Approach of a Pediatric Patient with Epidermolysis Bullosa: a Clinical Case

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Introduction

Epidermolysis Bullosa(EB) is a rare genetic disease characterized by a recurrent formation of blisters due to structural fragility of the skin and mucous membranes. It is caused by mutations in genes that alter the cohesion of the epithelial cells. It can be classified into four types: EB simplex, junctional EB, dystrophic EB (DEB) and Kindler syndrome. There is no predominance by sex or race. The most common clinical manifestations are blisters, although there may also occur changes in the shape and structure of teeth, ankyloglossia, susceptibility to tooth decay, limited mouth opening, among others.(1,2,3,4) In recessive dystrophic epidermolysis bullosa (RDEB) oral lesions appear with minimal trauma, requiring specific care in dental treatments.(5)

Description of the Clinical Case

- ✓ **General information and medical history:** caucasian male patient, followed since he was 5 years old. Diagnosed with RDEB at the age of 23 months old.
- ✓ **Chief complaint:** presence of tooth decay and difficulty in oral hygiene.
- ✓ **Clinical history and objective examination:** tooth 72 twinned (Fig.1), bullous and hemorrhagic lesions in the oral mucosa (Fig.2, 3, 4, 9), painful ulcers and skin scars that evolved over time (Fig.7, 8). High plaque index, tartar deposition, gingivitis, tooth decay (Fig.1, 5, 6, 10), left lateral crossbite (Fig.6) and mouth opening limitation.
- ✓ **Clinical procedure:** oral hygiene and dietary instructions, scaling, wear of cusps that traumatized tissues, and topical applications of fluoride. The treatment of decays and extraction of deciduous teeth that hadn't exfoliated. Patient undergoing treatment aimed at the cause. Appointments were scheduled quarterly, although given the patient's lack of cooperation, they were limited to urgent treatments.

Discussion

EBD can be inherited in an autosomal dominant or recessive manner.(4) In the present case, the mutation has maternal origin, and it is atypical, given the benignity of the phenotype.

Clinical and radiographic findings showed that we were facing a patient with a high risk of tooth decay. Brushing has always been very difficult. The patient was unable to use dental floss and didn't tolerate manipulation the oral cavity. As such, preventive measures were adopted. Treatments were sometimes postponed due to painful symptoms prior to the appointments. Currently, scaling is performed with *Guided Biofilm Therapy*®, which was well tolerated by the patient and didn't form new bullous lesions.

For a correct monitoring of these patients, dentists and caregivers must be aware of the preventive measures and recommendations shown in Table 1.

Preventive measures and guidelines

For patients	For dentists
○ Perform mouth opening and closing exercises 30min before the appointment. ○ Sit alone in the dental chair. ○ Use alcohol-free chlorhexidine mouthwash twice a day for 2 weeks, every 3 months. ○ Use a brush with a small head and soft bristles. ○ Soft and non-cariogenic diet.	○ Minimal manipulation of the oral cavity. ○ Local anesthesia should be administered deeply and slowly into the tissues. ○ Lubricate mucous membranes and instruments with vaseline. ○ Avoid pulling or applying air directly to tissues. ○ Place the aspirator on teeth and never on mucous membranes. ○ Polish restorations and cusps that traumatize the oral mucosa. ○ If new blisters appear, they should be drained with a sterile needle or scissors. ○ Topical applications of fluoride every 4 months. ○ Prescribe medication in oral suspension or syrup.

Table 1

Bibliographic references

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4.Krämer S, Lucas J, Gamboa F, Peñarrocha Diago M, Peñarrocha Oltra D, Guzmán-Letelier M, et al. Clinical practice guidelines: Oral health care for children and adults living with epidermolysis bullosa. Spec Care Dentist. 2020;40 Suppl 1(Suppl 1):3-81.
5.Brad W. Neville DDD, Carl M. Allen, Angela C. Chi. Oral and Maxillofacial Pathology. 4th ed: Saunders; 2015.

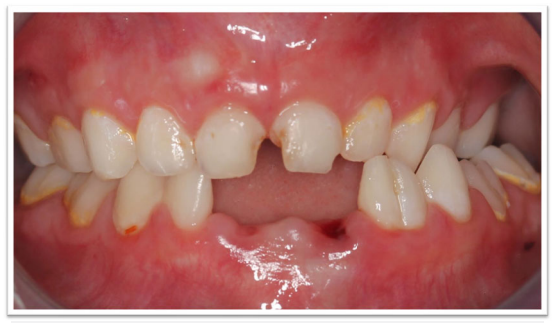

Figure 1: Presence dental plaque, decay and dental twinning (5 years old).

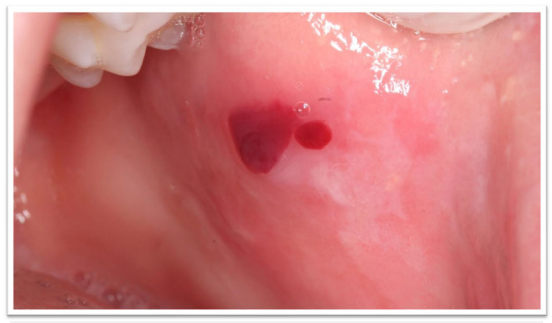

Figure 2: Hemorrhagic blister in the left buccal mucosa (5 years old).

