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Case report

Recessive dystrophic epidermolysis bullosa (RDEB): Oral manifestation and management rules in oral surgery: A case report

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ABSTRACT

Introduction and importance: The recessive dystrophic epidermolysis bullosa is a severe form of hereditary epidermolysis bullosa characterized by deformities of the skin, blisters and erosions on the mucous membranes. Oral manifestations are frequent and extensive vary from small discrete vesicles to large bullae, associated with microstomia, ankyloglossia and a depapilled tongue.

The purpose of this case report is to describe oral health status of patient with recessive-dystrophic epidermolysis bullosa, and the measures that dentists should adopt with the purpose of provide an effective dental treatment.

Case presentation: We present a clinical case of a patient with recessive-dystrophic epidermolysis bullosa who underwent a bone regularization and whose follow-up was carried out until healing.

Clinical discussion: The management of patients with RDEB in oral surgery requires the adoption of an atraumatic technique and certain precautions to limit the formation of bullae and promote mucosal healing.

Conclusion: We can conclude that oral management of patients with recessive dystrophic epidermolysis bullosa presents a challenge for the oral surgeon and the management is generally multidisciplinary and relies essentially on the adoption of a preventive and atraumatic approach.

1. Introduction

Hereditary epidermolysis bullosa (HEB) is a rare genetically determined disorder of the skin and mucosal membranes, characterized by fragility of tissues and the development of blisters following minor or insignificant trauma of the skin or mucosal surfaces [1].

It is mainly caused by mutations of genes involved in the production of proteins which allow cohesion between the different layers of the skin. Traditionally, these diseases are classified into four groups according to the zone of cutaneous cleavage [2,3].

The dystrophic form of epidermolysis bullosa is inherited in both dominant and recessive forms. Clinically is characterized by deformities of the skin, blisters and erosions on the mucous membranes. The lesions also affect the oral cavity and vary according to the severity of the (RDEB) [4].

Through our article, we report a clinical case of a patient with recessive-dystrophic epidermolysis bullosa who underwent a bone regularization and whose follow-up was carried out until healing, which provide an update on the specificity of the management these patients in oral surgery. This case report was reported in accordance with the SCARE criteria [4].

2. Presentation of case

A 27-year-old patient was referred by the department of removable Prostheses for localized bone regularization. According to the medical records, the patient was diagnosed with recessive dystrophic epidermolysis bullosa with two of her brothers and underwent multiple surgeries under general anesthesia in her childhood.

It's important to note that the patient had severe anemia with a hemoglobin value of 7.8 g/dL, which put her at risk for bleeding after the intervention. The patient was referred to her treating physician for a blood transfusion. 15 days after the transfusion a complete blood count (CBC) was performed to objectify the improvement of the hemoglobin value.

Physical examination revealed multiple regional blisters and ulcerations in various areas of the body (Figs. 1, 2, 3). Her fingernails were absent, and her hands showed some degree of deformity (Fig. 4).

Extraoral examination revealed a limited mouth opening.

Intraoral examination showed:

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- the presence of a bullae on the left jugal mucosa with an oar diameter of 8 mm (Fig. 5), and denuded tongue with hemorrhagic bullae located in the base (Fig. 6).
- An alveolar exostosis in the right area of the alveolar ridge of the maxilla (Fig. 7).

The difficulties in performing adequate oral hygiene caused the destruction of almost all the teeth that were extracted afterwards. Only the 47 and 37 that persist in the oral cavity free of any carious lesion.

A comprehensive radiographic examination (panoramic radiographs) was accomplished to outline a logical and realistic dental treatment planning and no bone pathology was observed.

After intraoral examination and the evaluation of the panoramic radiograph band a treatment plan was established.

Subsequently, the patient underwent bone regularization with certain special precautions: first, local anesthesia was infiltrated slowly and gently to avoid trauma to the fragile oral mucosa, then the flap was detached with minimal pressure on the tissue and the bone ridge was regularized using a round ball bur mounted on a low-speed handpiece (Fig. 8). At the end the flap is repositioned, and simple sutures are made without tension on the edges.

