Dental manifestations of dermatologic conditions

Anatoli Freiman, MD, FRCPC,[a] Daniel Borsuk, MD,[b] Benjamin Barankin, MD, FRCPC,[c] Geoffrey H. Sperber, DMD, PhD,[d] and Bernice Krafchik, MD, FRCPC[a]
Toronto, Ontario; Montreal, Quebec; and Edmonton, Alberta, Canada

Background: Cutaneous disorders can be associated with a wide variety of dental manifestations that should be familiar to dermatologists.

Objective: We sought to describe the development of the teeth, explain current dental terms, and review the dental manifestations of some dermatologic conditions.

Methods: A MEDLINE search (1966-May 2007) was performed to find relevant articles pertaining to dental manifestations of dermatologic conditions.

Results: Dental manifestations are associated with a wide variety of skin diseases that include genetic, infectious, inflammatory, and immune disorders.

Limitations: The review is broad and focuses on commonly described manifestations.

Conclusions: An appreciation and understanding of dental signs can aid in the diagnosis and treatment of many skin conditions. (J Am Acad Dermatol 2009;60:289-98.)

The common embryologic neural origin of the ectoderm includes the epidermal layer of the skin and the amelodentinal (the enamel and dentine) components of the teeth that result in a variety of conditions affecting both skin and dentition. The diseases derived from these components and neuroectodermal mesenchyme are named "neurocristopathies," signifying any disease arising from maldevelopment of the neural crest.

This article provides an overview of the epithelial elements of the mouth and dentition, a glossary of dental terms, and discusses a variety of conditions that affect the teeth and the skin that have been reported in the literature from 1966 to 2007. The diseases include the following: congenital erythropoietic porphyria (CEP), congenital syphilis, ectodermal dysplasia (ED), epidermolysis bullosa (EB), Gardner syndrome, hypomelanosis of Ito (HI), incontinentia pigmenti (IP), Langerhans cell histiocytosis (LCH), Naegeli-Franceschetti-Jadassohn syndrome, nevoid basal cell carcinoma syndrome (NBCCS), Papillon-Lefèvre syndrome (PLS), primary immune deficiency syndromes, Sjögren-Larsson syndrome (SLS), and tuberous sclerosis (TS).

Abbreviations used:

CEP:	congenital erythropoietic porphyria
EB:	epidermolysis bullosa
ED:	ectodermal dysplasia
HI:	hypomelanosis of Ito
IP:	incontinentia pigmenti
LCH:	Langerhans cell histiocytosis
NBCCS:	nevoid basal cell carcinoma syndrome
PLS:	Papillon-Lefèvre syndrome
SLS:	Sjögren-Larsson syndrome
TS:	tuberous sclerosis

Dermatologists should be aware of dental abnormalities that affect the number, structure, and color of teeth, as well as the degree of mineralization and caries susceptibility of enamel and dentine that are present in conditions associated with skin disease.

EPITHELIUM OF THE ORAL MUCOSA, AND TOOTH DEVELOPMENT

The tissue of the oral cavity is lined by a mucous membrane that consists of two layers: epithelium

From the University of Toronto,[a] University of Montreal,[b] The Dermatology Centre, Toronto,[c] and University of Alberta.[d]
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Reprint requests: Anatoli Freiman, MD, FRCPC, Division of Dermatology, University of Toronto, 131 Bloor St W, Suite 703, Toronto, Ontario, Canada, M5S1S3. E-mail: anatoli.freiman@gmail.com.
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and connective tissue. This tissue is extremely mobile, permitting free movement of the cheeks and lips. In other areas it serves as an organ of taste. The mouth is defined as the area that runs posteriorly across the roof of the oral cavity in a horizontal line just beyond the junction of the hard and soft palate, to a corresponding line on the back of the tongue. Histologically the oral mucosa is classified into 3 types: masticatory, which has a heavily keratinized epithelium; a lining mucosa, which is not keratinized and performs the function of protection; and a specialized mucosa that covers the surface of the tongue. The latter contains taste buds and papillae consisting of hyperkeratinized epithelium. Masticatory mucosa is commonly subdivided into the orthokeratotic hard palate and the parakeratotic gingiva, knowledge of which is important when reading histologic slides. The hard palate has a keratinized surface and the soft palate and buccal mucosa is parakeratinized. The floor of the mouth is lined by a nonkeratinized mucosa. It is commonly held that the lining mucosa of the cheeks, lips, floor of the mouth, and undersurface of the tongue are nonkeratinized although their stratum superficiale may show a tendency toward parakeratosis depending on functional and pathological conditions. In addition, the oral epithelium contains melanocytes, Langerhans cells, and Merkel cells.

