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DERMATOLOGICAL DISORDERS WITH ORAL MANIFESTATIONS IN PAEDIATRIC PATIENTS

Dental Science

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ABSTRACT

Dermatological diseases, including the skin and its supplements also involve the oral cavity, which needs special attention, they may be the early presenting clinical feature or the only sign of these disorders. The oral mucosal lesions in dermatological disorder can be life-threatening and also affect the quality of life in terms of pain, discomfort, social, and functional limitations. The rarity of the conditions and lack of awareness are the major complications in the management of these disease. Various dermatological disorders of diverse etiologies are associated with oral lesions, thus improving the knowledge about them will strengthen and enhance interdisciplinary and multisectoral approach and lead to better management of such patients. This review elaborates about various dermatological lesions in paediatric patients with oral lesions.

KEYWORDS

INTRODUCTION:

Dermatological diseases, including the skin and its supplements also involve the oral cavity, which needs special attention, they may be the early presenting clinical feature or the only sign of these disorders. The oral mucosal lesions in dermatological disorder can be life-threatening and also affect the quality of life in terms of pain, discomfort, social, and functional limitations.The rarity of the conditions and lack of awareness are the major complications in the management of these disease.[1] Various dermatological disorders of diverse etiologies are associated with oral lesions, thus improving the knowledge about them will strengthen and enhance interdisciplinary and multisectoral approach and lead to better management of such patients.[2]This review elaborates about various dermatological lesions in paediatric patients with oral lesions.

Vesiculobullous lesions

Pemphigus vulgaris:

Pemphigus vulgaris (PV) is an autoimmune disease in which the autoantibody, immunoglobulin G, is directed against the keratinocytes in the epidermis. The classic presentations of PV are flaccid vesicles or bullae over the oral mucosa, trunk, groin, and extremities. It is characterised by blister formation that occurs within the epidermis caused by acantholysis, that is, the loss of cohesion between epidermal cells. PV often starts in the oropharynx and then spreads to areas that involve the trunk, head, and intertriginous regions. Stomatitis is the presenting symptom in more than 50% of cases. Epistaxis and hoarseness are also seen owing to involvement of the nose, pharynx, and larynx.Most patients present with painful oral erosions, and for some, this is the only clinical manifestation. Months elapse before skin lesions occur. The diagnosis of PV is often delayed when oral lesions are the only manifestation, and the index of suspicion may be even lower in a young patient because of the infrequent incidence of PV in children. PNP has been associated with non-Hodgkin lymphomas, thymomas, Castle man's disease, and chronic lymphocytic lymphoma.[3]

Mucous membrane pemphigoid:

Mucous membrane pemphigoid is chronic inflammatory, sub epithelial blistering diseases predominantly affecting mucous membranes oral, ocular, urogenital, anal, nasopharyngeal, oesophageal and laryngeal with or without clinically observable scarring. It is characterised by linear deposition of IgG and C3, and IgA to a lesser degree, along the epithelial basement membrane zone (BMZ).

The oral cavity is most frequently affected, and in many cases constitutes the single presentation of the disease. Oral MMP typically manifests as erythematous patches, blisters, erosions and pseudomembrane-covered erosions, located mostly on attached gingiva, where it is described as desquamative gingivitis (DG). Other oral sites, such as palate, lips, tongue and buccal mucosa, can also be involved, but with a lower incidence. Prognosis of oral childhood MMP is good, although long-term follow-up is recommended. Early diagnosis of MMP may avoid severe and irreversible involvement of other mucous membrane sites.[4]

Paraneoplastic pemphigus :

Paraneoplastic pemphigus encompasses a multitude of mucocutaneous and systemic findings. Mucocutaneous involvement in paraneoplastic pemphigus is prominent, with painful erosive lesions involving the oral, nasal, upper gastrointestinal, respiratory, ocular, and genital epithelium. Histopathologic examinations classically reveal acantholysis, intraepidermal blister formation, and immunoreactant deposition along the basement membrane and within epithelial intercellular spaces. The occurrence of PAMS in childhood has been reported infrequently. When PAMS occurs among children, however, Castle man's disease is the most common underlying neoplasm, with progressive bronchiolitis obliterans resulting in pulmonary destruction and death. Patients have hemorrhagic crusting of the lips, severe ulcerative stomatitis/gingivitis, desquamative esophagitis and tracheobronchitis, scarring conjunctivitis, and genital erosions. A recent study showed that autoantibodies are found in kidney, bladder, smooth muscle, and striated muscle, in addition to skin, upper digestive tract, and respiratory tract epithelium.[5]