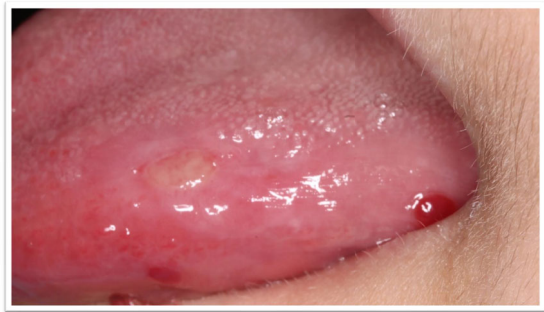

Figure 3: Bullous and hemorrhagic lesion on the left lateral border of the tongue (5 years old).

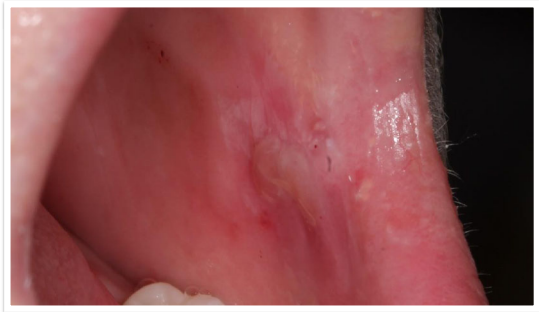

Figure 4: Blister in the left buccal mucosa (5 years old).

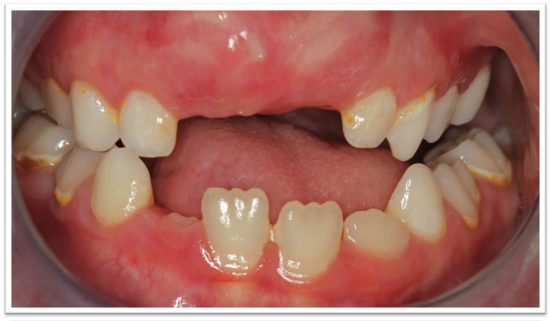

Figure 5 : Accumulation of dental plaque and tooth decay (6 years old).

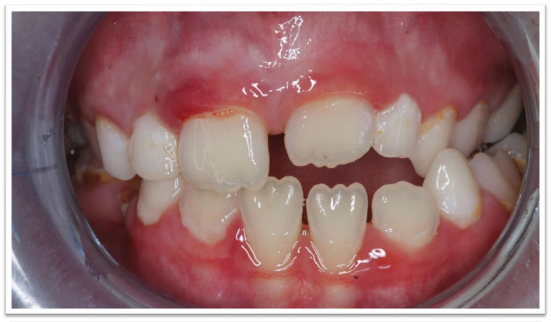

Figure 6: Gingivitis (7 years old).

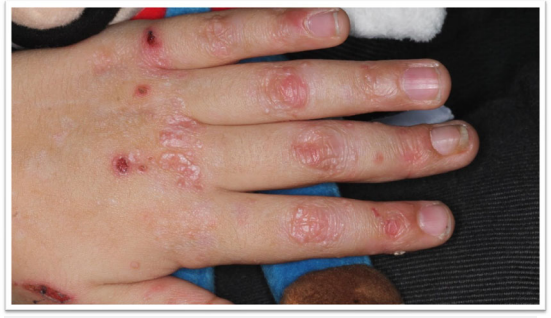

Figure 7: Cicatricial lesions and erosions on the hands caused by minimal trauma (8 years old).

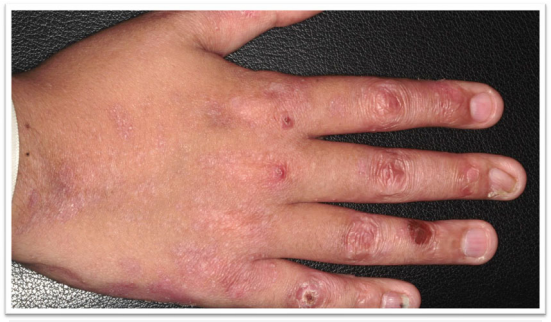

Figure 8: Cicatricial lesions. Signs of deformity in the hands are already observed (10 years old).

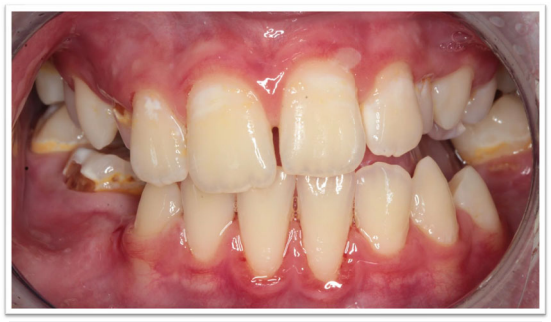

Figure 9: Blister on the gingival margin of the tooth 21 that didn't rupture after scaling (10 years old).

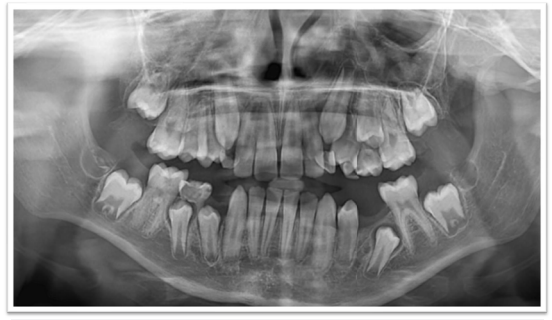

Figure 10: Panoramic X-ray (10 years old).

Conclusions

Considering the severe fragility of the oral mucosa in patients with EB, especially in the recessive dystrophic type, preventive dentistry plays a fundamental role in reducing the need for dental treatments and improving the quality of life of patients. It is essential that the dentist knows the necessary precautions he needs to take.