

General anesthesia management with laryngeal mask airway in a patient with epidermolysis bullosa: Case report

Fulya Yılmaz, Burcu Ulugolge

Health and Science University, Izmir Bozyaka Training and Research Hospital, Department of Anaesthesiology and Reanimation, Izmir, Tukey

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Abstract

Epidermolysis bullosa (EB) is a rare (1/17000-1/300000) genetic disease which is characterized by an abnormal collagen and extreme fragility of skin and mucous membranes. We report successful management of a 24-year-old-woman with epidermolysis bullosa presented to operation room for local resection of mass on tuberositas tibia under general anesthesia with laryngeal mask airway. Anesthesia management of EB cases requires features at all stages of anesthesia management. A multidisciplinary approach and careful planning are essential for safely management of surgery.

Keywords: Epidermolysis bullosa; general anesthesia; laryngeal mask airway

INTRODUCTION

Epidermolysis bullosa (EB) is a rare (1/17000-1/300000) genetic disease which is characterized by an abnormal collagen and extreme fragility of skin and mucous membranes (1-4). Antiesthetic management of these patients requires more attention than other cases. Because they have co-morbidities, patients may have complications during monitorization, venous cannulation, airway management, anesthesia management, surgical concerns (1,2,5). We report a case with EB, managed uneventfully under general anesthesia using laryngeal mask airway.

CASE REPORT

Written informed consent was obtained from the patient for hid anonymized information to be published in this case report. A 24-year-old woman (35 kg, 140 cm) with EB presented to operation room for local resection of mass on tuberositas tibia. Besides anemia and history of using drugs for EB associated skin lesions (Antibiotic and analgesic impregnated gauze was applied for dressing), she had no other medical or surgical history. Significant

preoperative findings were generalized scars, bed sores, skin and joint contractures, autoamputated fingers, poor oral hygiene, microstomia, limited mouth opening(peroral skin contractures due to EB) and neck extension. No premedication was given on the operation day. The patient positioned herself on the operation table in a supine position. She was monitored with clip-on pulseoximeter, non-invasive blood pressure with padding underneath the cuff (measured when required) electrocardiogram was not monitored to avoid skin touch.

Peripheric intravenous line was achieved (20 G cannula) on right tibia and was secured with gauze (Figure 1). Preoxygenation was followed by induction with propofol (80 mg) under gentle mask ventilation. Lubricated laryngeal mask airway No: 3 (Supreme LMA No:3, Teleflex, French) was used as a airway device (Figure 2). Because of the skin contractures her eyes were not closed spontaneously so ophthalmic gel was used to prevent ophthalmic complications. Anesthesia was maintained with oxygen,air (50%:50%) sevoflurane (Sevoraneliquide %100 (250 mL solution) (Abbvie) Istanbul, Turkey, England) and remifentanil (Rentanil 2mg, vialcontaininglyophilizedpowder, Vemilaç,

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Corresponding Author: Ersin Gundogan, Health and Science University, Izmir Bozyaka Training and Research Hospital, Department of Anaesthesiology and Reanimation, Izmir, Tukey **E-mail:** fulya.dr@gmail.com

Istanbul) infusion. At the end of the surgery under deep anesthetisalaryngeal mask airway device was removed. Postoperative analgesia was provided with IV tramadol. Postoperatively, oral examination was normal. Patient was observed for 2 hours in recovery room for signs of respiratory complications.



Figure 1. Intravenous cannula localization and wrapping



Figure 2. Perioperative monitorization and airway management

DISCUSSION

Anesthesia management is important in patients with EB. They frequently require anesthesia for multiple surgeries (dressing, dental procedure, contracture release, ophthalmic surgery vs) (3-5). Their oral intake is frequently decreased secondary to oropharyngeal and esophageal lesions; so they are prone for malnutrition, anemia and electrolyte imbalance (4). On the other

hand, they have gastro-esophageal reflux so antacid prophylaxis is advised (3-6). Difficult venous access may necessitates inhalation induction but difficult airway management and "no touch" principle to avoid skin and mucosa lesions must be kept in the mind (3,4). Reasons for difficult airway are: mouth, pharyngeal and laryngeal involvement; limited mouth opening and neck extension secondary to contractures (2,3). Maintenance of skin and mucous membrane integrity and prevention of heat and fluid loss are other major concerns (2,5).

