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Anesthesia management in dystrophic epidermolysis bullosa: A case report

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Abstract

Epidermolysis bullosa (EB) is a rare hereditary disorder characterized by an abnormal increase in the fragility of the skin and mucosal surface and recurrent blistering. Blisters and scar formation can be seen in tissues even after a minor trauma. Due to the healing of this disease by leaving a contracted scar tissue, the findings, especially on the airway, may cause important anesthetic problems. In addition, the protection of the skin and the mucosa during the procedures to be performed, prevention of new bulla formation and prevention of infection constitute the main difficulty in anesthesia management. This case report describes the anesthesia management for the dental treatment of a patient diagnosed with dystrophic epidermolysis bullosa (DEB) and aims to review the current information.

Keywords: Epidermolysis Bullosa, Dystrophic Epidermolysis Bullosa, General anesthesia

Introduction

Dystrophic Epidermolysis Bullosa (DEB) is an autosomal recessive disease characterized by diffuse dystrophic scarring, deformities, and severe involvement of mucous membranes. Even a minimal trauma can cause massive separation of the skin and mucosa from the underlying tissue [1, 2]. The biggest challenge in anesthesia management in EBD cases is the protection of the skin and mucous membrane integrity and airway control.

In this case report, our anesthesia approach in an EBD patient scheduled for dental treatment under general anesthesia is presented and the current literature is reviewed.

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Informed Consent
The authors stated that the written consent was obtained from the parents of the patient presented with images in the study.

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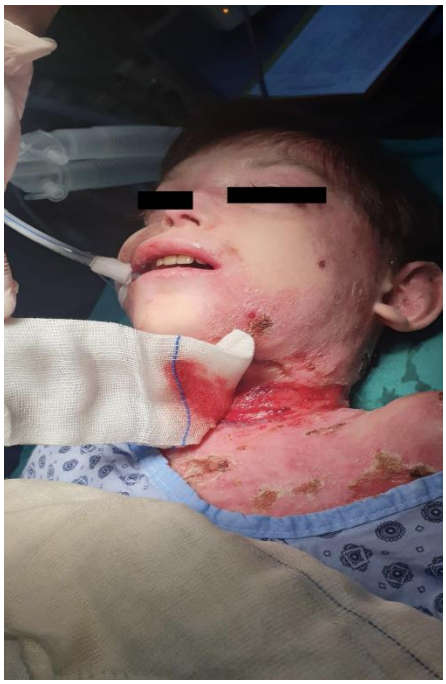
Case presentation

Dental treatment under general anesthesia was planned for a 17-year-old male patient with EBD weighing 20 kg. He had a history of surgery due to right knee joint contracture. On physical examination, there were extensive scar tissue and eroded lesions accompanied by hemorrhagic dryness on the extremities. Neck extension and mouth opening were limited due to contracted scars and ongoing hemorrhagic, drying lesions. Therefore, mallampati assessment could not be made. In indirect laryngoscopy, the vocal cords were bilaterally mobile, and the passage was open. In the laboratory examination, blood biochemistry, PT, PTT, INR, and hemogram values were within normal reference ranges.

In the preoperative unit, a tourniquet was applied, an intravenous (IV) line was opened with a 22-G cannula and fixed with vaseline gauze. Due to the expected difficult airway, various size masks, laryngoscope, endotracheal tube (ETT), flexible fiberoptic laryngoscope, flexible laryngoscope, and appropriately sized styles were prepared. Tracheotomy preparation was made.

The head of the patient, who was carefully placed on the operating table, the leg with flexion contracture, and the heel of the foot were supported with a silicone cushion. Oxygen saturation was monitored with a clip probe on different fingers instead of an adhesive finger probe. For electrocardiography (ECG) monitoring, the non-gel adhesive parts of the ECG electrodes were cut and fixed with a silk patch. Noninvasive blood pressure (NIBP) monitoring was planned before, after intubation, and when needed and was performed by placing wet cotton under the blood pressure cuff.

Figure 1: The endotracheal tube was fixed with a vaseline bandage.