Epidermolysis bullosa :

Epidermolysis bullosa describes a heterogeneous group of inherited blistering mucocutaneous disorders which have a specific defect in the attachment mechanisms of the epithelial cells, either to each other or to the underlying connective tissue. This dermatological condition is a severe autoimmune disease. It is characterised by blistering of the skin and mucosa in response to trauma. EB patients require special precautions during dental treatment because of the high risk of lesioning the soft tissue when handling cutting instruments close to the skin and oral mucosa.

The oral manifestation includes oral mucosal erosions, reduced vestibular sulcus depth and also have dental anomalies of shape, position, and structure (hypoplasia and hypomineralization); tongue denudation and limited mobility, ankyloglossia, and microstomia.[6]

Linear IgA disease:

Linear immunoglobulin A (IgA) dermatosis (LAD) is a rare chronic autoimmune disorder, which presents with vesicles and bullae

formation affecting both the skin and the mucous membrane, with a characteristic immunofluorescence finding of homogeneous linear deposits of IgA in the cutaneous basement membrane zone.It is divided into two types, based on the age of onset, namely Linear IgA bullous dermatosis of childhood and Linear IgA bullous dermatosis of adult.[7] Vesiculobullous rash usually appears in the lower trunk, extensor faces of arms and legs, buttocks and genital zone. Although it is less frequent, lesions may also be observed in face, scalp, feet dorsum or anus zone.Lesions may or not be symmetric respect to the medial line of the body.

Oral Manifestation- Proximately in 80 % of patients mucosal lesions can be observed oral, ocular, nasal or genital mucous. A 60 – 70 % of LAD patients have oral mucosal lesions. The most frequent oral lesions consist in painful erosive or ulcerative lesions caused by the rupture of bullae. These ulcerative or erosive lesions may appear anywhere in the oral mucous, including vestibular mucous, and the tongue. Occasionally, LAD can be manifested in the mouth as a chronic desquamative gingivitis, which cannot be clinically distinguished of the desquamative gingivitis produced by lichen planus, pemphigoid or other mucocutaneous diseases.[8]

Connective tissue disorders

Juvenile Systemic lupus erythematosus:

Juvenile Systemic lupus erythematosus is one of the most common autoimmune diseases in children and has a clinical course ranging from mild, gradual onset to rapid, progressive multi-organ failure. Approximately 20% of systemic lupus erythematosus (SLE) patients are diagnosed in childhood and adolescence, with a median age of 11–12 years. Systemic lupus erythematosus patients usually present with mucocutaneous lesions, renal involvement, central nervous system disorders and haematological abnormalities. The mucocutaneous manifestations in JSLE are classified as lupus erythematosus (LE)-specific and LE-nonspecific skin diseases.

Lupus erythematosus specific- A palatal erythematous ulcer is the typical oral ulcer and it is described as a painless, single or multiple lesions on masticatory or keratinised mucosa, especially the hard palate. It is an acute sign occurring when disease is active, and sometimes is the first clue of SLE without skin lesions . The early lesion may be haemorrhaging before developing into the ulcer. If the ulcers are coalesced into a large erythematous patch at the hard palate and extend to the soft palate, the early onset of oral Discoid lupus erythematosus should be considered.

Lupus erythematosus non specific-

i. **Aphthous ulcer-** The lesion is usually presented with multiple lesions or in a group, and tends to involve primarily lining tissue: the soft palate, buccal and labial mucosa. Aphthous ulcers are more common in JSLE patients. The most common type of aphthous ulcer in JSLE patients is multiple minor ulcers and less than ten lesions.
ii. **Lupus cheilitis-** It is an inflammation of the buccal lips. The typical lesion is small or diffuse, erythematous and oedematous, and may turn into crusty painful ulcers. Cheilitis often involves the vermilion zone of the lower lip (typical lupus cheilitis). As the lesion is found in the sun-exposed area, lupus cheilitis is usually associated with photosensitivity in JSLE patients.[9]

Systemic sclerosis:

Systemic sclerosis is a rare connective tissue disease of unknown etiology characterised by increased collagen deposition leading to fibrosis and subsequent degeneration of the skin and internal organs.

