Case Report
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A Case of Epidermolysis Bullosa Posted for Contracture Release and External Fixator Application of Hand under General Anaesthesia

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Abstract

Epidermolysis bullosa is a rare autosomal inherited disorder involving skin and mucous membrane, and characterized by blister and bullae formation all over the body. Meticulous attention needs to be given during perioperative period to ensure that no new bullae are formed and the existing ones are not disturbed. We present successful management of a case who presented for multiple procedures at different time intervals.

Keywords: Airway, Anaesthesia, Epidermolysis Bullosa, Skin

1. Introduction

Epidermolysis bullosa is autosomal inherited or acquired pediatric disease characterized by bullae formation in the skin or mucous membranes[1,2].

The reported incidence is about 20 cases per one million inhabitants[1].

The acquired forms are due to autoantibodies produced against basement membranes of skin and mucous membrane. Patients can have blisters in mouth resulting in potential difficult airway, multisystem involvement can result in anaemia, renal dysfunction and oesophageal dysmotility. The anaesthetic management of these patients can be challenging and requires meticulous care to avoid positional injuries[3].

2. Case Report

A 12 year old boy weighing 15 kgs, with history of Epidermolysis Bullosa and fusion of both hands and toes since 5 years presented for release of syndactyly. The patient had blisters all over the body since birth, syndactyly of all

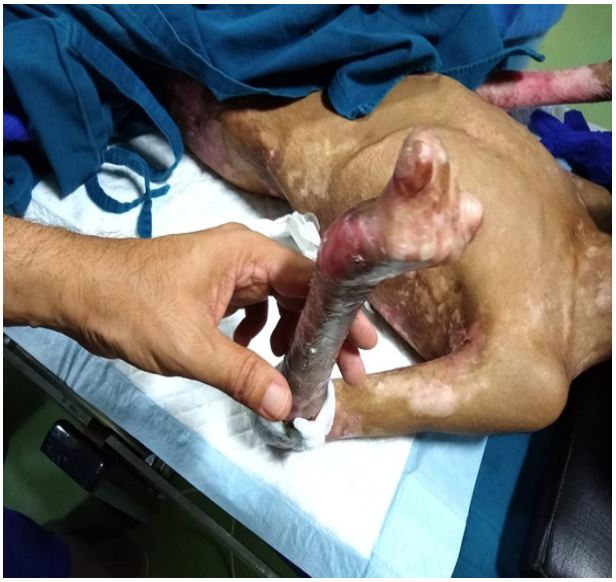

Figure 1. Image showing the contracture of elbow and syndactyly.

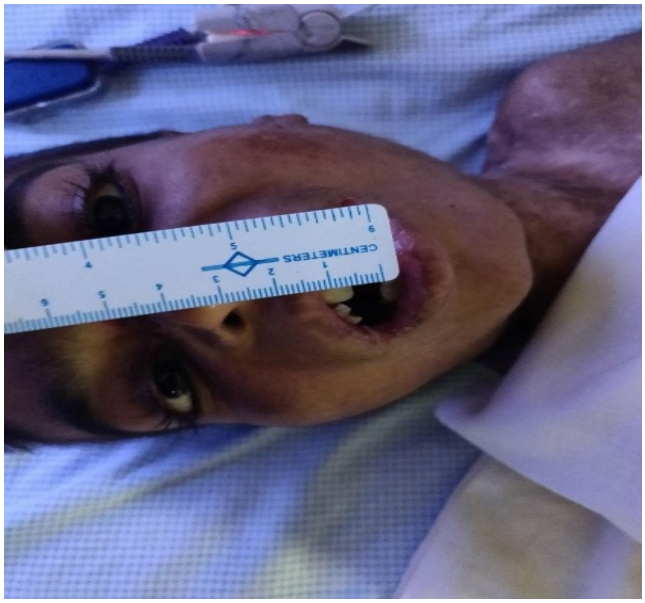

Figure 2. Mouth opening and airway parameters.

