Periodontal Aspects of Hereditary Epidermolysis Bullosa

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Hereditary epidermolysis bullosa, a rare dermatologic disease is discussed. Its oral manifestations and association with periodontal alterations, and possible systemic and periodontal treatment are described. An example of a case in which conventional periodontal therapy was effective is presented.

Key Words: oral pathology, hereditary epidermolysis bullosa, periodontitis, therapy.

Introduction

Epidermolysis bullosa syndrome is a mechanobullous disease which appears on the skin and mucosae as a result of trauma or irritation in the area of the lesions. This disease manifests itself in different forms, and is classified into groups and subgroups on the basis of these variations. A simple classification was proposed by Petges and Lecoultant (1936), and a subsequent classification by Gedde-Dahl and Anton-Lamprecht (1981) and Gedde-Dahl (1978). The latter and more complex classification system considered epidermolysis bullosa atrophicans generalisata mitis as a subgroup, and described alterations involving the lips, mouth, tooth enamel, and other body structures.

Hereditary epidermolysis bullosa refers to a group of inherited heterogenous bullous disorders in which lesions are the result of minimal trauma. A more accurate description of this condition would be mechanobullous disorder in which blister formation is associated with mechanical trauma. However, the term epidermolysis bullosa is the commonly accepted designation.

According to Briggaman (1983) the mechanobullous diseases characteristically present an inherent fragility of the skin and/or mucosae occurring on three levels:
intraepidermal, junctional, and dermal. The alterations observed at these different levels have been confirmed in the recent past through the use of ultrastructural and immunofluorescent studies utilizing antibodies of known specificity to epidermal-dermal junction components.

According to the Petges and Lecoulant (1936) classification system, the case described in this paper would be considered as epidermolysis bullosa dystrophica. Under this classification we could mention the work of Baptista (1928), Ramos (1931), Rossetti (1938), Rabello (1939), Mourão (1947), Castro (1950) and especially Bechelli and Belliboni (1954), in which epidermolysis bullosa dystrophica was reported.

The classification of the several varieties of epidermolysis bullosa was reviewed recently by Gedde-Dahl and Anton-Lamprecht (1981), and probably represents the most accurate system for defining the various mechanobullous disorders on the basis of their clinical, genetic, and ultrastructural aspects. Epidermolysis bullosa (EB) is divided into the following groups: EB simplex, EB junctional, EB dystrophic recessive, and EB dystrophic dominant. Each one of these groups may be further subdivided into several varieties, each of which represents recognized distinctive syndromes. Discussion of those disorders falls outside the scope of this paper.

The variety of epidermolysis bullosa atrophicans generalisata mitis reported by Schnyder and Anton-Lamprecht (1983) belongs to the group of newly recognized syndromes. This subgroup involves patients who present blistering at birth but whose condition tends to improve as they grow older. Cutaneous atrophy appears more frequently in the areas of repeated blistering, but other features present in EB dystrophica are absent. Involvement of the lips presents structural defects in some patients. All of the patients affected by this type of EB exhibit several degrees of nail alteration. The condition is inherited as an autosomal recessive Mendelian trait.

Oral manifestations of epidermolysis bullosa dystrophica have been described by Drury and Prieto (1964), Haas (1968), Schow and Fay (1968), Arwill et al. (1965), Kaslick and Brustein (1961), and Komori et al. (1966). In the periodontal literature, Levy et al. (1969) reported a case diagnosed at that time as epidermolysis bullosa dystrophica where dental treatment consisted of extraction of all permanent teeth and construction of full dentures. We find our case interesting because besides being the only one reporting periodontal alterations in a patient suffering from epidermolysis bullosa atrophicans generalisata mitis, conventional periodontal therapy was successful.