Teeth comprise 20% of the surface area of the mouth with the upper teeth occupying more space than the lower. They pierce the oral mucosa and are the only structures in the human body that pierce the epithelium.

To fulfill their function of chewing, the teeth need to be hard. This is provided by a hard, inert, acellular enamel, formed by epithelial cells and supported by the less mineralized but more resilient and hard connective tissue dentin. The latter is formed and supported by the soft connective tissue central pulp. In mammals teeth are anchored to the jaw by connective tissue supporting tissue consisting of cementum, a periodontal ligament, and alveolar bone. This provides enough flexibility to withstand the forces of mastication. In human beings there are two successive rows of teeth to fill the growing mouth. The first are called primary or deciduous teeth followed by the secondary or permanent teeth. Anatomically the tooth consists of a crown and a root, and although teeth vary considerably in shape and size, histologically they are all similar.

Enamel is the covering of the tooth and is the most highly mineralized tissue in the body. It consists of 90% inorganic hydroxyapatite crystallites. Cells responsible for the formation of the enamel are called ameloblasts. They cover the entire surface of the enamel as it forms, but are lost as the tooth emerges. Dentin, which is avascular, forms the bulk of the tooth, supports the central pulp chamber, and compensates for its brittleness. The pulp chamber is enclosed by dentin and is filled with a soft connective tissue.

Prenatal development is divided into 3 successive phases; the first begins at fertilization and ends at 4 weeks. It is unusual to see congenital defects this early, as a severe defect would result in loss of the embryo. The second phase from 4 to 8 weeks is characterized by the differentiation of all major external and internal structures. It is in this phase that many congenital defects develop. From 8 weeks to term, growth and maturation take place and the embryo is called a fetus.

During tooth bud development, neural crest-derived mesenchyme cells provide the dentine-secreting odontoblasts and the dental pulp, while discrete segments of the oral epithelium differentiate into the enamel-secreting ameloblasts. The ectodermally derived components of the dentition comprise the outer enamel cap of teeth and the inner neural crest-derived dentine.

In the last 15 years tooth development has been increasingly understood at the gene level and the number of recognized genes determining the position, shape, and/or number of teeth is increasing rapidly. All these genes exhibit important functions in cell communication and are the most important mechanism driving embryonic development.

GLOSSARY OF DENTAL TERMS PERTINENT TO SKIN CONDITIONS

Amelogenesis: the embryologic production of dental enamel by ameloblasts

Anodontia: congenital absence of teeth

Caries: a demineralization of the tooth surface caused by bacteria

Cementum: the thin layer of calcified tissue covering the roots of teeth and formed by cementoblasts

Crown: the area of the tooth that is visible

Cuspids (*canines*): third teeth from the midline

Deciduous teeth (*primary teeth* or *baby teeth)*: first set of (usually) 20 teeth in childhood; these are replaced later in life with 32 permanent teeth

Dentine: the major hard tissue component of teeth formed by odontoblasts, positioned between the enamel or cementum, and the pulp

Enamel: a hard, translucent layer composed of inorganic calcium phosphate crystals that covers and protects the dentine of the crown of the tooth; it is the hardest substance produced by vertebrates and is secreted by ameloblasts

Gingivitis: inflammation of the gums characterized by redness and swelling induced by bacteria

Hypodontia: congenital absence of one or more teeth, also known as oligodontia

Impaction: blockage of tooth eruption

Incisor: a tooth for cutting or gnawing; located in the front of the mouth in both jaws

Macrodontia: enlarged teeth

Malocclusion: malposition and/or incorrect relation between the mandibular and maxillary teeth

Marginal gingiva: the terminal edge of gingiva surrounding the teeth in collarlike fashion

Microdontia: decrease in the normal size of teeth

Molar: the last 3 upper and lower teeth on both sides of the mouth

Odontoma: a benign tumor of odontogenic origin

Periodontitis: a collection of inflammatory diseases affecting the tissues that surround and support the teeth; caused by bacteria and the host response to the inflammation