While oropharyngeal and esophageal mucosa is stratified squamous; laryngeal and tracheal mucosa are pseudostratified, columnar, ciliated epithelium. So, laryngeal and tracheal involvement in EB is quite rare (4). Tracheal intubation is safe and supraglottic airway device can be used for airway management. Both endotracheal tube and supraglottic airway device should be preferred half to one size smaller than the predicted by standarts (4,5).

Minimal monitoring is advocated (4). Clip-on pulseoximeters preferred to self-adhesive probes, electrocardiogram electrodes should have either paraffin gauze/gel pads or silicon-mesh or needle electrodes can be preferred, non-invasive blood pressure monitoring and tourniquets can be used after application of PVC film or padding underneath the cuff, invasive arterial monitorization can be an alternative to NIBP in long surgeries (2,3,5,6).

All equipment (laryngoscope, endotracheal tube, nasogastric tube, laryngeal mask etc) should be lubricated (1,5) and catheterizations (airway, urinary vs) should be done after lubrication (4). Preferred lubricants are water soluble and petroleum jelly (3,5). Sedative premedication has previously been reported useful in younger patients to avoid restlessness (3-6). Patients with EB are risk for eye injuries so care must be taken (2,6). Postoperative pain is other major concern (2,3). Rectal administration of analgesia should be avoided. Regional techniques alone or with general anaesthesia are useful for postoperative analgesia. Analgesia can be given intramuscular, intravenous or peroral route (5,6).

Surgery can be done under general (Endotracheal tube, supraglottic airway devices), local anesthesia, central neuroaxial block and/or regional anesthesia (3). Bipolar or dry electrocautery electrodes are recommended during surgery (3,5); avoid using monopolar diathermy (1). If general anesthesia is preformed; propofol is preferred iv induction agent over thiopental and ketamine. EB maybe linked with porphyria and so thiopental is thought relatively contraindicated. Ketamine can cause excitation during recovery which leads to trauma. Non-depolarizing neuromuscular blockers can be used safely (6).

Saraf et al (6) reported successfully management of a 6-year old male child with EB presented for syndactyly under general anesthesia with endotracheal intubation. Araujo et al (1) report an elective cesarean delivery in

a woman with EB, managed successfully with spinal anesthesia. Strupp et al (2) describe a patient with EB undergoing major orthopedic surgery. They nasally intubated the patient for general anesthesia. They preferred combination of interscalene, superficial cervical plexus block and T2-T3 paravertebral blocks for regional anesthesia under ultrasound guidance and a brachial plexus catheter was placed by the surgeon at the end of the surgery for postoperative pain management. Ames et al (6) evaluated their anesthesia management for patients with EB. According to their report; they prefer general anesthesia over regional techniques, patients were often monitored poorly unlike the advices and propofol was preferred as IV induction agent.

In our patient, no premedication was given on the operation day. She was encouraged auto-positioning. We preferred laryngeal mask airway because the surgery time is short and we wanted to avoid using neuromuscular blockers. We didn't prefer regional anesthesia because approximately full body of the patient was dressed. We used clip-on pulse-oximeter, NIBP with padding underneath the cuff (when required) as monitorization tools. We could not place the ECG electrodes because when we touch the patient mucocutaneous blisters was formed)Ophthalmic eye gel was used to prevent ophthalmic complications. This case report describes a successful management of surgery with laryngeal mask airway under general anesthesia with "no touch" principle. Although experience on larengeal mask use was very limited and avoided that may lead to blister formation in oral and pharangeal mucosa), we applied laryngeal mask airway for airway management. Because all the body was dressed with antibiotic and analgesic impregnated gauze so we could not apply regional anesthesia, surgery time was too short (15 minutes) and intubation might be much more harmful than larengeal mask airway to avoid no touch principle. We also prepared alternative airway devices (fiber opticbronchoscope, fast track laryngeal intubation set etc) for possible difficult airway management. We thought that while planning an operation for these patients, it is much more appropriate

to plan the operation date multidisciplinary (anesthetist, dermatology, plastic surgeon, dentist) because of the probable exacerbation of the present disease.

CONCLUSION

Anesthesia management of EB cases requires features at all stages of anesthesia management. A multidisciplinary approach and careful planning are essential for safely management of surgery.

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Fulya Yilmaz ORCID: 0000000269017404

Burcu Ulugolge ORCID: 0000000182154844

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