Clinical findings

A 19-year old female came to the Department of Periodontology of the School of Dentistry of the Federal University of Rio de Janeiro with a complaint of generalized gingival bleeding. The patient's medical history revealed that she had been suffering from several clinical problems, such as abnormalities of the nails (Figure 1), which had led to a tentative diagnosis of pachyonychia congenita, frequent diarrhea since childhood, persistent anemia, dilatation of the esophageal mucous membrane, and palmar-plantar blistering following mild trauma.
Oral examination revealed high plaque index, severely inflamed gingiva that bled easily on touch (Figure 2) and periodontal pockets varying in depth from 2 to 6 mm, with the deepest pockets found around the upper and lower incisors. The incidence of dental caries was high, and many teeth had been extracted. Anterior overbite as well as diastemata, particularly between the maxillary incisors, were also noted (Figure 3). In the posterior region of the mandible, a sizeable gingival enlargement extending from the left second bicuspid to the left second molar was noted, the first molar being absent. The mandibular right molars were also absent, and the patient reported that they had been extracted following similar gingival enlargement.

The radiographic examination revealed areas of horizontal bone loss, especially on the distal surface of the lower right lateral incisor and between the upper central incisors (Figure 4).

**Histopathological examination**

Two fragments of mandibular gingiva were obtained for histologic examination at the Oral Pathology Laboratory. They exhibited the following macroscopic features:
Figure 2 - Pre-operative photographs. Note the periodontal condition, dental caries and enamel hypoplasia on the upper left central incisor. R, Right, L, Left.
whitish-yellow in color, firm consistency, each measuring 1.3 x 1.2 x 6.0 cm. Both fragments were hemi-sectioned, embedded in paraffin, cut in sections of 6 μm, and stained with hematoxylin and eosin.

This histologic examination revealed several features. Massive chronic inflammatory infiltrate was evident in some sections, and most of the areas exhibited extensive ulceration in the surface bullae, as well as clefts at the epithelium-connective tissue interface, thereby indicating a subepithelial bullous lesion (Figure 5). The inflammatory infiltrate was composed of mononuclear cells, few macrophages, plasma cells and predominantly lymphocytes. The inflammatory infiltrate was more prominent underneath the ulcerated areas. A few inflammatory cells were seen within the bullae.

Treatment

Systemic treatment consisted of medication and improved diet to alleviate the patient's anemia, besides complementary instructions to preclude trauma and the resultant appearance of new blisters, guidance concerning proper care in cleaning occasional blisters, and plastic surgery of the nails.
Dental care consisted of periodontal treatment, including instruction in oral hygiene, plaque control, quadrant by quadrant scalings and root planing, crown polishing, and fluoride application. After this phase of treatment, the patient began the maintenance phase and returned every three months for prophylaxis. At a recent reevaluation (Figure 6), new pocket measurements were performed. Generalized pocket reduction had occurred, with the exception of the distal surface of the lower right lateral incisor, where a 5-mm pocket remained. A modified Widman flap was performed to reduce the remaining pocket. Following basic periodontal treatment, the patient was referred to the Departments of Endodontics and Operative Dentistry for completion of her dental treatment.

Discussion

This case is remarkable not only because very few cases have been reported describing the oral manifestations of this rare disease, but also because it responded well to conventional periodontal therapy. From this observation one can speculate that the combination of bacterial plaque and dermatologic disease provoked exacerbated periodontal destruction and that the removal of bacterial plaque was sufficient to reverse
Figure 5 - Subepithelial blistering observed on the histological examination of the gingival biopsy.
Figure 6 - Improved clinical aspect at reevaluation, requiring further therapy to reduce remaining pockets and to improve gingival architecture.
or at least to stop further periodontal breakdown. Despite the patient's age (19 years old), the prognosis appears to be favorable since she has responded well to therapy and at present has clinically normal gingiva with no bleeding and no pockets. The patient has been seen every three months for 2 years for maintenance, and follow up evaluations will continue to be scheduled.

This case also underscores the vital importance of the physician-dentist relationship, since the diagnosis was arrived at based on the histopathological findings of the gingival biopsy.

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