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Lower-Leg Amputation Performed Under Regional Anesthesia in a Patient with Epidermolysis Bullosa: A Case Report

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Data Collection B
Statistical Analysis C
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Manuscript Preparation E
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Patient: Male, 31-year-old
Final Diagnosis: Epidermolysis bullosa
Symptoms: Multiple blisters and scars
Medication: —
Clinical Procedure: —
Specialty: Anesthesiology

Objective: Rare disease

Background: Epidermolysis bullosa (EB) is a group of rare genetic conditions that can cause eruption of blisters on the skin and mucous membranes by the slightest mechanical stimulus. In these patients particular attention should be paid to potential complications, from monitoring of vital signs to anesthesia procedures in the perioperative period.

Case Report: A 31-year-old man with EB underwent lower-leg amputation for squamous cell carcinoma. Multiple blisters and scars had appeared all over his face and body, and his extremities were contracted. The patient's mouth could open only up to approximately 5 mm, and laboratory examination showed a high inflammatory response. In addition, he had anemia and hypoalbuminemia with a serum albumin concentration of 1.4 g/dL. We planned sciatic and femoral nerve blocks with sedation for anesthesia management because of the anticipated difficulty of intubation and concern about postoperative upper-airway obstruction due to changes in the oral cavity. While protecting the skin from external force application, we performed sciatic and femoral nerve blocks (1.7 mg/kg) using 0.25% levobupivacaine, 10 mL (3.5 mg/kg) of 1% mepivacaine, and 6.6 mg of dexamethasone. Good analgesia was achieved, and the patient was stable during the operation. The patient was discharged 12 days postoperatively without additional signs of infection or new blister formation, although surgical wound healing was delayed.

Conclusions: For patients with EB who have had repeated blistering and scarring, even from a minor external force, attention should be paid to airway management and avoidance of additional skin damage caused by external forces.

Keywords: Anesthesia, Local • Epidermolysis Bullosa • Amputation

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936722

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Background

Epidermolysis bullosa (EB) is a type of hereditary skin disease characterized by extracutaneous manifestations of skin fragility and blistering, first reported by Von Hebra in 1870 [1,2]. The overall prevalence of EB varies by region, but is reported to be 4-11 cases per million [3]. EB is a rare condition in which eruption of blisters on the skin and mucous membranes can be caused by the slightest mechanical stimulus. Eruption of blisters in the oral and nasopharynx due to endotracheal intubation and airway insertion is a concerning issue for anesthesia management. Because of skin fragility and blisters all over the body, it is necessary to consider the method of vital sign monitoring and fixing of catheters. We report the anesthesia management of lower-leg amputation for squamous cell carcinoma in a patient with recessive dystrophic impaired type EB (RDEB) under sciatic and femoral nerve blocks.

Case Report

The patient was a 31-year-old man with a height of 150 cm and weight of 30 kg. He was diagnosed with EB as a newborn and frequently underwent treatment for systematic skin erosion and blisters under general anesthesia with spontaneous mask ventilation using halothane and sevoflurane in childhood. In adulthood, he had also received general anesthesia several times for treatment of esophageal strictures. All anesthesia had been managed under spontaneous breathing without tracheal intubation or mask-assisted ventilation. Tumor resection and radiation therapy were performed for the treatment of squamous cell carcinoma on the left heel several times under local anesthesia in adulthood (**Figure 1**). During the present hospital admission, lower-leg amputation was planned because of uncontrolled pain associated with ulceration and calcaneal bone melting.

Multiple blisters and scars had appeared all over his face and body, his extremities were contracted, and his digits were fused together. The patient's mouth could open only approximately 5 mm due to mandible contracture. According to the otorhinolaryngologist's assessment, no lesions were detected from the oral cavity to the larynx, and nasotracheal fiberoptic intubation with a small-diameter tracheal tube was possible, although the patient had a mild head-tilt restriction if tracheal intubation was needed. There had been no radical treatment for EB; only ointments were applied to the blisters, and there was no history of surgical treatment.

Laboratory examination showed a high inflammatory response with white blood cell count of 9.14×10^3/μL and C-reactive protein concentration of 12.25 mg/dL.

The patient was fed finely chopped or mashed meals and a liquid diet because of severely restricted mouth opening. He had anemia with hemoglobin concentration of 7.9 g/dL and hypoalbuminemia with serum albumin concentration of 1.4 g/dL. He was being administered oral oxycodone 45 mg/day and oral morphine 20 mg/day.