Periodontium: the specialized tissue that surrounds and supports the teeth between the cementum of the tooth and the alveolar bone; it consists of connective tissue and periodontal fibers

Polydontia: supernumerary teeth

Premolar: a tooth having two cusps or points; located between the incisors and the molars

Pulp: the inner substance of the tooth containing blood vessels, lymphatic and nerve tissue

Talon cusp: a developmental anomaly of tooth shape seen on the lingual surface of anterior teeth

DERMATOLOGIC CONDITIONS WITH DENTAL MANIFESTATIONS

Congenital erythropoetic porphyria

CEP is a rare autosomal-recessive disorder first described by Günther in 1911.[1] The defect is in heme biosynthesis, resulting from an inborn error of porphyrin metabolism. The clinical presentation includes hemolytic anemia, photosensitivity (manifested as blistering of the skin), skin fragility, mutilating scarring, hypertrichosis and hyperpigmentation, and deposition of red-brown pigment in the bones and teeth.[2] The pigments are an accumulation of the isomer I porphyrinogens, which are spontaneously oxidized to water-soluble photosensitizing porphyrins with a reddish hue. The red appearance of the teeth (erythrodontia) in individuals with CEP, combined with an increased hair growth and necessity to only venture outdoors at night to avoid photosensitivity, have given rise to the legend of the werewolf[3] (Fig 1).

The dental features of CEP are helpful in establishing the diagnosis. The oral mucosa is pale and the teeth have a red to maroon color (erythrodontia).[4]

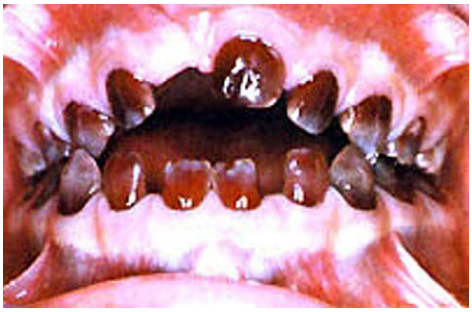

Fig 1. Porphyria; red discoloration of teeth. Courtesy of Marquette University School of Dentistry.

Incisors are almost completely stained, whereas the canines are colored only at the cusp tips and the molars vary in discoloration. The dental discoloration is thought to be caused by the affinity of porphyrins for calcium phosphate in the teeth.[2] Dental treatment of patients with CEP consists of aesthetic measures to mask the discoloration.

Congenital syphilis

Congenital syphilis arises from transplacental fetal infection with *Treponema pallidum* acquired during pregnancy from an untreated mother. The disease is divided into an early stage that usually occurs before 3 months, but may be seen up to 2 years, and late-stage disease that occurs after 2 years. Early clinical manifestations are often absent at birth and appear around 2 to 6 weeks. Cutaneous findings are seen in 38% of infants and present with a red macules and papules, papulosquamous eruption, or a desquamating dermatitis. A diagnostic sign in early syphilis is hemorrhagic bullae on the palms and soles. Rhagades occurs early but persists into late childhood and adult life. It consists of fine linear lines seen periorificially. Mucocutaneous lesions; snuffles (rhinitis); mucous patches that are present on the lips, mouth, tongue, and palate; and condylomata lata mainly in the anogenital area and at the angles of the mouth are characteristic. Prematurity, growth failure, bony involvement, hepatosplenomegaly, and jaundice are often seen as part of the multisystem disease. Pneumonia, enteritis, pancreatitis, nephritis, edema, ascites, uveitis, chorioretinitis, glaucoma, aseptic meningitis, and hematologic abnormalities may all be present in severely ill infants.[5]

The signs of late congenital syphilis affect every organ system including the eyes; interstitial keratitis and optic atrophy; the bones - frontal bossing, short maxilla, high palatal arch, saddle nose, saber tibias, scaphoid scapula, palatal perforation, and Clutton joints; the ears - eighth cranial nerve deafness; and the central nervous system - mental delay,

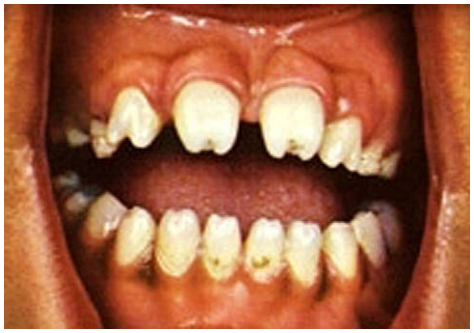

Fig 2. Syphilis; notched incisors (Hutchinson teeth). Courtesy of Marquette University School of Dentistry.

