



Case Report

Crown Lengthening Procedure in Epidermolysis Bullosa: A Case Report

Kamran Lahoopoor¹, Nazli Rabienejad¹, Sayed Ali Moosavi^{1*}

¹Department of Periodontology, School of Dentistry, Hamadan University of Medical Sciences, Hamadan, Iran

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***Corresponding author:**

Sayed Ali Moosavi,
Email: s.a.moosavi@protonmail.com



Abstract

Epidermolysis bullosa (EB) is a spectrum of conditions characterized by mechanical fragility and blistering of the skin. Individuals suffering from EB display a wide range of symptoms based on affected proteins in different organs and tissues in the body, including the craniofacial complex and the oral cavity.

In this case-report, a 22-year-old girl with Dominant Dystrophic Epidermolysis Bullosa is presented. She suffered no other medical complication and psychological examination was normal. Atraumatic extraction of hopeless teeth was performed and patient was referred for endodontic treatment of maintainable teeth. Surgical crown lengthening in anterior segments was performed in two sessions with specific precautions to avoid soft tissue trauma and irritation both during surgery and after it. Post-surgery healing was uneventful and acceptable. In conclusion, with a few precautions, surgical crown lengthening can be performed in these patients with minimal soft tissue trauma and optimal healing post-surgery.

Keywords: Epidermolysis Bullosa, Crown Lengthening, Surgery

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Introduction

Epidermolysis bullosa (EB) consists of a spectrum of conditions that are characterized by mechanical fragility and blistering of the skin (1). EB, which was first reported by Hebra (2), displays a tremendous genetic and clinical phenotype variety in its family of disorders. The main types of EB are intraepidermal (simplex EB), Junctional, dermolytic (dystrophic EB), and mixed (Kindler syndrome) EB (1).

EB symptoms are often manifested at birth or during the first year of life (3). Tremendous diversity is observed in involved organs and tissues and phenotypic severity in individuals suffering from EB disorders (4-6). Likewise, different EB types result in various craniofacial and oral manifestations with varying characteristics and severity (7,8). There can be significant morbidity and complications involving both soft and hard tissues of the craniofacial complex depending on the type of EB (1).

A specific molecular basis is currently known for 13 EB subtypes (1). The clinical phenotype of the disorder in affected individuals and the involved tissues and organs depends largely on the specific absent or abnormal proteins as a result of causative genetic mutations (9). For example, although type VII collagen is critical for maintaining the integrity of the skin and the oral mucosa, it is not essential in the development of the forming tooth bud. Therefore,

individuals with mutations affecting type VII collagen have normally developed dentition but may show severe oral soft tissue involvement. On the other hand, laminin 322 plays an essential role during tooth development, and hence, individuals with mutations affecting laminin 322 may demonstrate defects in the development of the enamel of their teeth (9).

Case Report

A 22-year-old girl was referred to the periodontology ward with the chief complaint of pain in her anterior teeth. The patient had dominant dystrophic EB, which is one of the mildest forms of dystrophic EB. Her parents were identified to be first-degree cousins. On routine medical examination, the patient appeared normal. She had a few blisters and multiple disfiguring scars on her body (Figure 1).

Psychologically, the patient appeared to have a normal cognitive function and was attending college. She was taking supplements and vitamins but not any medications. The radiological panoramic view represented numerous remaining roots in the maxilla and mandible (Figure 2).

The patient was very knowledgeable about her condition. She reported frequent intraoral bullae formation as a result of inflicted minor traumas. Minor dental procedures such as oral prophylaxis often caused bullae formation with





Figure 1. General appearance of the patient



Figure 2. Initial panoramic radiography

subsequent discomfort. Cutaneous involvement was limited, and bullae would form under heavier traction forces. Consultation with the patient's dermatologist conveyed no contraindications to periodontal surgery, with a recommendation to minimize the manipulation of soft tissues.

Oral evaluations revealed multiple ulcers on the buccal mucosa, tongue, and palate. The palate was dystrophic and had severe scarring due to chronic trauma and bullae formation (Figure 3).