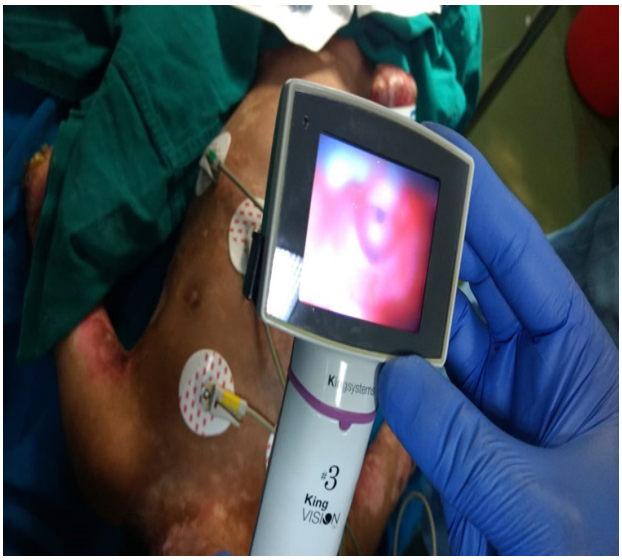

Figure 3. View of Larynx through video laryngoscope.

fingers of both hands and all toes of both feet contracture of elbow and knee, erythematous lesion in nape of neck and sacral area Figure 1. Airway assessment showed stained upper central incisors, restricted mouth opening and decreased neck movements Figure 2. Thyromental distance and jaw movements were normal.

Systemic examination was unremarkable. His laboratory investigations revealed HB - 7.3 gm%, S.Cr - 0.7 mg/dl Bl. urea - 20 mg/dl S. Bilirubin - 0.17 mg/dl Serology markers were negative.

General anaesthesia with endo-tracheal intubation and post operative routine monitoring was planned. After thorough pre anaesthetic evaluation, Patient relatives were explained about the risk of intubation, postion related problems and anaesthetic complications then consent was taken. Patient was kept fasting for 6 hours before induction of anaesthesia. Patient was shifted to operation theatre with all pressure area covered with cotton pad. In operation theatre patient placed in supine position NIBP connected to thigh by covering skin with cotton pad, SpO2 probe placed in left ear. Patient was pre-oxygenated and Premedicated with iv inj.glycopyrolate 0.1mg, inj.midazolam 0.5 mg, inj.fentanyl 30 mg. General anesthesia was induced with inj. Propofol 30 mg, and intubation facilitated with inj. Atracurium 7.5 mg. Patient was intubated with well lubricated 5.0 size cuffed endotracheal tube using kings vision video laryngoscope and endotracheal tube position was confirmed (Figure 3). Patient was ventilated with volume control ventilation. General anaesthesia was maintained with oxygen 50% and nitrous oxide 50% and sevoflurane 1% and inj. Atracurium bolus was given intermittently. Patient was positioned in supine, with adequate pads to protect eye and bony prominence. Intravenous cannula and endotracheal tube were secured meticulously to prevent injury to skin (Figure 4) 300 ml of crystalloids was administered and calculated blood loss was around 30ml. Intraoperative analgesia was supplemented with diclofenac suppository 12.5 mg and intravenous fentanyl boluses of 10mcg. Patient was covered with cotton sheet and forced air warm blanket to prevent hypothermia, temperature set at 32degree centigrade. Surgery lasted for 3 hours and patient was haemodynamically stable throughout surgery. At the end of surgery, neuromuscular block was reversed with neostigmine 0.75 mg and glycopyrolate 0.2 mg., trachea was extubated after ensuring adequate recovery. Ondansetron 2mg (iv) was administered for prevention of post-operative nausea and vomiting[5].

Subsequently patient was posted for staged procedures viz external fixator application, removal and prepuciotomy at one month intervals. General anaesthesia with endotracheal intubation was administered as mentioned earlier.

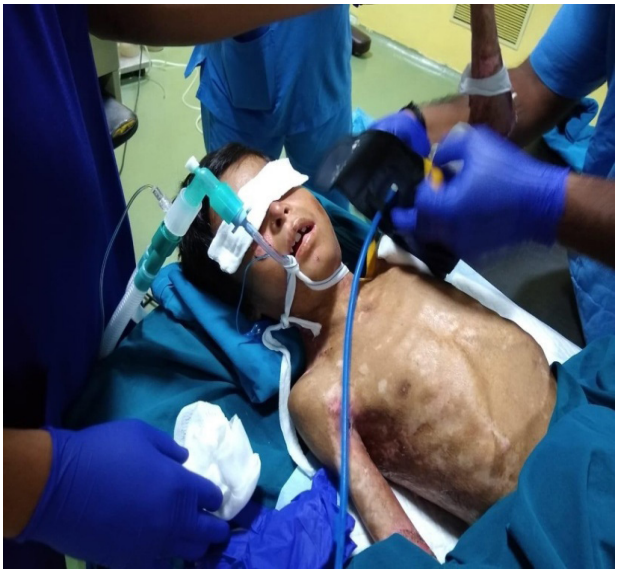

Figure 4. Endotracheal tube secured with gauze dressing.