Systemic features include sclerodactyly, Raynaud's phenomenon, telangiectasia, calcinosis, myositis, arthritis, tenosynovitis, renal failure, oesophageal hypomotility, pulmonary fibrosis and heart failure. Oral manifestation include reduced interincisal distance, xerostomia increased periodontal width, resorption of the mandible ,periodontal disease ,and increased decayed ,missing, and filled teeth (DMFT). The prognosis is difficult to predict because spontaneous remission has been documented, but death may result from extensive visceral involvement (heart, kidney, and lung)[10].

Genodermatoses

Goltz syndrome:

The Gorlin-Goltz syndrome, nevoid basal cell carcinoma syndrome, or basal cell nevus syndrome is an autosomal dominant condition disorder with high variability expression. It presents a series of relevant clinical manifestations that suggest its diagnosis in cutaneous, bone, dental, soft tissue, nervous, and ocular system disorders.

The diagnostic criteria for the Gorlin-Goltz syndrome were divided into major and minor.

i. The major includes the presence of more than two basal cell carcinomas or a history of one basal cell carcinoma below the age of 20 years, odontogenic keratocysts of the jaw three or more palmoplantar pits, bifid, fused, or markedly splayed ribs. Basal cell carcinomas are usually diagnosed after puberty. Due to the susceptibility to the development of neoplasm and the syndrome becoming very aggressive as age progresses, the diagnosis should be precocious.
ii. The minor criteria are falx cerebri calcification, macrocephaly, congenital anomalies, cleft lip palate, frontal bossing, coarse face, hypertelorism, skeletal anomalies, impacted or ectopic teeth, bilateral coronoid hyperplasia and oligodontia deformity, pectus deformity, hemivertebra and combined vertebral corpiovarian fibroma, and medulloblastoma.[11]

Hypohidrotic ectoderm dysplasia:

Ectodermal dysplasia (ED) comprises a large group of clinical and genetically heterogeneous diseases affecting the ectoderm-derived structures, such as the hair, nails, sweat glands, and teeth. The most common forms of ED are the X-linked hypohidrotic (Christ-Siemens-Touraine syndrome) and hidrotic (Clouston syndrome) types. The X-linked hypohidrotic presents with hypodontia or anodontia, hypotrichosis, and hypohidrosis or anhidrosis, while the hidrotic type is more severe, involving nail dystrophy, hypotrichosis, and palmoplantar keratoderma.[12]

Ectodermal dysplasia may be defined as conditions with at least one the following features Trichodysplasia, dental defects, onychodysplasia or dishidrosis as well as atleast one sign showing involvement of other ectodermal structure.Patients with HED usually have skin that is soft, thin, dry, either with complete or partial absence of sweat glands. As a result, they can- not perspire normally and may have heat intolerance and hyperplasia. Hair follicles and sebaceous glands are often defective or absent and the hair of the scalp and eyebrows tends to be fine, scanty and blond.[13]

Oral manifestations of HED include partial or complete absence of teeth. This can affect both the primary and permanent dentition. The teeth which are present may be conical in shape with large pulps, sometimes hypoplastic and delayed in eruption. Prosthetic dental treatment for ED is necessary to improve chewing, facial aesthetics, speech, nutrition, and social integration, the last of which can influence the patient's emotional health.[13]

Dyskeratosis congenita

Dyskeratosis congenita is an inherited bone marrow failure syndrome characterised by abnormal skin pigmentation, nail dystrophy, oral premalignant leukoplakia, BMF, and cancer predisposition, with increased risk for squamous cell carcinoma and hematolymphoid neoplasms.[14]

Approximately 90% of patients exhibit nail dystrophy, which affects the finger nails first and then the toenails in most cases. Nail dystrophy begins with ridging and longitudinal splitting and progresses, resulting in small, rudimentary, or absent nails. Mucosal leukoplakia is a pathognomonic feature and occurs in approximately 80% of patients. It typically involves the buccal mucosa, tongue, and oropharynx. The treatment of these lesions is symptomatic. However, approximately 30% of the leukoplakic areas progress to malignant transformation with the development of a squamous cell carcinoma in 10–30 years. Patients may have gingival inflammation, bleeding, recession, and bone loss that simulate juvenile periodontitis. In addition, there may be defects in decreased root/crown ratio and mild taurodontism. The evidence of multiple permanent teeth with decreased root/crown ratios may suggest a diagnosis of DC. These patients also may have more incidence of and more severe periodontal disease.[15]

