HEREDITARY EPIDERMOLYSIS BULLOSA AND PERIODONTAL REPERCUSSIONS

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Background:

Dystrophic epidermolysis bullosa (DEB) is characterized by a plane of cleavage below the lamina densa in the part of the dermis. It can be predominantly or recessively inherited, varying in severity, with considerable phenotypic overlap between types. Oral mucosal involvement is frequent, often severe in the recessive forms, less symptomatic or absent in the dominant forms.

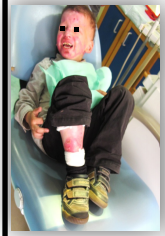


figure 1

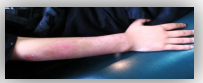


figure 2

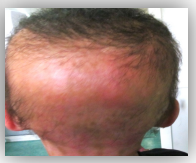
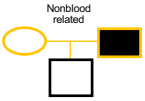


figure 3

Case report :

History: Yasser is the first child from nonblood related parents cut born by cesarean section. Was operated for an inguinal hernia around the age of 1 month



Family tree of a 3 years old boy

Diagnosis :DYSTROPHIC EPIDERMOLYSIS BULLOSA HALLOPEAU-SIEMENS

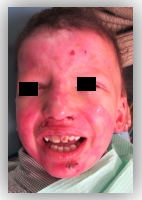


figure 4

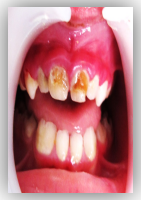


figure 5

Clinical manifestation:

- Erythematous scar appearance with grains of milia (figure 1) , Similar lesions on the left shoulder (figure 2)
- Crusty lesions of the scalp (figure 3)
- Post-bullous erosive lesions on the forehead (figure 4)
- Oral mucosal involvement (figure 5)
- First erupted teeth at 14 months

Discussion and Conclusion:

DEB recessive severe generalized is the most severe form of EBD, formerly called the Hallopeau-Siemens form. Extreme skin fragility is expressed from birth by bubbles and diffuse erosions, often hemorrhagic. Gingival lesions in DEB patients are a relatively common entity and may have multiple clinical aspects, emphasizing the need for thorough attention and awareness among dentists.

Fine JD, Johnson LB. Pseudosyndactyly and musculoskeletal contractures in inherited epidermolysis bullosa: experience of the National Epidermolysis Bullosa Registry, 1986-2002. J Hand Surg Br 2005;30:14-22.