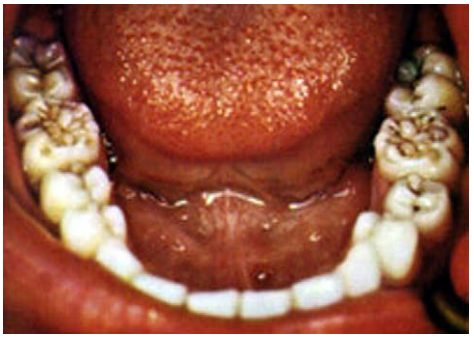

Fig 3. Syphilis; mulberry molars. Courtesy of Marquette University School of Dentistry.

convulsive disorders, paresis, and paralysis.[5] Gummas may be seen in the later stages.

Notwithstanding the many manifestations of late congenital syphilis, the dental signs are easily identified and may help to expedite the diagnosis.[6] Late congenital syphilis affects the amelogenesis of the molars and incisors. Both Hutchinson teeth and mulberry molars are seen in about 65% of patients. Hutchinson incisors are named after Sir Jonathan Hutchinson who first associated the defect with congenital syphilis in 1858.[7] These characteristic teeth present at around 6 years; they are centrally notched, widely spaced, peg-shaped upper permanent central incisors (Fig 2). The triad of Hutchinson teeth, interstitial keratitis, and sensorineural hearing loss is pathognomonic of congenital syphilis. Patients with congenital syphilis may also have mulberry molars, which are first molars dwarfed by a small occlusal surface, and are characterized by roughened lobulated hypoplastic enamel leading to caries.[8] The surface has numerous poorly formed cusps surmounting a dome-shaped tooth, which is considerably narrower at the grinding surface than at its base (Fig 3). Management of the dental abnormalities can hide the obvious notched teeth.

Ectodermal dysplasias

EDs represent a large group of hereditary conditions characterized by congenital defects of one or more ectodermal structures including skin appendages. The original constructional theme encoded in ectoderm diverges into epidermis, hair, sweat and milk glands, and the mineralized crystalline anvils of teeth, under the direction of local signals emanating from the underlying mesoderm. The intimate origins of these diverse ectodermal structures account for the wide spectrum of dysplasias. Clinically the hair (hypotrichosis, partial or total alopecia), nails (dystrophic, hypertrophic, or abnormally keratinized), teeth (enamel defects or absence), and sweat glands (hypoplastic or aplastic) are usually affected.[9] An estimated incidence of ED is about 7 in 10,000 births and all mendelian modes of inheritance have been reported.[10] Of more than 190 EDs described, the molecular basis has been elucidated for more than 30 of them.[11]

Dental defects represent a core clinical feature of many EDs: anodontia, polydontia, dysplastic teeth, retained primary teeth, deficient enamel development (amelogenesis imperfecta), dentine deficiency (dentinogenesis imperfecta), and underdevelopment of the alveolar ridge.[12] In some EDs, the number of erupted teeth is reduced, the spacing of the teeth disrupted, and the peridontium affected. In one case-controlled study of 68 persons with oligodontia, 57% had disturbances in hair, nails, and/or sweat production in addition to defective teeth, and were classified as having ED.[13] Disturbance of the enamel matrix may occur, making the teeth more susceptible to caries, and altering the shape of the teeth, leading to a pegged appearance and additional accessory cusps.

X-linked hypohidrotic ED (Christ-Siemens-Touraine syndrome) is the most frequent form; the diagnosis is usually made with the identification of hypotrichosis, characteristic facial features, hypohidrosis (and more rarely anhidrosis), and teeth abnormalities. The nails are usually normal. Abnormalities in the development of tooth buds result in hypodontia and peg-shaped or pointed teeth.[14] The hypodontia varies in each case, but usually only 5 to 7 permanent teeth are present, the teeth are smaller than average, and the eruption of teeth is often delayed[15] (Fig 4). The extent of hypodontia may be useful in assessing the severity of the disease, and is best done with dental radiographs.[16] It is extremely important for children with ED to be seen by a dentist early. Radiographs after 2 years reveal the state of the permanent teeth and allow children to be fitted with a plate early; this is helpful for both aesthetic reasons, and to maintain the alveolar ridge, allowing for later tooth implantation.