Prophylactic intravenous ranitidine (1 mg/kg) and cefotaxime (50 mg/kg) were administered. Since difficult airway was considered, induction was performed with inhalation anesthetics. Using sterile gloves, the face mask was placed on the face with vaseline gauze without causing any damage to the skin, and anesthesia was induced with 4-8% sevoflurane and oxygen. Eyes were protected with moisturizing eye pomade and protective gauze. After securing the airway, muscle laxity was achieved with 0.5 mg/kg rocuronium bromide. A lubricated Macintosh was used to prevent mucosal trauma. Endotracheal intubation was performed with a no. 5 endotracheal tube, which was fixed with a vaseline bandage (Figure 1). Anesthesia was maintained with O2: N20 (40:60 ratio) and 1–1.5% sevoflurane. Methylprednisolone (1 mg/kg) was administered. Perioperative hemodynamic parameters were stable. Postoperative analgesia was provided with 10 mg/kg paracetamol. At the end of the operation, the neuromuscular blockade was antagonized with 2 mg/kg sugammadex. Extubation was performed when there was sufficient spontaneous breathing. There were no complications perioperatively, except for the development of fresh blisters on the buccal mucosa. The patient was observed in the postoperative recovery unit for 2 hours (Figure 2), then transferred to the ward, and was discharged after 24 hours. The parents were informed about the case report, and their written consent was obtained.

Figure 2: Observation of the patient in the postoperative recovery unit

Discussion

Epidermolysis bullosa is an autosomal dominant or recessive disease characterized by recurrent bullae and scar tissue formation due to increased fragility of the skin and mucosa. A bulla is formed as a result of mutations in genes encoding the proteins in the epidermis or the basement membrane (dermo epidermal junction). Repetitive bulla formation with minimal mechanical trauma is the characteristic finding in all types of EB. Four major types have been defined: EB simplex (EBS), Junctional EB (JEB), dystrophic EB (DEB), and Kindler syndrome. Other than cutaneous involvement, tracheolaryngeal complications, oral mucosal involvement, anemia, malnutrition, osteoporosis, growth and growth retardation, and cardiomyopathy may be observed [3]. Infection is common, and wound healing is delayed due to decreased immunity and long-term corticosteroid use. Recessive dystrophic EB is characterized by widespread dystrophic scarring, deformities, and severe involvement of mucous membranes, beginning at birth. Adhesion of the fingers and toes and limitation of movement due to pseudosyndactyly are frequently observed. Skin biopsy, tooth extraction, and care, eye surgery, contracture and adhesion surgery in the extremities, skin grafting, esophageal dilatation due to esophageal stricture are common surgical procedures. Anesthesia procedures can be extremely difficult due to airway distress, ankyloglossia, excessive skin sensitivity, deformities, and wounds. Because of oropharyngeal and esophageal lesions, patients have malnutrition, anemia, electrolyte disturbance, and decreased

immunity, which may change the pharmacokinetic effects of anesthetic agents.

The type of anesthesia technique is determined by the expected duration and type of surgery. In their study in which they reviewed the anesthesia records of patients with EB, Van Den Heuvel et al. reported that only patients requiring orofacial surgery were managed with standard general anesthesia, and deep sedation/analgesia technique was preferred in procedures such as wound dressings, esophageal dilatation, and syndactyly [4]. Regional anesthesia techniques can be preferred with or without general anesthesia in cooperative adult patients [5].

It is essential to know the EB subtype in EB patients. In autosomal recessive DEB, studies state that oropharyngeal bulla formation after endotracheal intubation, and postoperative stridor is less common, and dilated cardiomyopathy association is more frequent. Oral involvement, ankyloglossia, early tooth decay development, and imperfect tooth alignment are more common than other types. Subglottic stenosis, choanal and nasal stenosis, excessive paratracheal and intranasal granulation tissue, gastroesophageal reflux, and coexistence of renal and cardiac diseases are more common in JEB. EBS is more frequently associated with muscular dystrophy [5, 6].

The IV cannulation should be performed by a manual tourniquet procedure and fixed with non-adhesive silicone tapes in the preoperative preparation phase. Preoperative sedation may be beneficial in children with vascular access [6].

Atropine or glycopyrrolate, and antacid prophylaxis such as sodium citrate in case of a history of reflux or esophageal stricture, are useful to reduce excessive saliva release. Hydrocortisone treatment may be required in patients on long-term steroid therapy.