We note the appearance of Mucosal sloughing immediately after the operation (Fig. 9).

An analgesic for pain control, an alcohol-free mouthwash, and a hyaluronic acid-based gel were prescribed postoperatively to promote healing. After 20 days the healing process is achieved, and the patient did not complain about any pain (Fig. 10).

3. Discussion

Hereditary epidermolysis bullosa (HEB) refers to a group of hereditary diseases which are characterized by blistering of tissues containing stratified epithelia. It is a rare disease which the incidence is estimated at 19.6 per 1 million live births (about 1:50.000) and the prevalence is 11 cases per million inhabitants. HEB affects all racial and ethnic groups, there is no gender predominance and often manifests at birth or during the first year of life [2,16].

Classification schemes of HEB were first introduced by Pearson in 1962. Since then, various consensus classifications have been published. The 2020 classification system recognizes four major types, 35 subtypes, and five other disorders with skin fragility [2,3,5].

The hallmark feature of inherited EB is mechanical fragility of the skin and the appearance of blisters and bullae. Other systemic manifestations are reported in the literature, including:

- Acral deformities, anemia, gastrointestinal complications, and cardiac damage… etc. [11–13].

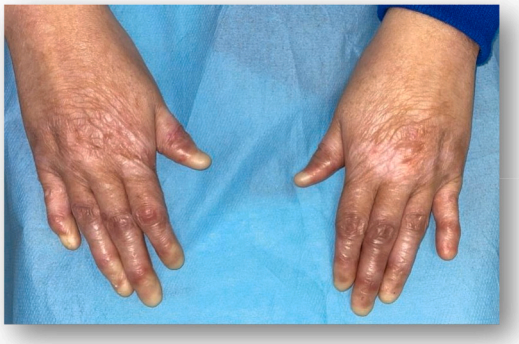


Fig. 4. View of the hands showing absence of nails, scarring and deformity.

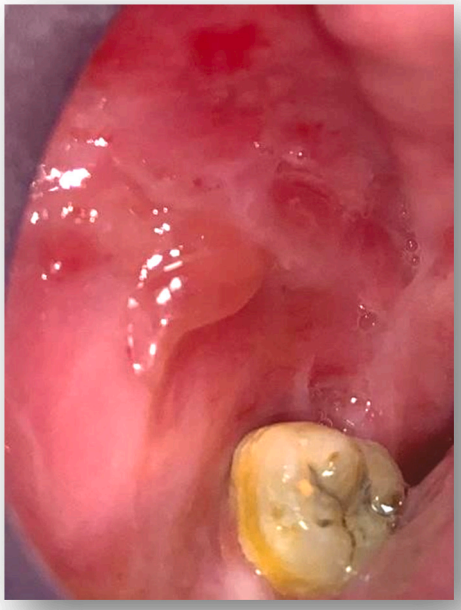
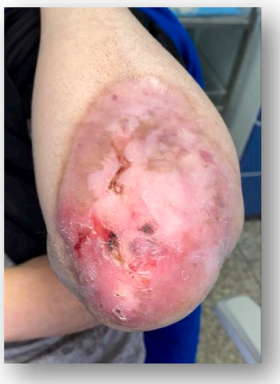
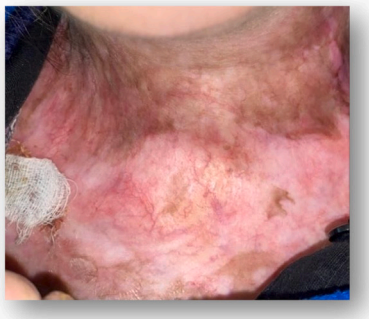
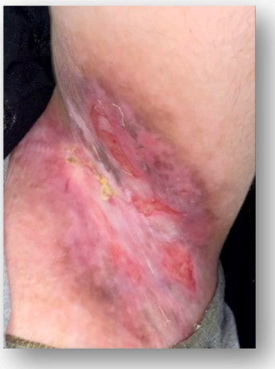


Fig. 5. Bullae on the buccal mucosa of a patient.







