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Anesthetic Approach to a Patient with Epidermolysis Bullosa: A Case Report
Epidermolizis Büllozalı Hastada Anestezik Yaklaşım: Olgu Sunumu

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ÖZET

Epidermolizis bülloza (EB); cilt ve mukozalarda frajilite ve bül oluşumuyla karakterize bir hastalıktır. Özellikle havayolundaki büller, yapılacak girişimlere bağlı yeni bül oluşumu anestezi yönetimini güçleştirir. Bu sunumda rekonstrüktif cerrahi planlanan, 6 yaşındaki, EB tanılı olgunun anestezik yönetimi anlatıldı. Anestezi ve analjezi optimal monitörizasyon ve girişim sonrası intravenöz (iv) yoldan sağlandı. Ventilasyon ise orofaringeal lezyonların varlığı ve ağız açıklığındaki kısıtlılık nedeniyle yüz maskesi ile gerçekleştirildi. Yeni lezyon veya komplikasyon oluşmadı.

Anahtar Kelimeler: anestezi; epidermolizis bülloza; preoperatif değerlendirme

ABSTRACT

Epidermolysis Bullosa (EB) is characterized by fragility and formation of blisters in the skin and mucosal membranes. Due to blisters on the airway and occurring new blisters as a result of anesthetic or surgical procedures may complicate anesthesia management. In this case report, we present the anesthetic approach of a 6 years old case with EB who underwent reconstructive surgery. After premedication and optimal monitorization, intravenous (IV) anesthesia and analgesia was administered. Associated with the oropharyngeal lesions and limitation to reach an adequate mouth opening, we preferred the facemask to maintain ventilation. Neither new lesions nor complications were detected during or after surgery.

Key words: anesthesia; epidermolysis Bullosa; preoperative evaluation

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INTRODUCTION

Epidermolysis Bullosa (EB) is a rare, chronic and heterogenic mucocutaneous disease, characterized by abnormal fragility caused by autosomal dominant or recessive mutations in the genes coding structural proteins of the skin and mucosal membranes[1"5]. The layers of the skin detaches spontaneously or as a result of minor mechanical abrasion or trauma; it is followed by intradermal fluid accumulation, formation of blisters and finally to the formation of scars. Several complications which includes; pain, malnutrition, secondary wound infection, sepsis, vulnerability to cancer (squamous cell carcinoma/malign melanoma), growth and developmental delay may accompany to the skin lesions[5"7]. Anesthesiologists encounter these patients during reconstructive surgery to treat cases such as pseudosyndactyly, surgery to increase the opening of the mouth, esophageal balloon dilatation, placement of gastrostomy tubes and dental restoration procedures[8]. These cases, however, are difficult cases as they may develop new bullous lesions as a result of anesthetic or surgical procedures[9].

CASE REPORT

Preoperative Evaluation:

A 6 years old 20 kg child with congenital EB underwent reconstructive surgery for bilateral hand lesions at the Plastic and Reconstructive Surgery clinic. Medical history was clear except an negligible anemia. Mallampati score was III, neck movements and thyromental distance were normal but mouth opening was limited (2 cm). Physical examination revealed bullous, erythematous and dry skin lesions of different sizes covering all of the body. Additionally, mitten hand and foot deformity (pseudosyndactyly), and joint contractures related to scar formation especially on the right hand as well as aphthous lesions in the oropharyngeal mucosa were seen. Laboratory test results indicated only anemia with an hemoglobin level of 9.7 g/dL.