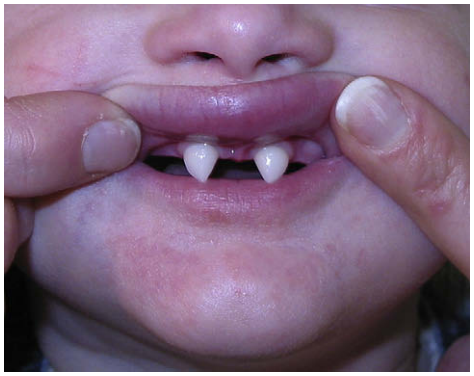

Fig 4. Ectodermal dysplasia; oligodontia and peg-shaped incisors.

Tooth and nail syndrome (Witkop syndrome) is another type of autosomal-dominant ED associated with specific dental findings. The primary dentition is usually unaffected, although the teeth may be small or peg-shaped, whereas the secondary dentition is often partially or completely absent.[17]

Oculodentodigital dysplasia is a rare autosomal-dominant subtype of ED, characterized by bilateral microphthalmos, nose malformations, hypotrichosis, syndactyly, and dental abnormalities particularly enamel hypoplasia.[18]

Dental treatment is often necessary in patients with some forms of ED and some children may need dentures as early as 2 years of age.[19] It is important to seek dental advice early as maintenance of the alveolar ridge is important for later dental intervention. Prosthetic teeth are implanted in adults for mastication and speech. Importantly, aesthetic dental interventions in patients with ED and malformed teeth and malocclusion helps with the development of a positive self-image and overall oral health.[20] A treatment protocol for meeting patients' functional and aesthetic needs as they grow into adulthood has been described by Hickey and Vergo.[12]

Epidermolysis bullosa

Hereditary EB includes a heterogeneous group of genetic bullous disorders characterized by blister formation in response to mechanical trauma. There are 3 main types of EB: simplex, junctional, and dystrophic, and numerous subgroups of these main types. The latter two cause skin breakdown and scarring and have associated organ dysfunction. Severe oral involvement often occurs in EB, leading to alterations in the soft and hard tissues. There is a risk of oral and esophageal carcinoma development in the chronically scarring disease of dystrophic EB of Hallopeau. Oral and dental manifestations and treatment of patients with EB have been comprehensively reviewed by Wright et al.[21]

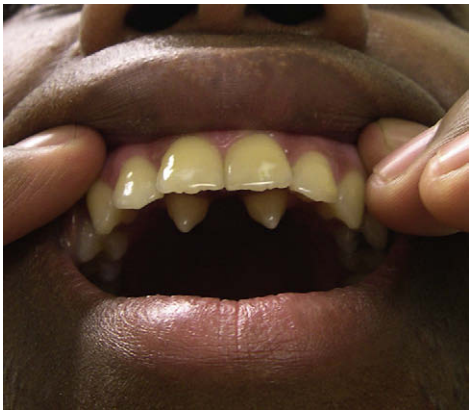

Fig 5. Gardner syndrome; supernumerary teeth. Courtesy of Douglas Hoffmann, MD, www.dermatlas.org.

Enamel hypoplasia is present in all forms of junctional EB causing pitting of the surfaces of all primary and permanent teeth. If untreated, teeth are lost during childhood as a result of caries.[22] Generalized enamel hypoplasia is limited to junctional EB,[21] whereas the prevalence of dental caries is significant in both junctional and recessive dystrophic EB.[22] The latter is also typically associated with significant mucosal blistering and soft tissue scarring.

Appropriate interventions and early dental therapy may help prevent destruction and loss of the dentition. Early referral of patients with EB for dental evaluation is important. Aggressive preventive measures and management of developing malocclusions using serial extractions can reduce the likelihood of rampant caries and achieve an acceptable occlusion, allowing patients to maintain a healthier dentition.[21] Of note, prophylactic dental care is difficult in patients with dystrophic forms of EB. Ankyloglossia and microstomia seen in these patients makes it challenging for dentists to work in their mouths, which frequently requires general anesthesia.