Based on the above preoperative evaluation, he was classified as ASA-PS (American Society of Anesthesiologists Physical Status) 3. We planned sciatic and femoral nerve blocks with sedation for anesthesia management because of the anticipated difficulty of intubation and concern about postoperative upper-airway obstruction due to changes in the oral cavity.

The operating table was covered with pressure-dispersing polyurethane foam (TEMPUR-MED(R) Mattress; TEMPURSEALY JAPAN, Hyogo, JAPAN) for pressure dispersion. A 20-gauge needle was inserted intravenously in the left forearm with nonadhesive dressing (3M™ Coban™ Self-Adherent Wrap, MN, USA). Electrodes were placed on a jelly to prevent damage to the skin. The manchette for noninvasive blood pressure measurement was applied on a bandage because EB patients are relatively tolerant of vertical pressure against the skin. A pediatric probe for pulse oximetry monitoring was wrapped around the remaining finger without exposing the sealing surface. A urinary catheter was not placed, as it was not recommended because of the risk of urethral stenosis.

A sciatic nerve block was performed with the patient in a semi-right lateral position because of severe pain in the foot wound. Chlorhexidine was poured on the site without rubbing. Under real-time ultrasound guidance with sufficient gel to minimize skin abrasion, left sciatic and femoral nerve blocks (Sonolect needle USG-type CCR®; HAKKO, Tokyo, JAPAN) were performed with a mixture of 20 mL (1.7 mg/kg) of 0.25% levobupivacaine,

Figure 1. The scarred tissue before surgery.

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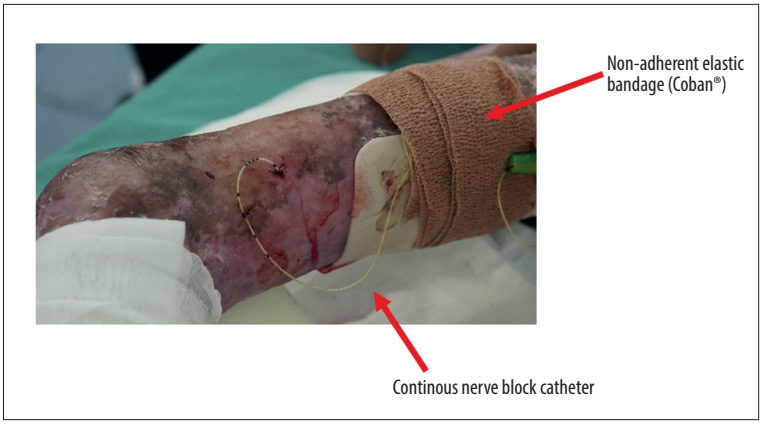

Figure 2. Fixed continuous sciatic nerve block catheter. Red arrows indicate non-adherent elastic bandage and continuous nerve block catheter.

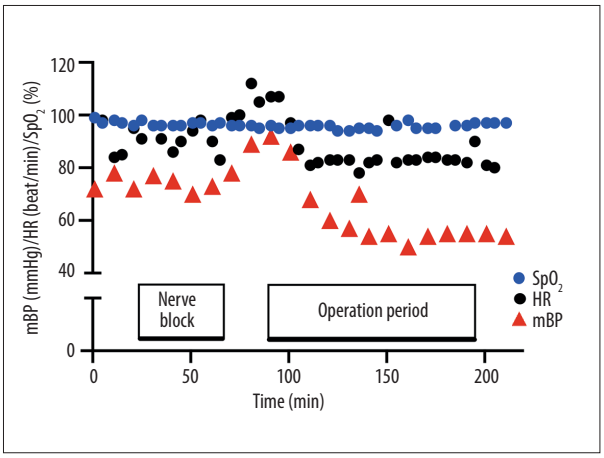

Figure 3. Anesthesia record. Red triangles: Mean blood pressure; black circles: heart rate; blue circles: SpO_2. mBP – mean blood pressure; HR – heart rate.

10 mL (3.5 mg/kg) of 1% mepivacaine, and 6.6 mg of dexamethasone. A catheter (Contiplex C®; B BRAUN, Pennsylvania, USA) was placed near the left sciatic nerve for postoperative analgesia and fixed to the healthy skin with a thread and wrapped with a non-adherent elastic bandage (3M™ Coban™ Self-Adherent Wrap, MN, USA) (**Figure 2**).

