CASE REPORT

Dental Management in a 7-year-old Child with Ichthyosis Vulgaris: A Rare Case Report

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Abstract

Ichthyosis Vulgaris is a rare cornification disorder affecting the skin characterized by the accumulation of hyperkeratotic fish-like scales on the skin surface. It is a minor form of ichthyoses disorder. There is little evidence related to oral and dental manifestations in these patients. As their general manifestations can lead to oral and dental manifestations also, it is important to take various precautions by the dentists while managing these children. Also, preventive therapies can be initiated in them in the early stages to limit the acquired dental manifestations.

This case reports various dental considerations and management carried out in a 7-year-old female patient who was a known case of Ichthyosis Vulgaris.

Keywords: Dental caries, Dry lips, Filaggrin, Fishy-scales, Ichthyosis Vulgaris, Rhinitis.

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Introduction

Ichthyosis is a group of inherited conditions constituting, both clinically and etiologically, a wide heterogeneous group of cornification disorders that affects the skin and are clinically characterized by accumulation of hyperkeratosis or scales.[1] The term ichthyosis has been derived from the Greek word "ikhthus" which means "fish" and indicates the similarity of skin appearance fish scales. The worldwide first Ichthyosis Consensus Classification was approved in 2009 which was based on the recent pathogenic aspects and clinical presentation. In India, the incidence of ichthyosis is approximately between 1:300000.[2] It is caused due to loss-of-function mutations in the filaggrin gene (FLG) leading to abnormal keratinization and exfoliation of the horny cell layer. This causes scaling and thickening of the cornified layer and is commonly associated with inflammation of the skin which presents as erythroderma. The management aims at the removal of the external scales and maintaining soft and pliable skin. This is done using topical treatments like urea or lactic-acid based emollients, and/or keratolytics. Oral retinoids may be preferred in severe cases.[3]

Case Description

A 7-year-old girl reported to the outpatient Department of Pedodontics with the chief complaint of pain in the left upper back tooth region for 5 days. Her parents are a nonconsanguineous couple and she was born with normal delivery. Her mother reported that the child developed generalized scaling all over the body within a year after birth and was diagnosed with Ichthyosis Vulgaris by a dermatologist. No positive family history was elicited. The child did not have any ocular, otolaryngeal, or any other systemic abnormalities and had normal IQ and physical growth. The treatment advised to her by the dermatologist was specific soaps and moisturizers. Also, the child has frequent episodes of rhinitis. Dental history revealed restorations of carious teeth before 2 years. The child did not have any history related to oral habits, although intermittent episodes of breathing through the mouth during rhinitis were reported.

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Cutaneous examination revealed dry fishy scales involving the entire body surface including the upper and lower limbs, face, scalp, and body trunk. The hair was sparse and had scanty eyebrows. There were no abnormalities involving palm, sole, or nails (Figs 1A – C).

Extraoral examination elicited normal mouth opening, absence of a nasal bridge, dry lips with bilateral multiple fissures at the corner of the mouth. Malar deficiency was noticed in the lateral profile (Figs 1D and E).

Intraoral examination showed mixed dentition with dental caries in 55, 74, 36; restored 64; fractured restoration in 84; preshredding mobility of 51 and mild gingivitis (Figs 2A – C). No pathology was noticed in the tongue.

Radiological examination depicted mixed dentition with multiple carious lesions and no other defects (Fig. 3A).

The dental management was initiated with preventive measures such as oral prophylaxis and fluoride application as the patient had high caries risk followed by restorations in relation to 55, 64, 36, 84, Mineral Trioxide Aggregate pulpotomy in 74 (Fig. 3B), followed by postoperative fluoride application. 51 had undergone natural exfoliation. Crown w.r.t 84 was avoided as parents were not willing for esthetic crowns due to economic factors and the child's physician advised to prevent stainless steel crown, if possible. Oral hygiene instructions, maintenance, and follow-up protocols were informed to the parents and the child. Adequate moisturizing of lips using petroleum jelly was advised. Also, the possible correlation

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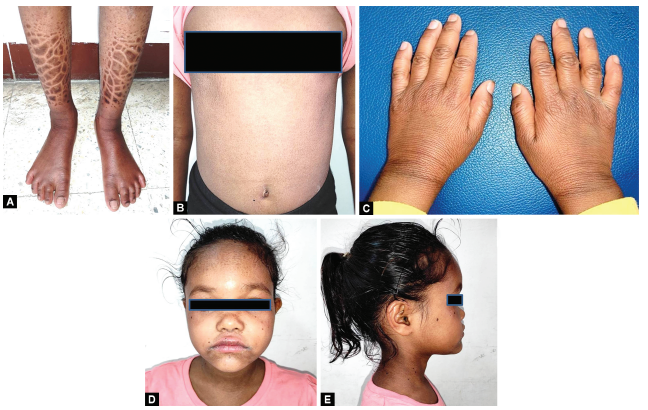
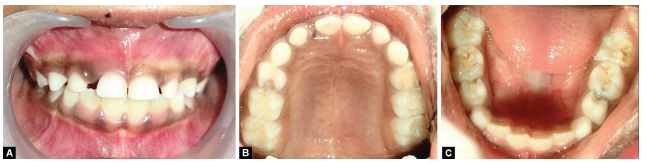


Figs 1A to E: General manifestations (A)Thick, armor-like brownish polygonal scales on the legs. (B) Scales on the anterior trunk. (C) Scales on the hand region. (D) Extraoral frontal profile. (E) Extraoral lateral profile



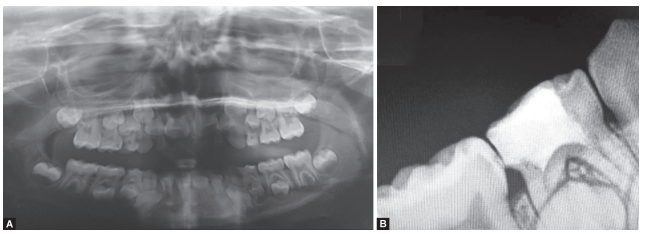
Figs 2A to C: Preoperative intraoral images (A) Dentition in occlusion. (B) Multiple carious maxillary dentition. (C) Multiple carious mandibular dentition

between rhinitis leading to mouth breathing and increased caries incidence in the child was informed to the parent.