The treatment plan consisted of the extraction of hopeless teeth, root canal therapy, crown lengthening, and fixed partial prosthesis. The treatment plan was presented and accepted by the patient.

Local anesthesia was obtained using 2% lidocaine with 1:80,000 epinephrine (Persocaine-E, Darou Pakhsh Pharmaceutical Mfg Company, Tehran, Iran). All hopeless roots were atraumatically extracted in two sessions (Figure 4).

The patient was referred to the endodontics ward to undergo root canal treatment of the remaining periodontally maintainable teeth. Four weeks after the completion of root canal therapy sessions (Figure 5), the patient underwent surgical crown lengthening.

For the crown lengthening of the anterior segment of



Figure 3. Initial intra-oral view



Figure 4. Extracted hopeless teeth

the maxilla, anesthesia was achieved using buccal and palatal supra-periosteal infiltration of lidocaine 2% with epinephrine 1:80000 (Persocaine-E). First, sub-marginal scalloped internal bevel incisions were made 1 or 2 mm apically from the remaining caries-free tooth structure; sufficient tooth structure would be available for a ferrule effect and consider leaving at least 2 mm of keratinized gingiva. Then, sulcular incisions were performed to detach the attachment apparatus from the root. Next, interdental incisions were made to detach the interdental tissue and the gingival collar from the bone. Full-thickness flap reflection was performed to expose the alveolar bone beyond the mucogingival junction. Bone contouring was mainly performed using a long-shank round diamond bur on a high-speed dental handpiece by utilizing water spray as a coolant. The two hopeless central incisors were atraumatically extracted, and final adjustments to the bony contour were made using hand instruments such as chisels and bone files. After ensuring a minimum of 4 millimeters of the available supracrestal tooth structure, all root surfaces were checked for the remaining bone spicules and widow peaks. Following achieving the optimal bone contour, the soft tissue was adjusted by trimming and thinning so that it optimally adapts to the underlying bone and allows for primary closure, except for the extraction sites. For wound closure, 4-0 absorbable polyglycolate-coated suture (Supabon, SUPA Medical Devices Company, Tehran, Iran) was selected, and closure was obtained using interrupted simple-loop sutures



Figure 5. Intra-oral pre-op view



Figure 6. Maxillary immediate post-op results

(Figure 6).

Crown lengthening of the mandibular anterior segment was performed after the initial healing of the maxillary mucosa. Anesthesia was achieved using lidocaine 2% with epinephrine 1:80000 (Persocaine-E) by means of bilateral incisive/mental nerve block and lingual infiltration. Moreover, incisions and flap reflections were performed as described previously. Bone recontouring was also conducted based on previous explanations. No teeth needed to be extracted in this region. Copious irrigation was performed after verifying sufficient available supracrestal tooth structure, and wound closure was achieved, as previously described, using 4-0 absorbable polyglycolate-coated sutures.

To minimize soft tissue irritation and manipulation during and after surgeries, a few steps were taken as follows:

- Incisions were performed sharply and continuously, accompanied by firm pressure to complete the incision in a single stroke.
- Tongue, cheek, and lip retraction were conducted using large-surface smooth blunt vaseline-coated instruments to avoid pin-point pressure on these tissues.
- Flap reflection was solely performed using large blunt periosteal elevators leaning directly on the bone.
- If there was a need for a flap to be held or secured in place (e.g., when a needle had to be passed through), it was attempted to be conducted using instruments applying force from inside the flap (i.e., connective tissue + periosteum surface), and the use of instruments which would cause pressure on the epithelial side (e.g., tissue forceps) was kept to a minimum.
- Absorbable sutures were selected to avoid an extra session of suture removal and the resulting soft-tissue irritation.
- No periodontal dressing was placed to minimize soft tissue irritation and trauma and avoid the possible separation of epithelium/bullae formation.