Steven – Johnson syndrome:

SJS begins with a prodrome of fever, malaise, anorexia, pharyngitis, and headache lasting for 2–3 days, at times, extending to 10–11 days. Mucosal lesions usually precede skin lesions. Usually, two mucosal membranes are involved, most commonly conjunctiva and oral

mucosa Oral lesions occur in crops. Thickly walled vesicles rupture into irregularly shaped ulcers covered by pseudomembranes are seen in lips, buccal mucosa, palate, the anterior, and lateral border of the tongue. Gums are spared in contrast to herpetic gingivostomatitis. Oral mucosal involvement is seen in 90% of SJS.[16]

Skin lesions are accompanied by pain, which is a characteristic feature of SJS and TEN. Flat atypical target lesions or erythematous macule of irregular shape and size with are seen. Skin lesions are seen initially on the face, neck, chest, trunk, and proximal extremities. This is followed by confluence and development of flaccid blisters. SJS should be differentiated from erythema multiforme major which is recognised by the presence of acrally distributed typical target lesions, and are not associated with significant skin detachment. Drug-specific CD8+ cytotoxic T-cells release perforins and granzyme on direct contact with keratinocytes expressing specific MHC alleles is also a possible mechanism for the development of drug-induced SJS.[17]

Behect syndrome :

Behçet's Disease (BD) is a rare systemic vasculitis disorder of unknown etiology characterised by recurrent attacks of oral aphthous ulcers, genital sores, and ocular lesions (triple-symptom complex). Clinical manifestations of acute inflammation are typically self-limiting in time with relapsing episodes of varied intensity leading to sequelae. Earlier age onset is correlated with more severe clinical manifestations and mortality. BD may affect almost all vascularised systems and is characterised by episodes of relapses and remissions with sequelae. Although several clinical manifestations are associated with BD, the triple-system complex of oral and genital aphthae, and uveitis. Children exhibit more frequent perianal aphthosis and arthralgia, less frequent genital ulcers and vascular involvement and a more severe course of uveitis.[18]

Skin disease occurs in about 80% of patients with Bechet disease, and lesions often occur in combination. Erythema nodosum is common, particularly in females. It usually affects the lower limbs and may resolve leaving hyper pigmented areas. Superficial thrombophlebitis is also common and may be confused with erythema nodosum. Papulopustular lesions and acneiform nodules also occur. These are morphologically identical to adolescent acne, although their distribution may be more widespread, affecting the arms as well as the face, torso and buttocks.[19]

Moderate anaemia of chronic disease is common, and a neutrophil leukocytosis is seen in 15% of patients. Serum immunoglobulins may be non-specifically elevated. Autoantibodies such as rheumatoid factor, anti-nuclear antibody and anti-neutrophil cytoplasmic antibody are usually negative. Importantly, non-specific markers of inflammation such as C-reactive protein level and erythrocyte sedimentation rate can be normal.[19]

Incontinentia pigmenti:

Incontinentia pigmenti is a rare genodermatosis in which the skin involvement occurs in all patients. Additionally, other ectodermal tissues may be affected, such as the central nervous system, eyes, hair, nails and teeth. The disease has a X-linked dominant inheritance pattern and is usually lethal to male fetus . The dermatological findings occur in four successive phases, First phase - vesicles on an erythematous base, second phase - verrucous hyperkeratotic lesions; third phase - hyperchromic spots and fourth phase - hypo chromic atrophic lesions.

Cutaneous findings are characterised by four stages

Stage 1- known as inflammatory or vesicular stage, is characterised by the development of papules, vesicles and pustules on an erythematous base, distributed linearly along the lines of Blaschko.

Stage 2- also known as verrucous stage, is characterised by plaques and warty papules linearly arranged over an erythematous base, also following the lines of Blaschko.

Stage 3 - hyper pigmented stage is defined by the development of linear or whorled lesions, with a brownish pigmentation, which may be accompanied by atrophy. This stage occurs in 90-98% of patients with IP.

Stage 4 - known as atrophic or hypopigmented, is characterised by areas of hypopigmentation, atrophy and absence of hair, most frequently observed on the lower extremities.[20]

Oral manifestations are the most frequent, which are observed in 80% of patients and usually affect both dentitions. The most frequent alteration is hypodontia (up to 43% of patients), followed by pegged or conically crowned teeth (30% of patients).[21]

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