Discussion

Ichthyosis Vulgaris is an autosomal semi-dominant disorder. It is the most commonly inherited keratinization disorder which is mainly caused due to functional loss mutations in the FLG and clinically presents during the first years of life,[4] and the reported patient also manifested symptoms within 1 year of birth. FLG mutations are prevalent in approximately 7.7% of Europeans and 3.0% of Asians, but infrequent in darker-skinned populations. Its overall prevalence is 1:250–1000.[5] The xerosis manifests itself as fine (powdery) or coarse (polygonal) scaling of the extensor surfaces of the extremities, the scalp, the central part of the face, and the trunk. The extensor surfaces of the lower limbs are more commonly affected in adults than in children.[6] The scales are centrally adherent and have loose edges, which appear smaller in children when compared to adults. The scales involving the scalp may simulate dandruff but the predisposition to fungal infections is not known.[7] A similar presentation of scales involving various parts of the body and scalp resembling affected with dandruff was noticed in our patient.

The diagnosis is based on clinical manifestations, histopathological examination, and genetic testing. Genetic testing can be utilized to provide genetic counseling to the parents. Also, light microscopy, immunofluorescence, and electron microscopy (EM) can be used. EM can be utilized to differentiate between heterozygous and homozygous carriers.[8] It can be differentiated from Cleidocranial dysplasia, Neurodevelopmental delay and dysmorphic facial features, Achondroplasia, Conradi-Hünermann-Happle syndrome, Cornelia de Lange syndrome, Osteogenesis Imperfecta, and



Figs 3A and B: X-rays (A) Preoperative orthopantomogram. (B) MTA pulpotomy

Klippel-Feil syndrome by the clinical features of skin, genetic and histopathological examination.[9]

The treatment objective of ichthyosis is the removal of the scaliness externally and maintaining a soft and pliable skin condition. Hence, moisturizers are the preferred mainstay management. Keratolytics and humectants have proven to be efficient but cause irritation and increased plasma urea levels.[10] Oral retinoids are known to be the most effective therapy for most of the ichthyoses but may have side effects like angular cheilitis. The dentist should advise the patient to apply petroleum jelly or local anesthetic gel in the affected areas, as advised in the reported case.

Various studies have reported increased penetration of allergens and chemicals in filaggrin-deficient skin, increased levels of hand eczema, allergic rhinitis, irritant contact dermatitis, nickel sensitization, and serum vitamin D levels.[8] Similarly, the child mentioned in this case has a history of frequent common cold/rhinitis. Also, it is necessary to inform the patient about the increased risk of developing dermatitis when exposed to nickel or irritants. Dental consideration should be given while choosing crowns or other restorations involving metals known to cause allergy.

There is a very limited amount of knowledge about the association between Ichthyosis Vulgaris and dental manifestations. The various dental and oral findings mentioned in Ichthyosis Vulgaris patients are gingivitis, periodontitis, enamel hypoplasia, increased risk and incidence of caries, delayed primary and permanent tooth eruption, bruxism, bifid teeth, varied morphology of teeth, fish mouth appearance, mouth breathing, dry mouth, and hyperkeratotic plaques on the tongue, fissures around the corner of the mouth.[11] Literature has few cases reported by various authors to find a correlation between ichthyosis and its dental manifestations. List et al. reported abnormal deciduous and permanent teeth with the presence of notches and pits in three individuals diagnosed with ichthyosis.[12] Bolgul et al. mentioned missing teeth, dental caries, and retained deciduous teeth in a 14-year-old boy.[13] Similarly, in the reported case, the presence of multiple caries and dry lips with fissures around the corner of the mouth, perioral dryness with the absence of a nasal bridge, and malar prominence were noticed. The eruption of permanent teeth does not seem to be affected.

Since these children have a possibility of hepatic toxicity because of the use of retinoids, the pedodontist must be aware of this and make an informed choice of local anesthetic agents and medicines in them.[14] The possible cause of increased dental caries in the reported child can be frequent rhinitis leading to mouth breathing and dryness of the mouth which was informed to the parents. Regular follow-up with required preventive measures should be carried out for early diagnosis of any oral or dental manifestations and their management. Manipulation

of the patient's skin or oral and perioral tissues should be avoided during dental treatment as these areas can be tender or friable. Dentists can moisturize the oral and perioral tissues with petroleum jelly before commencing the treatment to avoid discomfort to the child.

Conclusion

Ichthyosis Vulgaris, although is a dermatological disease, can affect the oral and perioral tissues. There is little knowledge about the association between the disease and dental manifestations but involving a dentist in early diagnosis and treatment planning with preventive measures will help in alleviating oral and maxillofacial disorders, as the reported cases have shown some extent of oral and maxillofacial involvement.

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