Gardner syndrome

Gardner syndrome, a subtype of familial adenomatous polyposis, is an autosomal dominant condition characterized by gastrointestinal polyps, multiple osteomas, skin epidermal cysts, mixed muscle and fibrous desmoid tumors, and soft tissue fibromas. Dermatologic manifestations include epidermoid cysts and other benign tumors. Dental anomalies are seen in approximately 20% of patients and include multiple nonerupted teeth, polydontia (Fig 5), follicular odontomas, dentigerous cysts, and caries.[23,24]

Hypomelanosis of Ito

First described in 1951, HI is a neurocutaneous disorder characterized by linear, patchy, or swirling

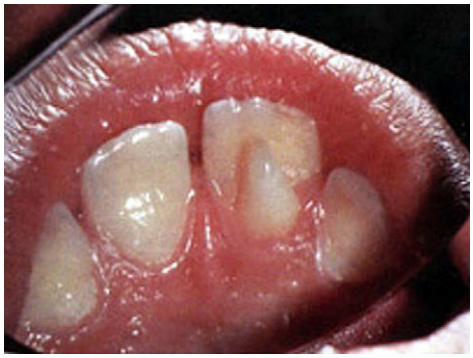

Fig 6. Hypomelanosis of Ito; talon cusp on central incisor. Courtesy of Marquette University School of Dentistry.

areas of hypopigmentation on the skin.[25] These patches are often not present at birth but develop in the first few years. "Pigmentary disorder following Blaschko lines" is a term sometimes used to describe this condition. Ophthalmologic, musculoskeletal, neurologic, and dental anomalies may be associated with the syndrome. The dental abnormalities found in HI include talon cusps,[26] a single maxillary central incisor,[27] enamel defects, hypodontia, and irregularly spaced teeth.[28] The talon cusps (Fig 6) have been described as protuberances appearing on the palatal surface of the incisor crowns, and have been suggested as specific markers for HI by Happle and Vakilzadeh.[26]

Incontinentia pigmenti (Bloch-Sulzberger syndrome)

IP is a rare X-linked dominant genodermatosis that affects mostly female patients and is usually lethal in male fetuses in utero.[29] IP is characterized by abnormalities of the tissues and organs derived from the ectoderm and neuroectoderm. Clinical presentations vary considerably, with neurologic, ophthalmologic, cutaneous, and dental findings usually present. Cutaneous features are divided into 3 distinct phases: an early bullous phase that occurs in a linear distribution on the limbs, a verrucous stage again mainly seen on the limbs, and a pigmented phase that is usually on the trunk and is described as "Chinese lettering" because of the splashes of pigmentation. In adults a fourth stage is described where women have atrophic, linear, white lines on the legs.

Although many of the physical manifestations of IP are variable, dental features occur in more than 80% of cases.[30] These are of paramount diagnostic importance, as, in contrast to the dermatologic features, they persist through life.[31] Typical dental manifestations include hypodontia, delayed eruption, impaction, malformation of the crowns or peg-shaped teeth, and accessory cusps.[32] There are reports of the absence of teeth, especially the lateral incisors and premolars, in otherwise unaffected siblings and mothers of patients with the syndrome.[33] Unlike many other dermatologic diseases with dental features, IP is not associated with increased incidence of enamel hypoplasia.[31]

In a recent review of 40 cases of IP, information about examination of the teeth was available for 17 of the 40 patients. Dental abnormalities were present in 10 (59%): 7 had partial anodontia of deciduous or permanent teeth and 5 had conical teeth. Microdontia and delayed eruption of permanent teeth were noted in one patient.[34] Dental supervision and management is aimed at cosmetic correction of the teeth changes.

Langerhans cell histiocytosis (histiocytosis X)

LCH is a rare proliferative disorder, the hallmark of which is infiltration of various body organs by clonal Langerhans cells. Skin manifestations are protean; presentation with papules, vesicles, nodules, and a seborrheic-like pattern on the scalp and diaper area are all seen in LCH. Ulceration and crusting with studded petechiae are typical of the disease. Dental problems may be seen in approximately 30% of patients with LCH[35]; these can be the presenting features of the disease.[36,37] The most common dental manifestation of LCH is a destructive periodontitis, which results from osseous infiltration by proliferative cells; this can lead to almost total destruction of mandibular and maxillary periodontal support and loosened teeth known as "floating teeth." When periodontal tissue is involved, clinical manifestations can vary from gingival recession and pocket formation to severe alveolar bone loss. Although bacteria are seen in patients with LCH their role in producing periodontitis is not understood. Oral and palatal ulcerations also occur.[38,39] Knowledge of the oral lesions associated with LCH and biopsy of the periodontium can be helpful in establishing the diagnosis.

