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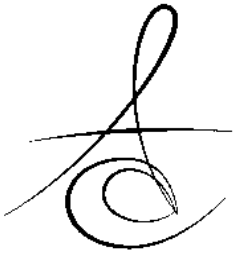

ANESTHETIC APPROACH FOR ORAL SURGERY PROCEDURE OF PATIENT WITH EPIDERMOLYSIS BULLOSA

BÜLLÖZ EPİDERMOİDLİ HASTAYA ORAL CERRAHİ OPERASYONU İÇİN ANESTEZİK YAKLAŞIMIMIZ

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ABSTRACT

Epidermolysis bullosa is a congenital, hereditary disease, characterized with blisters of the skin and mucosal membranes. Abnormal healing of these blisters results with contracted scars and erosion. Whether diagnostic, treatment or anesthesia, it is likely that the patients will require different procedures in certain phases of their lives; every process can result with new blisters. In this study our aim is to present a 19 year-old patient with Simplex type Epidermolysis bullosa and taken various precautions in order to prevent new blister formation during minor oral surgery procedure.

Precautions were taken in order to minimize new bulla formation during preprosthetic surgery, which involved lengthening crown height with gingivectomy for all erupted teeth and removal of excess mucosal tissue over the unerupted first molars for obtaining eruption. After healing was observed prosthetic treatment was carried out in order to fulfill esthetic demands.

Precautions such as using pillows under pressure areas such as heels and ankles, using paddings under the blood pressure cuff, using smaller electrocardiogram pallets, using hand pressure instead of tourniquet for catheterization minimized new blister formation.

Keywords: Anesthesia, Epidermolysis Bullosa, Preventive Measures

ÖZ

Büllöz epidermoid deri ve mukoz membranlarda bül oluşumuyla karakterize konjenital, herediter bir hastalıktır. Büllerdeki anormal iyileşme kontrakte yara izleri ve erozyonlarla sonuçlanır. İster tanı amaçlı, ister tedavi amaçlı ister anestezik amaçlı olsun bu hastaların hayatları süresince değişik girişimlere ihtiyaç duymaları olasıdır ve her girişim yeni bül oluşumu ile sonuçlanabilmektedir. Bu çalışmada 19 yaşında simpleks tip büllöz epidermoid hastası genç bir erkeğin minör oral cerrahi için yapılan anestezisi sırasında yeni bül oluşumunu önlemek için alınan çeşitli önlemlerin paylaşılması amaçlanmıştır.

Minör cerrahi sırasında yeni bül oluşumunu engelleyecek tedbirler alınmış ve protetik tedavi öncesi hastanın sürmüş olan dişlerine kron boyunu uzatmak amacıyla gingivektomi uygulanmış, sürmemiş birinci molar dişler üzerindeki fazla yumuşak doku sürmenin gerçekleşmesi amacıyla uzaklaştırılmıştır. İyileşmenin gözlenmesinin ardından, protetik tedavi tamamlanarak hastanın estetik ihtiyaçları karşılanmıştır.

Topuk ve bilekler gibi basınç alanları altına konulan yastıklar, tansiyon manşonu altına konulan pedler, kullanılan küçük elektrokardiyogram peletleri, damar yolu açılması sırasında turnike yerine elle bası kullanılması gibi önlemler yeni bül oluşumunu azaltmıştır.

Anahtar Kelimeler: Anestezi, Büllöz Epidermoid, Önleyici Tedbirler

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INTRODUCTION

Epidermolysis bullosa (EB) is a genetic disorder, which pattern of inheritance may be either autosomal dominant or autosomal recessive, characterized by separation of the epidermis from the underlying dermis (1,2) resulting with blisters. The disease mainly affects the stratified squamous keratinizing epithelium of the skin; however, blisters can occur on virtually any mucosal surface, including the oral mucosa (3,4). EB is classified into four main groups, simplex, recessive dystrophic, dominant dystrophic and junctional, based on mode of inheritance, anatomic location and distribution of the lesions and associated morbidity (2).

