A Proposed Classification to Identify the Oral Manifestations of Genodermatoses

Sir,

Dermatology, being the specialized study of skin diseases, comprises of an important subdivision of the practice of medicine not only because of the various primary diseases affecting the skin, but also because of the common cutaneous manifestations of deeper visceral or systemic diseases.\(^1\)

There are several dermatologic disorders that have a genetic etiology or a genetic predisposition. Such genetically determined skin disorders are termed as “genodermatoses” (geno: genetic + dermatoses: skin lesions).\(^2\)

A genodermatoses can be defined as “a cutaneous phenotype caused by a single mutation, which may be a point mutation, deletion or a chromosomal aberration”.\(^3\)

The epidermis of the skin and the amelodentinal (the enamel and dentine) components of the teeth are derived from a common embryologic neural origin of the ectoderm. As a result, there exist many primary cutaneous diseases which find their manifestations in the oral cavity affecting the oral mucosa and dentition.\(^4\)

Therefore, it is of utmost importance for a dentist to recognize that not only some dermatoses exhibit concomitant lesions of the oral mucous membranes but also manifestations of some diseases may be preceded by oral lesions.

There are a plethora of genodermatoses with prominent and characteristic manifestations in both hard and soft tissues of the oral cavity. These genodermatoses have been well identified and classified. Although oral manifestations of genodermatoses have been mentioned by various authors, a comprehensive classification is required to segregate the manifestations of genodermatoses in oral cavity.\(^4,5\) This can enable dentists and dermatologists or any clinician, to identify the underlying genetic dermatopathy.

We hereby, propose a classification for oral manifestations of genodermatoses. We have attempted to accrue various genodermatoses in one classification suggesting their manifestation in hard and soft tissues of the oral cavity.\(^6,7\) We have also included the diseases with multiple manifestations in both teeth and oral mucosa.

Through this classification, we bring to light the various genodermatoses with oral manifestations which usually go otherwise unnoticed. Therefore, we are identifying and shedding light on the significance of oral genodermatoses which can aid in early diagnosis of the disease.

Proposed classification of oral genodermatoses

1. Genodermatoses affecting teeth and dentition
   - Ichthyosis
   - Sjogren-Larrson syndrome
   - Incontinentia pigmenti
   - Ehlers Danlos syndrome
   - Focal dermal hypoplasia syndrome
   - Gardner syndrome
   - Ectodermal dysplasia
   - Hyperimmunoglobulin E syndrome (Job syndrome)

2. Genodermatoses affecting periodontium and gingiva
   - Ichthyosis
   - Sjogren-Larrson syndrome
   - Papillon Lefevre syndrome
   - Tuberous sclerosis
   - Chediak-Higashi syndrome
   - Ehlers Danlos syndrome
   - Focal dermal hypoplasia syndrome

3. Genodermatoses affecting oral mucosa
   - Darier's disease
   - Neurofibromatosis type 1 and 2
   - Chediak-Higashi syndrome
   - Ehlers Danlos syndrome
   - Lipid proteinosis
   - Focal dermal hypoplasia syndrome
   - Multiple hamartoma syndrome (Cowden syndrome)
   - Pachonychia congenita
   - Epidermolysis bullosa
   - Multiple endocrine neoplasia syndrome
   - White sponge nevus

4. Genodermatoses affecting jaw bones and facies
   - Mccune-Albright syndrome
   - Ehlers Danlos syndrome
   - Marfan syndrome
   - Focal dermal hypoplasia syndrome
   - Gardner syndrome
   - Basal cell nevus syndrome
   - Orofacial digital syndrome type I

5. Genodermatoses causing pigmentation of oral mucosa
   - Carney complex
   - Neurofibromatosis type 1 and 2
   - Mccune-Albright syndrome
   - Lipid proteinosis
   - Pseudoxanthoma elasticum
   - Peutz-Jeghers syndrome
   - Congenital erythropoietic porphyria
   - Hypomelanosis of Ito
   - Sturge-Weber syndrome
   - Hereditary hemorrhagic telangiectasia syndrome

6. Genodermatoses with malignant potential
   - Xeroderma pigmentosum
   - Dyskeratosis congenita
REFERENCES