Post-operation instructions included general oral surgery post-op instructions with an emphasis on minimizing soft-tissue trauma and irritation. Medications

prescribed post-operatively included amoxicillin 500 mg three times per day for one week, ibuprofen 400 mg three times per day or as needed, and normal saline rinsing 3-4 times a day. Chlorhexidine mouthwash was prescribed as it would reduce the oral cavity microflora which may lead to less inflammation. The patient was revisited 3 days post-operation after each surgery to ensure proper wound healing. Figure 7 displays soft tissue healing was uneventful and without complications. The soft tissue condition at 1-month post-operation immediately after the cementation of two cast posts and before the prophylaxis session.

Discussion

Several researchers have reported various dental procedures performed on EB patients. However, to the best of our knowledge, periodontal plastic surgery and related complications in patients with EB are rarely mentioned in the literature. In dystrophic EB, blisters are found beneath the basement membrane within the dermis, exposing the underlying connective tissue (10-13).

The presented case demonstrated widespread dental caries that are directly related to the inability to maintain proper oral hygiene (14,15). Wright et al found that susceptibility to caries was significantly higher in EB patients (12). This might be due to the severity of the soft tissue ulceration and consumption of a soft diet rich in sucrose -necessary to provide sufficient energy intake- and the difficulty in mechanical removal of dental plaques due to their incapability in holding a toothbrush (3). The periodontal status of these patients could be well managed by accurate treatment and a good maintenance program (16). Comprehensive oral care instructions should be given to these patients as soon as possible, and they should be put in a preventive program. To enhance the oral clearance of food debris, an increased fluid intake during meals could be helpful (17). Water jets and water flossers, soft toothbrushes, and mouthwashes containing fluoride are recommended (3), all of which can help fight cavities. These patients tolerate the topical application of neutral sodium fluoride and fluoride varnish well (17). Chlorhexidine mouthwash may also help fight cavities (3,17). Sucralfate (the drug of choice for the treatment of



Figure 7. One month post-op results

active duodenal ulcers) has been proposed as an effective prophylactic and therapeutic agent for reducing pain and blistering in these patients (18).

Rapidly rupturing vesicles frequently develop in EB patients, leaving erosive surfaces on the mucous membranes of the lips and mouth. The other common oral manifestations of EB are vestibular obliteration, denudation of the tongue, microstomia, and ankyloglossia (3,17,19,20).

Ankyloglossia, severe scarring of the mouth and around the mouth, limited opening of the mouth, and vestibular obstruction of the mouth can impede or complicate dental care. Mouth ulcers are inevitable during dental treatment; however, they can be prevented by lubricating the mucosa with hydrocortisone cream, triamcinolone, or petrolatum before starting the procedure (21). Furthermore, retractors can be coated with petrolatum to minimize friction and soft tissue trauma.

Local anesthesia during outpatient dental treatment can also cause blisters. Therefore, it should be avoided whenever possible (20). The injections of local anesthetic solutions should also be slow and deep into the tissue to prevent mechanical damage to the tissue (3). Superficial injections can cause tissue ballooning and possible subsequent bullae formation.

The principles of surgical crown lengthening in EB patients do not differ from systemically healthy patients, but precautions and measures should be taken to minimize soft tissue irritation. These measures include but are not limited to sharp, continuous, and single-stroke incisions, using blunt instruments for flap retraction, flap retraction by applying force to the inner surface of the flap, avoiding tissue forceps whenever possible, using absorbable sutures, and avoiding periodontal dressings.

Conclusions

EB depicts a group of rare genetic disorders that involves the skin and mucous membranes by vesicle and bullae formation. The oral and dental manifestations of EB disrupt the patient's oral health and hygiene and exhibit a challenge for dental professionals. The administration of general anesthesia for patients with EB requires distinctive

attention and inclusive care. Surgical crown lengthening is possible in these patients with some precautions to minimize soft tissue irritation.

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Conflict of Interest Disclosures

The authors of this manuscript declare that they have no financial or other competing interests concerning this article.

Ethical Statement

Informed consent was obtained from the patient for the publication of this report.

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