Clinical manifestations range widely, from localized blistering of the hands and feet to generalized blistering of the skin and oral cavity, and injury to many internal organs (5), involvement of upper gastrointestinal system mucosa is very often. Abnormal healing often results with contracted scars and erosion. The disease may be associated with abnormal dentition and even complete loss of permanent teeth (6) and dental anomalies ranging from mild hypoplasia to missing teeth have been reported in individuals with simplex EB (2).

It is likely that patients will require different procedures in certain phases of their lives. Whether the purpose is diagnosis, treatment, anesthesia or surgery, every process can result with new blister formation. Precautions should be taken in order to prevent new blister formation.

In this study we present our anesthetic approach and taken precautions to a patient with simplex EB. In our patient, all teeth were affected with amelogenesis imperfecta and molar teeth were covered under thick mucosal or bony tissue. Patient required gingivectomy around the upper and lower incisors, canines and premolars in order to lengthen crown height and needed removal of the excess mucosa over the upper and lower molars, which were held accounted for impaction.

CASE REPORT

19 year-old simplex type EB patient who weighed 60kgs, measured 170cms, admitted to our clinic with aesthetic complaints about his image. He was a high school graduate and wanted to correct his image before applying to university. He was being examined by a medical doctor regularly and did not use any systemic medication for his condition. He was only using a soluble dressing containing nitrofurazon locally for his bullas. After radiological and clinical examination he was diagnosed with amelogenesis imperfecta (Figure-1). Crown lengths were too short for a healthy prosthetic restoration. Molars in both jaws and maxillary right canine were unerupted. Since the patient was 19 years old the treatment plan did not involve extraction for the unerupted teeth. Lengthening crown height with gingivectomy for all erupted teeth and removal of excess mucosal tissue over the unerupted first molars in order to obtain eruption and prosthetic restoration was planned. Because of the extensive operation area procedure was planned under general anesthesia with nasotracheal intubation and the patient was consulted to the anesthesiologist. Patient had no former history of surgery or anesthesia. He had widespread lesions on all extremities and joints (Figure-2). Laboratory tests revealed no abnormalities, patient was Mallampati Class 3. Patient did not have any limitation of his mouth opening due to his condition. After 6 hours of fasting patient was taken to the operation room without premedication. Pillows were placed under pressure areas such as ankles and heels. Instead of tourniquet use hand pressure over the vein trace was used and peripheral venous catheterization was achieved with 22-gauge needle. Wet padding was used under the blood pressure cuff, which was used to monitor non-invasive blood pressure. Smaller electrocardiogram pallets, which did not contain gel, were immobilized with sticking plaster. For fingertip oxygen saturation monitorization a single use probe, which did not apply pressure, was used.

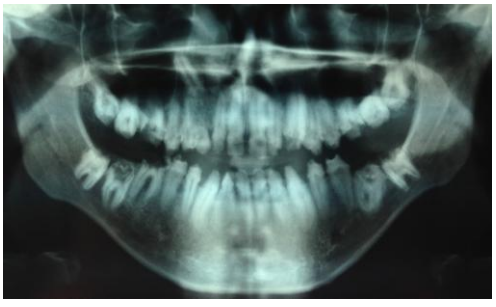

Figure 1. Panoramic radiograph of the patient

Figure 2. View of the patients' lesions

After anesthesia induction was achieved with 2mg/kg propofol, 0.9mg/kg rocuronium and remifentanil; nasal decongestant spray was applied and patient was intubated without complication using McCoy laryngoscope and special production 7.0 nasotracheal tube.

Eye pomade with antibiotic was applied and eyes were covered with wet padding, possible pressure areas; like head and heals were supported with small pillows. Anesthesia maintenance was obtained with % 50 O_2/air, % 2 sevoflurane and 0.05-0.1 µcg kg-1 dk-1 remifentanil infusions. He was administered 40mg metilprednisolon and 1mg ampicillin sodium before surgery. After implementation of infiltration anesthesia gingivectomy and removal of excess mucosal tissues over the molars was performed. After the 95-minute procedure inhalation agents were terminated, muscle relaxants were antagonized; patient was extubated after 4 minutes when his respiration became adequate. After 30 minutes of observation period patient became hemodynamically stabile and was sent to service in good condition. Post operatively new blister formation was observed only on the nose where mask was applied. Patient was discharged at postoperative day 2. After healing prosthetic treatment of the patient was finalized (Figure-3,4).