Figs. 1, 2, 3. Regional ulcerations and scarring throughout the body.

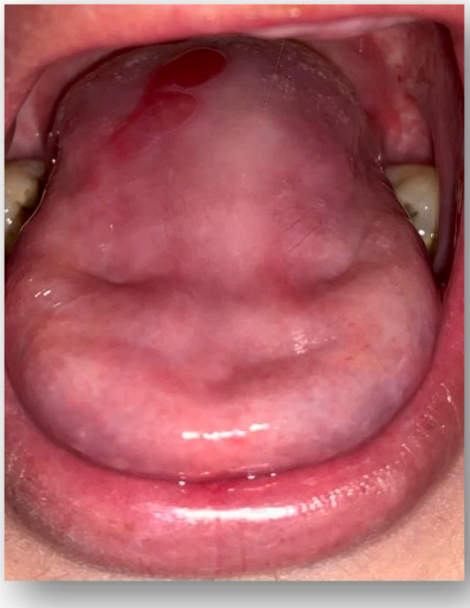


Fig. 6. Absence of tongue papillae.

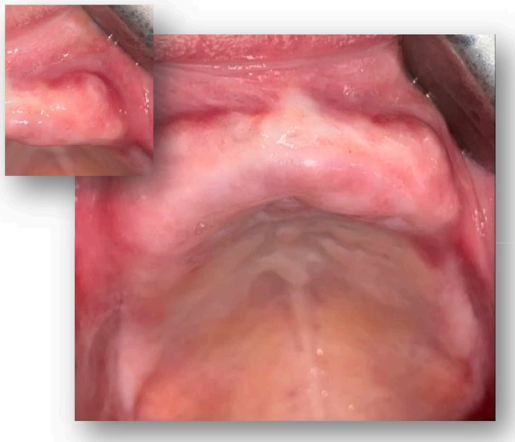


Fig. 7. Intraoral image showing the exostosis in the left maxilla.

Dystrophic EB is one of the major forms of EB and is characterized by a deep cleavage plane located at the level of the sub lamina densa which explains the severity of the cutaneous-mucosal manifestations in patients affected by the recessive form [9,10].

Oral manifestations of (RDEB) are frequent and extensive. Recurrent vesiculobullous lesions that range from small discrete vesicles to large bullae, microstomia, ankyloglossia, and a depapillated tongue, are often encountered. Squamous cell carcinomas may be the consequence of atrophic wounds that cannot heal properly. Oral tissue biopsies may be required when carcinoma is suspected [10,12,13].

In the reported case, the patient had most of these manifestations: blisters and ulcerations in various areas of the body (Figs. 1, 2, 3), deformity of fingers (Fig. 4), limitation of the mouth opening, and bullous lesions distributed on multiple regions of the oral mucosa (Fig. 5) associated with a depapilled tongue (Fig. 6).

The scientific literature regarding oral health care of people living with RDEB is relatively scarce. A preventive protocol is today's dental management approach of choice for patients with (RDEB) to reduce the risk of developing dental diseases [7,8].

The planning of surgical procedures including dental extractions involves contacting the treating physician and prescribing certain biological tests (CBC) to detect any hematological disorders (as anemia) that may complicate the dental surgery.

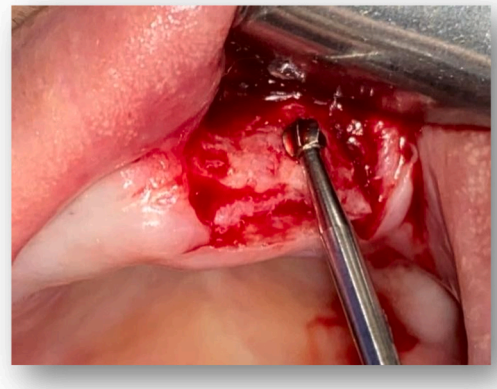


Fig. 8. Osteotomy using round ball bur mounted on a low speed handpiece.