Naegeli-Franceschetti-Jadassohn syndrome

Naegeli-Franceschetti-Jadassohn syndrome is an autosomal-dominant ED condition, characterized by reticulated hyperpigmentation, hypohidrosis, palmoplantar hyperkeratosis, abnormal teeth, and nail dysplasia. Dental abnormalities constitute one of the cardinal features of the syndrome, and include abnormally shaped teeth, polydontia, yellow spotted enamel, caries, and early total loss.[40]

Nevoid basal cell carcinoma syndrome (Gorlin syndrome)

NBCCS is an autosomal-dominant disorder characterized by a predisposition to develop basal cell

epitheliomas, medulloblastomas, and multiple developmental defects. Multiple odontogenic keratocysts are a hallmark of the syndrome and occur in upward of 80% of children; they were originally described by Gorlin and Goltz.[41] These usually occur during the second or third decades of life; however, one case report documented a 5-year-old with odontogenic keratocysts, which were the initial presentation of the syndrome. Dental examination may help establish early diagnosis as the cysts most often appear in the ramus of the lower jaw.[42] There are histologic differences between odontogenic keratocysts occurring in NBCCS and as single lesions in otherwise healthy persons.[43] Delayed tooth development is sometimes seen in NBCCS. The surgical treatment of odontogenic keratocysts is complicated and there is a considerable risk of recurrence.[44,45] A conservative approach is recommended by some surgeons in an effort to conserve permanent teeth.

Papillon-Lefèvre syndrome

PLS is a rare autosomal-recessive condition associated with cathepsin gene mutations.[46] Diagnostic features of PLS include palmoplantar hyperkeratosis and rapid periodontal destruction. Although the development of deciduous teeth proceeds normally, their eruption is associated with gingival inflammation and subsequent rapid destruction of the periodontium.[47] Severe periodontitis manifests at 3 to 4 years and results from the gingival inflammation, followed by the early exfoliation of the primary dentition.[48] After exfoliation, the inflammation subsides and the gingivae appear healthy.[47] Several reports document successful treatment with aromatic retinoids.[48] With the eruption of the permanent dentition, the process of gingivitis and periodontitis is usually repeated and there is subsequent premature loss of the permanent teeth, although the third molars are sometimes spared.[49]

A recently proposed treatment involves extraction of the primary dentition while on antibiotic therapy; this has been shown to be effective in maintaining the permanent dentition.[50] Early appreciation of associated dental pathology in patients with PLS is paramount because treatment can begin before eruption of the primary dentition. Any young child who exhibits palmoplantar hyperkeratosis should be carefully examined for periodontal breakdown.[51]

Primary immunodeficiency syndromes

Hyperimmunoglobulin E syndrome (Job syndrome) is a rare immunodeficiency disorder characterized by recurrent skin abscesses, an atopic dermatitis-like picture on the skin, pneumonia with pneumatocele development, various skeletal abnormalities, and high serum IgE levels. Dental abnormalities include retention of primary teeth, lack of eruption of secondary teeth, and delayed resorption of the roots of primary teeth. Another rare immunodeficiency disorder, leukocyte adhesion deficiency syndrome, is associated with repeated skin infections, hypomineralization of bones, and hypoplasia of cementum adjacent to the dentogingival junction.[52] Generalized progressive periodontitis and oral infections are common with subsequent loss of primary and permanent dentition.[53]

Sjögren-Larsson syndrome

First described by Sjögren in 1956[54] and by Sjögren and Larsson in 1957,[55] SLS is an autosomal-recessive neurocutaneous genodermatosis characterized by a triad of congenital ichthyosis, spastic diplegia or quadriplegia, and mental retardation. A mutation in the fatty aldehyde dehydrogenase gene on chromosome 17p leads to impaired fatty alcohol oxidation.[56] Ophthalmologic findings of glistening white dots in the fundus, speech defects, epilepsy, dental problems, and skeletal abnormalities are also observed in patients with SLS.