Figure 3. Panoramic radiograph after prosthetic treatment

Figure 4. View after prosthetic treatment

DISCUSSION

EB is a diverse group of disorders that have as a common feature blister formation with tissue separation occurring at variable depths in the skin and mucosa depending on the specific EB type (4). In this case our patient was a Simplex type EB patient. The site of tissue separation in patients with Simplex EB is within or just above the stratum basalis (4). Patients with mild forms of EB can usually be treated with local anesthesia like any other patient, and will easily tolerate dental procedures, however severe soft tissue involvements requiring surgical procedures are best managed with general anesthesia (4). In these patients, due to risk of esophageal stenosis, dehydration, malnutrition, anemia, hypoalbuminemia, electrolyte imbalance, thrombocytosis and infections; the need of attentive anesthetic selection for anesthesia induction and maintenance because of increased porphyria, myasthenia gravis and muscular dystrophy probability; assessment of mucosal fragility should be carefully made to form an altered approach and the surgeon should be alert for possible perioperative traumas during dental treatment (4).

Crowley et al. (7) reported a case of difficult intubation (Mallampati Class 4) of a patient with Epidermolysis bullosa due to limited neck extension secondary to scarring, a small mandible, and a severe range of motion limitation in the temporal mandibular joint. Implementation of minimal attempts with optimal monitorization is required in these patients. Similar to our study Lanier et al. (8) reported a study where they used cream, bandages and pads to minimalize postoperative trauma. They concluded that dental care of these patients could be carried out effectively with these precautions. In addition to our taken precautions Prabhu et al. (9) reported use of lubrication of the facemask and laryngoscope. Although precautions were taken they reported slight skin sloughing on the forehead during the chin lift-head tilt procedure while intubation, similar to our case which blistering occurred on the nose where facemask was applied. Detailed preoperative examination is crucial in EB patients. This detailed examination will reveal the difficulties for intubation before the procedure.

REFERENCES

1- Gache Y, Chavanas S, Lacour JP, Wiche G, Owaribe K, Meneguzzi G, Ortonne JP. Defective Expression of Plectin/HD1 in Epidermolysis Bullosa Simplex with Muscular Dystrophy. J Clin Invest 1996;97:2289-98.
2- Harris JC, Bryan RAE, Lucas VS, Roberts GJ. Dental disease and caries related microflora in children with dystrophic epidermolysis bullosa. Pediatr Dent 2001;23:438-43
3- Culpepper TL. Anesthetic Implications in Epidermolysis Bullosa Dystrophica. AANA Journal 2001;69:114-8.
4- Zidorio APC, Dutra ES, Leao DOD, Costa IMC. Nutritional aspects of children and adolescents with epidermolysis bullosa: literature review. An Bras Dermatol 2015;90:217-23.
5- Fine JD. Review:Inherited Epidermolysis Bullosa. Orphanet Journal of Rare Diseases 2010;5:12.
6- McGrath JA, Gatalica B, Li K, Dunnill MGS, McMillan JR, Christiano AM, Eady RAJ, Uitto J. Compound Heterozygosity for a Dominant Glycine Substitution and a Recessive Internal Duplication Mutation in the Type XVII Collagen Gene Results in Junctional Epidermolysis Bullosa and Abnormal Dentition. American Journal of Pathology 1996;148:1787-96.
7- Crowley KL, Shevchenko YO. Anesthetic management of a difficult airway in a patient with epidermolysis bullosa: A case report. *AANA Journal* 2004;72,261-263.
8- Lanier PA, Posnick WR, Donly KJ. Epidermolysis bullosa-dental management and anesthetic considerations: case report. Pediatric Dentistry 1996;12,246-249.
9- Prabhu VR, Rekka P, R, Swathi S. Dental and anesthetic management of a child with epidermolysis bullosa. J Indian Soc Pedod Prev Dent 2011;29:155-60.

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