The biggest challenge in anesthesia management in EB patients is airway management and prevention of mucosal damage. In this patient group, detailed airway examination should be performed for anesthesia risk assessment. Decreased mouth opening due to contracture formation and development of temporomandibular joint motion limitation may cause position difficulties in laryngoscopy [7]. It is essential to prepare for tracheotomy considering the possibility of airway complications such as intubation difficulty, post-extubation edema, and obstruction. Lubricants should be applied to the laryngoscope blade before use. The endotracheal tube should be smaller than standard formulas suggest. ETT should be fixed with non-adhesive silicone bandages so as not to cause lip or skin damage. If nasal intubation is required in dental procedures, suturing can be used for tube fixation. Using a hard face mask, mandibular thrust, and oral airway can easily damage the skin, lips, tongue, and mucous membranes. For this reason, soft masks should be preferred, and gelled oral airways should be used. Silicone-based tape should be applied to the areas where the mask will contact the face. NgLY et al. suggested using high flow oxygen as a preferable method instead of a face mask in children with difficult airways since it does not harm the mucosa and the skin [8].

Fiberoptic intubation is less traumatic than direct laryngoscopy and should be the first choice for EB patients with difficult airway. A video laryngoscope can be helpful. Studies are suggesting that the use of a laryngeal mask (LMA) should be preferred in a difficult airway scenario because the cuff increases the formation of blisters by putting pressure on the pharynx wall. There are also publications showing that LMA, pre-lubricated with a water-based gel that is smaller than the customarily prescribed size, can be used safely [6, 9]. After difficult intubation, 0.25 mg/kg (maximum 8 mg) dexamethasone can be used to reduce postoperative airway edema. In our case, induction of anesthesia with an inhalation agent was preferred for the control of the airway, a mask with a soft air cushion was carefully used on a vaseline bandage in areas where it contacted the skin, and no complications occurred due to mask ventilation and intubation.

In EB patients, pressure and friction can cause new blisters and wounds to form. Therefore, avoiding friction and trauma during anesthesia, surgery and other procedures and preventing the formation of new blisters is the most important part of these patients' anesthesia management. The patients' transportation should be managed carefully, silicone pads should be placed on the required areas on the operating table, and it should be ensured that the bed sheet in contact with the patient's skin is appropriately placed. Self-adhesive tapes and wraps that are applied directly to the skin that may cause friction should be avoided. Instead, silicone adhesive tapes, gel, or vaseline dressings should be preferred. Minimal intervention and monitoring are recommended for EB patients [2]. ECG follow-up should be done by fixing the electrodes, whose adhesive part is cut and only the gel part is left, with silicone adhesive tapes. Pulse oximetry can be measured by using a 'clip probe' instead of an adhesive finger probe. NIBP tracking can be performed by placing cotton pads between the sleeve and the skin surface. Since no significant blood loss and hemodynamic deterioration were expected in our case, NIBP monitoring was used only when necessary. ECG electrodes were applied as recommended in the literature. Skin and mucosal damage were minimized by applying careful monitoring and handling.

Nasopharyngeal and rectal temperature probes should be used with caution and avoided if possible. Aspiration should be performed with lower pressure, protecting the mucosa.

EB patients are prone to ophthalmic complications. Especially in surgical procedures to be performed in the prone position, cornea and conjunctiva damage should be avoided, and adequate support should be provided with appropriate equipment and silicone pillows. It is important to cover the eyes with moisturizing eye pomade.

Anesthetic agents should be selected considering the accompanying systemic diseases, patient age, and difficulty in airway management. If there is IV access, intravenous induction is generally preferred. Propofol, thiopental and ketamine can be used safely in these patients [10, 11]. Induction with inhalation agents is safe in cases where a difficult airway is expected. In cases where the intravenous route cannot be provided, sevoflurane and isoflurane are the preferred agents. Inhalation anesthesia and TIVA can be used for the maintenance of anesthesia. In our case, after induction of anesthesia with sevoflurane, maintenance of anesthesia continued with inhalation anesthesia, and no complications developed. Suxamethonium, one of the muscle relaxants, should be used with caution due to its hyperkalemic potential. Muscle relaxants such as atracurium,

vecuronium, rocuronium bromide are frequently used. The duration of action of these drugs may be prolonged in the presence of hypoalbuminemia. However, it has been shown in some studies that this was not observed, and it can be used safely [11, 12].

Local anesthesia without a vasoconstrictor should be preferred for oral surgery. In postoperative analgesia, paracetamol, nonsteroidal anti-inflammatory drugs (NSAIDs) and opioids can be used safely [13].

Conclusion

Preoperative and perioperative care and teamwork, minimal monitoring, and preparation for difficult airway scenarios are important in the anesthetic management of EB patients to prevent skin and mucosal damage and formation of new blisters.

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