Oral pathology is not specific or helpful in making the diagnosis. Caries, periodontitis, malocclusion, and enamel hypoplasia have been reported in SLS.[57] Early recognition of SLS and treatment of the caries may help prevent further dental sequelae.

Tuberous sclerosis (Bourneville disease)

TS is an autosomal-dominant condition of phenotypic variability. This disorder of cellular differentiation and proliferation results in hamartomatous formation in numerous organs (skin, brain, eye, kidney, heart, lung, and bones). Mental retardation and seizures are commonly associated with the dermatologic findings in TS. These include hypomelanotic macules, facial angiofibromas, shagreen patches (connective tissue nevi), Koenen tumors (fibromas) surrounding the nail, and hard fibromas around the hair line. Consensus diagnostic criteria of TS were recently revised.[58]

Characteristic oral features of TS include gingival fibromas and dental enamel pits. The latter have been documented in 71% of patients with TS by Lygidakis and Lindenbaum,[59] and in 90% of patients reported by the TS Complex Consensus Group. Flanagan et al[60] found that most patients with TS had greater than 14 pits per person, whereas healthy control subjects have less than 6. Multiple randomly distributed pits in dental enamel have been

Table I. Dental manifestations of dermatologic diseases

Disease	Dental features
Congenital erythropoietic porphyria	- Red-colored teeth
Congenital syphilis	- Widely spaced upper incisors
	- Centrally notched upper incisors (Hutchinson incisors)
	- Microdontia of molars (mulberry molars)
	- Enamel hypoplasia of molars and incisors
Ectodermal dysplasias	- Hypodontia
	- Microdontia
	- Conical rows
	- Enamel defects and caries
	- Anodontia
	- Polydontia
	- Retained primary teeth
	- Peg-shaped teeth
	- Accessory cusps
Epidermolysis bullosa	- Enamel hypoplasia
	- Dental caries
Gardner syndrome	- Multiple nonerupted teeth
	- Polydontia
	- Odontomas
	- Caries
Hyperimmunoglobulin E syndrome	- Retention of primary teeth
	- Ankylosed permanent teeth
Hypomelanosis of Ito	- Hypodontia
	- Irregularly spaced teeth
	- Single maxillary incisor
	- Talon cusps
	- Enamel defects and caries
Incontinentia pigmenti	- Hypodontia
	- Delayed eruption
	- Impaction
	- Peg-shaped teeth
	- Accessory cusps
Langerhans cell histiocytosis	- Destructive periodontitis
	- Loosened teeth
Leukocyte adhesion deficiency syndrome	- Hypomineralization and hypoplasia of cementum
	- Generalized progressive periodontitis
Naegeli-Franceschetti-Jadassohn syndrome	- Abnormally shaped teeth
	- Polydontia
	- Yellow spotted enamel
	- Early total loss of teeth
Nevoid basal cell carcinoma syndrome	- Odontogenic keratocysts
Papillon-Lefèvre syndrome	- Gingival inflammation
	- Periodontal destruction
	- Premature exfoliation
Sjögren-Larsson syndrome	- Gingivitis
	- Malocclusion
	- Enamel hypoplasia
Tuberous sclerosis	- Large enamel pits on labial surface and away from gingivae
	- Decreased enamel matrix

designated as a minor feature of the diagnostic criteria.[58] The enamel pits are thought to result from a reduction in the amount of enamel matrix formed during amelogenesis, and not from decreased mineralization or defective maturation of the enamel matrix.[59] The dental enamel pits found in patients with TS differ from the pits found in unaffected individuals.[59] TS lesions have multiple large enamel defects farther away from the gingivae and on the labial surface of the teeth. There is usually no

alteration in color or texture of the enamel surrounding the pit, but they do acquire a dark stain from the oral environment resulting in a black, pockmarked appearance of the defects. These can be visualized as multiple pin-sized radiolucencies on radiographs.[59] Dentists can fill the enamel pits for aesthetic reasons.

Conclusion

Dermatologic diseases and syndromes are often diagnosed through the identification of systemic signs and symptoms. Dental pathology is seen in a range of dermatologic disorders, but is often overlooked. A number of skin diseases and syndromes, relevant to dermatologists, present with dental signs and periodontal pathology, as summarized in Table I. An appreciation of the specific dental signs associated with cutaneous diseases can aid in the diagnosis and assist in the treatment of patients with these conditions.

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