

DYSTROPHIC EPIDERMOLYSIS BULLOSA WITH SKIN CANCER – IMPLICATION ON ANESTHETIC MANAGEMENT – A CASE REPORT

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Summary

We report a patient with recessive dystrophic epidermolysis bullosa who developed multiple skin cancer on the upper arm. She was scheduled for arm amputation. Skin and mucosa lesions render more difficulty in anesthetic management, especially monitoring, positioning and airway instrumentation. General anesthesia based on ketamine enables us to avoid some of the problems.

KEY WORDS: *epidermolysis bullosa, malignant alteration, difficult airway, anesthetic management, ketamine*

DISTROFIČNA BULOZNA EPIDERMOLIZA S KOŽNIM KARCINOMOM – UTJECAJ NA ANESTEZIJU – PRIKAZ SLUČAJA

Sažetak

Opisujemo bolesnicu s distrofičnim oblikom buloznog dermatitisa u koje su se razvili multipli kožni karcinomi nadlaktice. Bolesnica je predviđena za amputaciju ruke. Kožne promjene i promjene sluznice otežavaju vođenje anestezije, a osobito promatranje, namještanje i pristup dišnom putu. Opća anestezija ketaminom omogućila nam je da izbjegnemo neke od navedenih problema.

KLJUČNE RIJEČI: *bulozna epidermoliza, maligna alteracija, otežan dišni put, anestezija, ketamin*

INTRODUCTION

Epidermolysis bullosa is an inherited or acquired (an autoimmune disease) blistering disease affecting the skin and mucous membranes. The prevalence is 1 : 300 000 people per year (1). A minor mechanical trauma leads to separation of skin layers and bulla formation caused by ineffective, intercellular collagen skin bridges. Later, it heals with scarring and contractures (1).

The site of interlayer separation defines epidermolysis bullosa as simplex, junctional or

dystrophic. Dystrophic epidermolysis bullosa is associated with scarring of the fingers and toes which lead to hand deformities. Patients with recessive dystrophic epidermolysis bullosa have insufficient amount of collagen type VII and increased collagenase (1, 2). Furthermore, they have increased propensity to malignant alteration to aggressive cutaneous squamous cell carcinoma with frequent metastases and local recurrences (3-5). The management includes surgical treatment - excision or amputation, whereas irradiation and chemotherapy are ineffective (4). Be-

sides, most patients undergo elective surgery of hand deformities, esophageal strictures and dental surgery. Great care should be taken to avoid further bulla formation during the surgery. Therefore, the surgeon and the anesthesiologist have to anticipate and prevent possible complications of their intervention.

CASE REPORT

We report a clinical case of a 28-year-old woman, height: 155 cm, weight: 40 kg, suffering from recessive dystrophic epidermolysis bullosa. Due to multiple skin cancers on the arm and axillary metastases, the patient was scheduled for right arm amputation and right axillary dissection under general anesthesia in the dorsal position.

Preoperative evaluation reveals multiple bullae and ulceration, small mouth and carious teeth. The patient has only one preserved finger on each hand, with contracture of other fingers. Pathologic preoperative laboratory test results are the following: hemoglobin = 10 g dL⁻¹, hematocrit = 29%, red cell count = 3.46×10^{12} L⁻¹, total protein = 71g L⁻¹, albumin = 32.4%. These tests with the body mass index = 16.64 kg m⁻², describe a poor nutritional status. Of medications, she was taking analgesics and a histamine receptor antagonist, ranitidine.

Preoperatively, we performed cannulation of the subclavian vein and intravascular catheter was anchored with sutures. Preoperative medication was avoided for the patient should be able to place herself in the best position. Monitoring included blood pressure, pulse and oxygen saturation with pulse oxymetry. Special attention was paid to the blood pressure cuff that was padded with cotton dressing and maximal measurement time interval. The operation was done in general anesthesia and lasted 90 minutes. We decided to avoid tracheal intubation and maintained spontaneous breathing during the surgical procedure. We took advantages of ketamine - it has a good analgesic effect and maintains spontaneous breathing. Oxygen was given through a nasal cannula. The anesthesia was induced by ketamine 2 mg/kg and midazolam 0.2 mg/kg IV and maintained with ketamine 0.5 mg/kg and midazolam 0.1 mg/kg IV. During the operation,

the patient was hemodynamically stable. After the surgery, the patient was discharged without any major complication first to the ICU and then to the surgical ward with everyday redressing. On postoperative day 15, patient was discharged from the hospital in good condition and without new cutaneous lesions.

DISCUSSION

Patients with recessive dystrophic epidermolysis bullosa have to undergo repetitive surgery of the hand, due to the correction of severe hand contractures or cancer (3, 6-9). The primary concern in anesthesia for the patient with epidermolysis bullosa is a careful management to prevent even minor skin and mucosa trauma due to the potential risk of bulla formation (10). Anesthetic management of patients is complicated by monitoring, positioning and difficult intubation. Scarring of the skin require special attention to placement of blood pressure cuff, intravascular catheters, ECG electrodes and pulse oxymetry (11,12). Adhesive material and sticking plasters are forbidden. The patient alone has to find the position what will not cause additional blistering due to rubbing (9). Lateral pressure is much more damaging than perpendicularly applied pressure (13). Of importance is a padding of all pressure points before the beginning of induction (10,13). This disease always carries specific anesthetic implication on difficult airway instrumentation. There are two reasons for difficult airway management - 1. scarring of the oropharyngeal mucosa with microstomia, decreased tongue mobility and poor dentition; 2. endotracheal instrumentation carries the risk of mucosal trauma with bleeding and of new blistering; 3. limited movability of the temporal mandibular joint (14,15). In dystrophic type of dermatitis bullosa, laryngeal mucosa is rarely affected, in contrast to oropharyngeal mucosa. On the other hand, patients with junctional type have affected laryngeal mucosa too. Regional anesthesia and ketamine anesthesia enable to avoid airway instrumentation. Epidural and spinal anesthesia could be performed for lower limb surgery depending of the required block height (10). Brachial block is often used for hand surgery (7,8,10). Another approach is the use of ketamine anesthesia features,

as an anesthetic with strong analgesic and amnesic effect, without respiratory depression (7,16). The main advantage is the ability to maintain spontaneous breathing and protective reflexes (16). Excitation on the recovery is possible, and therefore special care must be taken (9).

In summary, the patient with epidermolysis bullosa is a challenge to the anesthesiologist not only because of the difficult airway management, but also because of other specific implications of anesthetic management associated with vascular access, intraoperative monitoring equipment and positioning. We want to emphasize that the choice of anesthetics makes it possible to avoid airway instrumentation in the case of a patient with the susceptible and vulnerable airway. Detailed knowledge of the disease and special precautions to avoid any mechanical trauma of the skin and mucosa are necessary for successful anesthetic management of a patient with epidermolysis bullosa.

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Received for publication: December 7, 2004

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