Anesthetic Approach

Endotracheal intubation was avoided as the case was considered difficult airway with high risk of exacerbation or impairment in the oropharyngeal lesions. An anesthetic approach consisted to preserve and support spontaneous ventilation was planned. Premedication with oral midazolam at a dose of 0.5 mg kg"[1] was administered 30 minutes before surgery. Due to the skin lesions on the extremities, we used a 22 G sterile cannula toestablish vascular access via the right femoral vein. A non"allergenic plaster was used to stabilize the cannula (Cansin Fix Elastic Tape, Cansin Healthcare Company, Turkey). Pulse oxymeter probe was placed on the earlobe to monitor peripheral oxygen saturation (Nellcor Oximax MAX-P Pediatric Oxymeter Probe, United States). The probe was frequently switched between each earlobe to necessitate optimal oxygen monitorization. Heart rate and rhythm was monitored with electrocardiogram (ECG) using non-allergic plasters and gel to maximize contact to the skin. Blood pressure was monitored non-invasively on the arm which surgery was not scheduled at that moment (with long intervals between measurements). Wet cotton was placed between the cuff and the arm to prevent possible trauma that might occur due to the inflation of the cuff. All pressure points were supported with silicone pillows. After 3-minutes of pre-oxygenation with 100% O_2 and remifentanyl 0.1 pg kg^{-1} min^{-1} IV infusion, propofol 1 mg kg^{-1} IV was administered to induce anesthesia. A proper facemask was lubricated with Vaseline (Vaseline liquid ® Merkez Pharmaceuticals, Istanbul) and placed to the face with avoiding pressure. End-tidal CO2 monitorization was also performed. Anesthesia was maintained with 5 L/min 50% O_2 /50% air mixture, propofol 100125 pg kg^{-1} min^{-1} and remifentanyl 0.1 pg kg^{-1} min^{-1} IV infusions. During surgery,

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hemodynamic parameters were within normal limits. A total of 400 ml crystalloid solution was given to the patient. Bilateral hand deformities were repaired in 150 minutes. The Patient was observed in the recovery room for approximately one hour and afterwards taken to the in"patient service. Postoperative pain management was done using oral paracetamol (250 mg/ 6 hours). No complication was reported during the post"surgical period and the patient was discharged two days after surgery.

DISCUSSION

There is no effective treatment or cure for EB. In these patients, avoiding traumas that would trigger the formation of new bullous lesions, secondary infections and scar formation is the main principal of management of the disease [3]. Management of the airway and protection of the skin integrity can be difficult in these cases. After recovery period, bullous lesions around the mouth generate scars and contractures which may cause movement restriction at the temporomandibular joint and mouth resulting in difficulty in airway management. Thus, appropriate equipment should be made available before the initiation of anesthetic procedures. Anemia, electrolyte imbalance and hypoproteinemia should be treated before surgery[3,9,10]. In our case, the patients had limited ability to open her mouth, therefore, we made sure that all necessary equipments were kept ready to use. Pulse oxymetry, capnography, and non"invasive blood pressure monitorization (preparations should be made to protect the skin under the cuff such as the placement of wet cotton, Vaseline-soaked bandage, or Mepitel® on the arm before placing the cuff) is suggested for all patients with similar problems[10]. Intra-arterial blood pressure monitoring is not advised unless indicated otherwise. Routine ECG monitorization is not necessary in patients that would not be intubated. However, if necessary, electrodes should be placed using silicone-based adhesives or used after the removal of the adhesive or consolidating gel parts[8,9]. In our case pulse oxymetry, capnography, non-invasive blood pressure and ECG monitorization were performed. We did not observe any lesions or complications related to monitorization in our patient.

Intravenous analgesia and anesthesia is less harmful when compared to induction using face mask. However, the lesions may complicate the establishment of vascular access[11]. Therefore, if possible IV cannula should be placed during the preanesthesia period. In our case, we could not find sufficient area to access into the vessels and had to use the femoral vein.

Anesthesia should be defined on the basis of the patient's condition and the type of surgery scheduled[12]. General anesthesia is commonly used, however regional interventions can be preferred in suitable cases as they are safer procedures in terms of bullous lesions[10]. To decrease the risk of trauma, water-based gel or moisturizing creams can be used when replacing facial masks, laryngeal masks or intubation tubes. Facial masks should be properly inflated and smaller endotracheal tube should be preferred for intubation[8,12]. In our case, we used Vaseline for the parts of the facial mask directly coming in contact with the skin.

In conclusion, careful preoperative evaluation should be performed and unnecessary monitorization and interventions should be avoided to prevent complications in cases with EB.

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