

CASE REPORT

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Spinal anesthesia in a patient with epidermolysis bullosa

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Abstract

Epidermolysis bullosa (EB) is a rare genetic disorder characterized by skin and mucosal fragility that leads to exaggerated blister formation triggered by minor trauma or occurring spontaneously. Squamous cell carcinoma develops at a younger age compared to other populations, especially in recessive dystrophic EB (DEB) subtypes. Ensuring skin and mucosal integrity is crucial in anesthesia monitoring and airway management for EB patients. Here, we present the case of a 23-year-old woman diagnosed with DEB, presenting with intraoral blisters and scheduled for excision of squamous cell carcinoma of the foot. Spinal anesthesia was administered for the patient's anesthetic management. It should be remembered that spinal anesthesia can be a suitable method in patients with dry and infection-free skin at the puncture site.

Keywords: Epidermolysis bullosa, spinal anesthesia, squamous cell carcinoma

Introduction

Epidermolysis bullosa (EB) is an autosomal recessive mucocutaneous disorder clinically characterized by exaggerated blister formation following minor trauma. Anesthetic management of these patients requires attention due to scars limiting oral opening, pharyngeal and laryngeal blisters, dehydration, hypoalbuminemia, anemia, and thrombocytosis. Minimal monitoring that minimizes trauma should be preferred [1].

Case Presentation

A 23-year-old, 46 kg, 163 cm female patient was diagnosed with EB at 15 months of age and scheduled for mass excision by the plastic and reconstructive surgery team due to suspicion of squamous cell carcinoma in the right foot.

Preoperative evaluation included obtaining informed consent after explaining the operation to the patient and obtaining permission to publish the case. Physical examination revealed blistered lesions around the mouth, on the extensor surfaces of both upper and lower extremities, and on the elbows and feet, along with dystrophic changes in fingernails and toenails (Figures 1, 2 and 3). Oropharyngeal examination revealed lesions inside the mouth and decayed teeth. The Mallampati score was 2, with

no restrictions in neck movements. Laboratory test results were within normal limits.



Figure 1. Preoperative physical examination findings



Figure 2. Clip-on pulse oximeter with adhesive part on the glove

CITATION

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Figure 3. Restricted mouth opening and tooth deformities

The patient had a history of bleeding and postoperative swallowing difficulties due to intubation during previous surgeries. Considering the intraoral blisters and the absence of infection and dryness at the puncture site, spinal anesthesia was planned and discussed with the patient.

To minimize hypothermia, the operating room temperature was adjusted before the patient's arrival. A 22-gauge cannula was placed in the dorsal hand vein using thin gauze instead of adhesive tape for fixation. Pediatric ECG electrodes were applied over gauze, and a cotton layer was placed under the blood pressure cuff to prevent skin contact. A pulse oximeter was attached to an unaffected thumb using gauze (Figure 4).



Figure 4. Single layer cotton between blood pressure cuff and arm

The lumbar skin was gently prepared with povidone-iodine solution while preventing friction, allowing it to air dry. Sterile lubricating gel was used to facilitate atraumatic palpation and puncture. A single puncture was performed at the L3-4 interspace using a 25-gauge Quincke spinal needle, administering 10 mg heavy bupivacaine and 10 mcg fentanyl. The puncture site was covered with a non-adhesive dressing, and the patient was positioned supine.

The patient, exhibiting preoperative anxiety, received 1 mg midazolam and supplemental oxygen through a face mask. Cotton was applied to mask straps to avoid skin damage. The cotton was coated with Vaseline to prevent it from sticking to the skin and causing damage (Figure 5).



Figure 5. Single-layer cotton wrapped between the neck and oxygen mask

The 90-minute surgery proceeded without complications. Postoperatively, the patient was monitored for 30 minutes in the recovery room, with a motor block duration of 120 minutes. The patient was discharged without complications after two weeks of follow-up

Discussion

Epidermolysis bullosa encompasses dermatological disorders causing fragile skin and mucosa that are prone to injury. It primarily affects stratified squamous epithelium, including the skin, oral cavity, pharynx, esophagus, and genitourinary system. Extracutaneous manifestations, such as laryngotracheal stenosis, esophageal strictures, infections, hypermetabolic states, and cardiomyopathy, pose significant challenges for anesthesiologists [2].

Preoperative evaluation should include metabolic assessment, focusing on malnutrition, anemia, fluid and electrolyte imbalances, amyloidosis, hypercoagulopathy, porphyria, and neuromuscular disorders commonly associated with EB [3].

When planning anesthesia, all the characteristics of the patients should be taken into account. Our patient's history did not reveal any significant features.

In these patients, even minor trauma can lead to severe blister formation and infection. Anesthesia monitoring should be as minimally traumatic as possible. Necessary precautions should be taken to minimize friction-induced trauma under anesthesia. ECG electrodes should be applied with as few adhesives as possible and without using sticky substances. Invasive arterial blood pressure monitoring is not recommended, as subsequent scarring may impede arterial circulation. Literature reports do not mention blister formation induced by pulse oximeter application due to pressure. However, the use of clip-type pulse oximeters, which can open and close, is still recommended. In our case, we applied the ECG electrodes without adhesive, placed a single layer of cotton between the blood pressure cuff and the skin to avoid excessive pressure, and used a sticky pulse oximeter with the adhesive part facing the protective gloves, which allowed us to monitor SpO₂ and prevent excessive pressure from the clip-type pulse oximeter [4].

In Epidermolysis Bullosa (EB), the mouth opening may be limited due to contractures and scarring, so detailed examination and preparation should be done for the potential of a difficult airway. Aspiration risks related to esophageal strictures should also be considered. Dental deformities in these patients may also cause difficulties. Lin et al. reported that, during general anesthesia procedures for 121 surgeries in 25 children with EB, three patients required fiberoptic laryngoscopy [5]. It has been emphasized that spinal anesthesia may be more advantageous in EB patients undergoing elective or emergency cesarean sections. In our patient, due to the presence of blisters in the oral cavity and a previous history of oral bleeding and infection post-general anesthesia, we preferred spinal anesthesia [6].

Adequate postoperative analgesia can reduce unnecessary movements due to pain, which in turn helps minimize trauma [7].

For spinal anesthesia, we extended the duration of postoperative analgesia by adding 10 mcg of fentanyl.

Conclusion

In anesthetic management of patients with epidermolysis bullosa, protecting skin and mucosal membranes from mechanical damage is essential, achieved through minimal monitoring and minimal contact. It should be emphasized that spinal anesthesia can be a favorable option due to potential airway challenges.

Conflict of Interests

The authors declare that there is no conflict of interest in the study.

Financial Disclosure

The authors declare that they have received no financial support for the study.

Patient Informed Consent

The patient declares that she gives permission to share images and information about the disease in this study

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