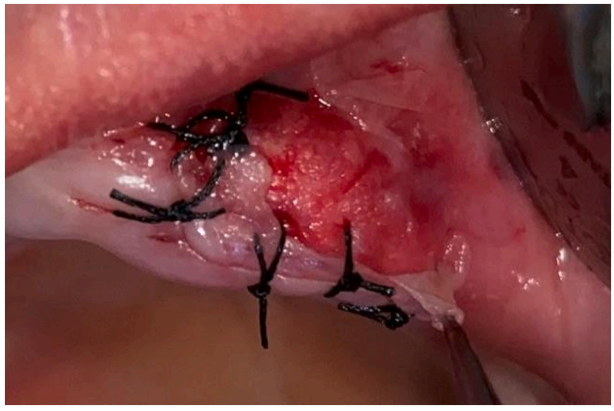


Fig. 9. Mucosal sloughing after dental surgery.

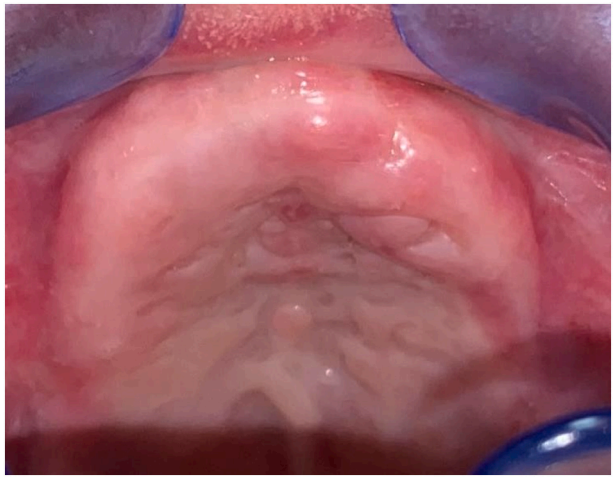


Fig. 10. Post-operative view 20 days after bone regularization, showing mucosal healing.

The use of postoperative antibiotics will depend on each individual case, and there is no need due to the patient's EB condition [9,15].

An atraumatic technique should be used throughout the procedure and certain precautions are recommended to limit bullae formation:

- Lubrication: Lips should always be lubricated with Vaseline or petrolatum or other appropriate lubricants before any procedure to reduce adherence and shearing forces that lead to lesions formation.
- Bullae: that occur during treatment should be drained with a sterile needle or by a cut with scissors to avoid lesion expansion because of fluid pressure (perforating the blisters contributes to accelerating the healing process).
- Instruments: Because of limited access, it is easier to use pediatric size instruments.
- Drug prescription should be adapted to the clinical situation, and for antiseptics, alcohol-free formulations are recommended for patients with oral lesions.

Healing of oral tissues occurs gradually after one to two weeks [10,14,15].

4. Conclusion

The oral management of patients with recessive dystrophic epidermolysis bullosa presents a challenge for the oral surgeon. The management is generally multidisciplinary and relies essentially on the adoption of a preventive and atraumatic approach.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Patient perspective

All patients were satisfied with the result after the intervention.

Provenance and peer review

Not commissioned, externally peer reviewed.

Ethical approval

Ethical approval is exempt at our institution.

Funding

N/A.

Author contribution

Dr. EL YACOUBI Oumayma designed the concept, analysed and interpreted the findings, wrote and reviewed the final paper under the supervision of Prof CHBICHEB Saliha.

Guarantor

El Yacoubi Oumayma.

Registration of research studies

N/A.

Declaration of competing interest

N/A.

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