Dexmedetomidine was used for sedation (dose 0.4-6 µg/kg/h) and propofol (6-8 mg/kg/h) intermittent administration of ketamine (4 mg/kg) was used for additional analgesia. Leg amputation was performed approximately 8 cm below the knee without a tourniquet to avoid skin damage. Intraoperative blood loss was 551 mL, and 4 units of erythrocytes were transfused. The operation time was 103 min and anesthesia time was 201 min (**Figure 3**).

For postoperative analgesia, continuous sciatic nerve block (0.125% levobupivacaine 2 mL/h) and intravenous oxycodone (0.48 mg/h) were used. Postoperative analgesia was well controlled, and the sciatic nerve block catheter was removed on postoperative day 1. The intravenous oxycodone was gradually transitioned to oral medication.

The patient was discharged 12 days after the operation without additional signs of infection or new blister formation, although surgical wound healing was delayed.

Discussion

At least 18 causative genes have been identified to be related to this disorder, and abnormalities in keratin, collagen, and laminin proteins have been indicated [4]. These proteins are responsible for the anchoring function in various parts of the skin. The loss of the anchoring function in various parts of the skin and the normal barrier against infection causes skin infection and delayed wound healing [2,5]. As in the present case, recessive dystrophic impaired type EB is caused by the type VII collagen gene, and the junction between the basement membrane and the upper dermis is fragile. The clinical manifestations are most prominent, and unlike other types of EB, the blisters tend to form severe scars as they heal. This often results in scar tissue in the skin, such as fused fingers.

Anesthesia management for EB patients is important to avoid as much as possible applying friction and shearing forces to the patient. In fixation of various routes to the skin, it is recommended to use nonadhesive dressings such as silicone-based tapes or other soft dressings (Mepitac®, Mepitel®, Mepilex®, Mölnlyche Healthcare, Västra Götalandslän, Sweden)[6]. ECG electrodes need to have a gel pad between them and the skin. Noninvasive blood pressure measurement can be used if there is a pad under the cuff. Monitoring accurately could be performed for all parameters in this case. In addition, restrictions of body position due to joint contractures should be considered.

Airway Management

The most important point of anesthesia management for EB patients is airway management. There are reports in which face mask, oral tracheal intubation, nasotracheal intubation, tracheostomy, and supraglottic devices were used in anesthesia for

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EB [7,8]. Unnecessary mask ventilation should be avoided because of eruption of blisters on the face due to physical compression, and risk of gastroesophageal reflux and difficulty in prolonged mask ventilation because of lubricated masks [9].

There are many reports of difficulty of intubation in EB patients because of difficulty in visual field securement due to lesions in the oral cavity, pharynx, and larynx, and difficulty in laryngeal deployment due to neck contracture, restricted mouth opening, and decreased tongue mobility.

Furthermore, it is important to note that the risk of difficult intubation is higher in adults than in children because of progressive neck contracture, restricted mouth opening, and shortening and contracture of the tongue [10].

On the other hand, there are reports that no serious tracheal intubation-related complications occurred [11-13]. It is possible that the pseudostratified columnar epithelium that covers the trachea and most parts of the larynx except the false vocal cords and parts of the epiglottis makes them less susceptible to friction-induced blisters than the oral cavity and pharynx, which are composed of multilayered squamous epithelium [14]. In this case, intubation via oral was impossible due to trismus and nasotracheal fiberoptic intubation could cause postoperative rhinostenosis. Thus, the choice of endotracheal intubation should be made with caution [10,15].

Dexmedetomidine was used because it causes less respiratory depression and loss of airway obstruction and has an analgesic effect. In addition to dexmedetomidine, we intermittently used ketamine, which provides appropriate sedative and analgesia level without loss of the gag reflex and pharyngeal reflex in patients with EB [14,16]. We chose it to prevent aspiration pneumonia because the patient had esophageal stricture and history of vomiting when he was anxious.

Regional Anesthesia

Epidural or spinal anesthesia and peripheral nerve blocks can be performed at areas with no local infection [6]. However, in addition to blistering at the planned puncture site and high inflammatory response, the skin of EB patients is prone to infection because of the loss of the normal infection barrier, which causes delayed wound healing [2,5]. Therefore, peripheral nerve block was chosen in this case because of the possibility of infection development caused by epidural and spinal anesthesia. Although the patient was at high risk for local anesthetic systemic toxicity because of severe hypoalbuminemia, we were able to manage the patient perioperatively without complications.

Conclusions

This report describes safe and effective perioperative management using peripheral nerve block in a patient with EB who had repeated blistering and scarring, even with minor external force, and had difficulty in positioning and intubation because of joint contracture. In addition to airway management, a method of vital sign monitoring and how to fix catheters should be considered.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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