of the anaesthesiologist and the cushion of the mask with liquid paraffin. Sterile gloves were used during airway management. Although all limbs had extensive skin lesions, intravenous access could be obtained on the right leg. Pulse oximetry was applied intermittently over different fingers, and paraffin gauze was applied under the blood pressure cuff to prevent trauma from friction. For the electrocardiogram (ECG) monitoring, the adhesive peripheral part of the ECG electrode was removed, leaving only the gel portion which was secured by placing cotton pads to prevent any mechanical injury. Minimal pressure for chin lift and head tilt was used for manual ventilation. After induction of anaesthesia, atracurium 1.5 mg was given to facilitate intubation with a 3.5 uncuffed tube using the Macintosh laryngoscope size 1 blade, which was lubricated with water-based jelly to prevent trauma. Anaesthesia was maintained with O_2:N_2O (40:60 ratio) and sevoflurane 1–1.5%, and fentanyl and atracurium iv were given in aliquots. A volume of 200 ml of crystalloids was infused over a period of 2 h. Following surgery, the neuromuscular blockade was antagonized, and the trachea was extubated after the patient was fully awake. She remained haemodynamically stable throughout the surgery. Postoperative analgesia was provided with intravenous paracetamol. Postoperative period was uneventful except for development of fresh bulla over left buccal mucosa which healed spontaneously over next 48 h. The patient was discharged on 7th postoperative day.

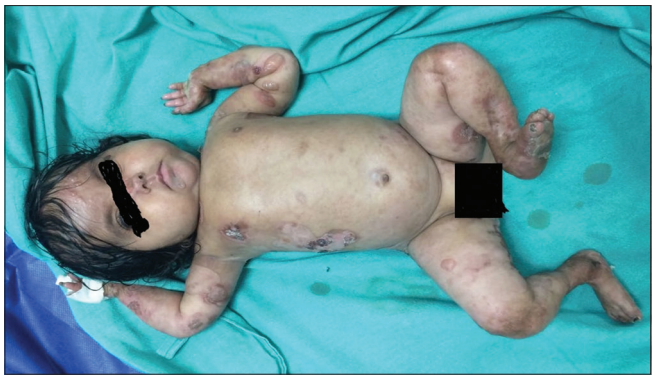

Figure 1: Multiple skin erosions, numerous bullae, widespread blisters and healed scars present on the body

The cause of malnutrition in our patient was due to decrease in the oral intake as a result of oropharyngeal and oesophageal lesions. Oral scarring with limited mouth opening, oesophageal stricture, anaemia and infection are also common, but rare in simplex type of EDB.[2] Nutritional deficiency leads to hypoproteinaemia, anaemia and electrolyte imbalance that may affect the pharmacokinetics and pharmacodynamics of anaesthetic agents. Infection is also common because of decreased immunity. Junctional type of EDB often has laryngeal and tracheal lesions. Possibility of airway complications such as oedema and obstruction due to blister and scars after extubation requiring emergent tracheostomy should be kept in mind.[3] Some authors reported that intubation might predispose the patient to formation of intraoral bullae, airway obstruction and extensive haemorrhage.[4] In our case, we performed intubation using the Macintosh laryngoscope with the blade well lubricated with water-based jelly to prevent trauma. Care should be taken during mask holding to avoid corneal abrasion. Changing the site of pulse oximeter probe frequently is helpful in preventing new onset lesions, to avoid burns, pressure erosion, skin necrosis and digital sensory loss.[5] Non-invasive blood pressure monitoring can be used over skin padded with paraffin gauze on manual mode to minimise friction. Utmost care was undertaken by us during mask ventilation to prevent injury to the contralateral eye, including application of moisturizing ophthalmic ointment and moistened protective gauze and lubricating the fingers of the anaesthesiologist and the cushion of the mask with liquid paraffin.

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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