3. Discussion

EB encompasses an array of autosomal dominant and recessive conditions that may have either localized or generalized dermatological manifestations. The loss or absence of normal intracellular bridges is due to a collagen abnormality, which makes patient susceptible for blister formation by friction/shearing forces and subsequent scarring.

Hallmark of blistering in response to minor injury, heat or friction, rubbing, and scratching from adhesive tapes[1,2].

These disorders can be categorised into three groups depending on where the actual skin separation occurs: (i) epidermolysis simplex, (ii) junctional epidermolysis, and (iii) epidermolysis bullosa dystrophica (DEB)[2]. DEB is caused by a defect in type VII collagen. DEB produces severe scarring of the fingers and toes with pseudo syndactyly formation, ankylosis of the interphalangeal joints, and resorption of the metacarpals and metatarsals Figure 1.

Involvement of the esophagus and heart resulting in dysphagia, esophageal strictures, dilated cardiomyopathy, and formation of intracardiac thrombi. Hypoalbuminemia, secondary to nephritis and protein loss into bullae is usual.

Anaemia is due to poor nutrition and repeated infections. Hypoplasia of tooth enamel results in carious degeneration of the teeth. DEB patients rarely survive beyond the third decade. Medical therapy for DEB has not been very successful.

Anaesthetic management in epidermolysis bullosa is always challenging due to problems associated with positioning, monitoring, iv access, infection, trauma to skin and mucosa, difficulty in airway. Further, Scarring of the oral cavity can cause microstomia and immobility of the tongue that increases the difficulty of tracheal intubation[5].

In order to avoid skin trauma and mucous membranes, ECG gel pads can be used. Blood pressure cuff should be padded with cotton dressing and intravascular catheters should be anchored with sutures or gauze dressing rather than tape Figure 4. Lubrication of the face mask, laryngoscope, endotracheal tube and patient's face can reduce the friction and resultant bullae formation. The use of upper airway devices should be avoided because frictional trauma to the oropharynx can result in the formation of intraoral bullae, airway obstruction, and extensive hemorrhage. We resorted to video laryngoscope to ensure atraumatic and safe intubation as the patient had difficult airway but sufficient mouth opening to admit size 2 video laryngoscopic blade Figure 3.

There are no reported complications attributable to anesthetic technique or drugs per se and use of regional techniques seem safe in these patients.

4. Conclusion

Epidermolysis bullosa is associated with potential complications and careful intraoperative management is associated with few adverse effects.Avoiding trauma to the fragile skin and mucus membranes is the key to success for providing atraumatic anesthetic care to DEB patients.

5. References

1. Epidmiology of Inherited Epidermolysis Bullosa Based on Incidence and Prevalence Estimates From the National Epidermolysis Bullosa Registry, JAMA Dermatol. 2016; 152(11):1231-38. https://doi.org/10.1001/jamadermatol.2016.2473. PMid: 27463098.

2. Inherited epidermolysis bullosa: updated recommendations on diagnosis and classification, J. AM. ACAD. Dermatol. 2014; 70(6):1103-26. https://doi.org/10.1016/j.jaad.2014.01.903. Epub 2014 Mar 29.
3. Anaesthetic management of children with epidermolysis bullos BJA Education, Copyright 2017 British Journal of Anaesthesia. 2018; 18(2):41-45. https://doi.org/10.1016/j.bjae.2017.11.005. PMid: 33456808, PMCid: PMC7808044.
4. Intraoperative cardiovascular collape in a patient with epidermolysis bullosa. Journal of clinical Anaesthesia, 2016; 34:492-493, Copyright 2016 Elservier Inc. https://doi.org/10.1016/j.jclinane.2016.06.003. PMid: 27687437.
5. Epidermolysis Bullosa, Dental and Anaesthetic Management: A Case Report. J. Dent. (Shiraz), 2014; 15(3):147-152.