A Case of Ichthyosis Vulgaris and Its Dental Manifestations

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Abstract

Ichthyosis vulgaris is a cornification disorder that is characterized by accumulation of hyperkeratotic fish like scales on the skin surface. It is clinically characterized by scaling, especially on the flexor limbs, with palm plantar hyper linearity. This paper illustrates dermatological as well as dental manifestations of a 14 year old boy affected with ichthyosis vulgaris which makes this case very much unique.

Keywords: Dental management; Dermatological disorder; Ichthyosis vulgaris

Introduction

The ichthyosis forms clinically and etiologically a large heterogeneous group of cornification disorders that are characterized by accumulation of hyperkeratotic scales on the skin surface [1]. Ichthyosis is derived from the Greek word “ikhthus” meaning “fish” and refers to the similarity in appearance of skin to fish scales. Ichthyosis is caused by abnormality in keratinization and exfoliation of the horny cell layer [2]. The relevant clinical feature of this disorder is scaling and thickening of cornified layer which is often accompanied by inflammation of the skin presenting itself as erythroderma [3]. The incidence of ichthyosis is approximately between 1:30000 in India [3]. This case report presents dental involvement in a rare case of ichthyosis vulgaris.

Case Report

A 14-year-old boy reported to the Department of Pedodontics, Bharati Vidyapeeth Dental College and Hospital, Sangli with the chief complaint of pain in the lower left back region of the jaw since 5 days. He was born to a non-consanguineous couple as a collodion baby. He had generalized scaling all over the body since his early infancy and was diagnosed as a case of ichthyosis vulgaris by dermatologist after a skin biopsy and was treated with different moisturizers. History of such disease was not reported in the family. Patient did not suffer from any ocular and otolaryngeal abnormalities. Systemic abnormalities were not detected. The patient's IQ and physical growth was normal. Cutaneous examination reveals dry scales covering the entire body surface including the face (Figure 1), the body trunk (Figure 2), the shoulder (Figure 3) the upper and lower extremities of the body (Figures 4 and 5). Patient’s hair was dry, sparse and eyebrows were scanty (Figure 1). There were no nail abnormalities. The mouth opening of the patient was normal with multiple fissures on corner of the mouth bilaterally.

Figure 1: Extra-oral pre-operative image of the child.

Figure 2: Scales over the chest region.
Intraoral examination revealed multiple carious lesions (Figures 6-8). Root piece with 54, 55, 64 and 65 (Figure 7); over-retained 75 and 85 (Figure 8), grossly decayed 36, lingually erupting 35 were seen. No soft tissue abnormalities were present. Treatment plan was explained to the child's parents and a written consent was obtained from them. The dental management was initiated with a preventive approach as the oral hygiene was compromised and patient was under high caries risk. Full mouth scaling and polishing was done. Pit and fissure sealants on 17, 27, 37 and 47 were applied. Composite restoration with 16, 26 and 46 was done. Root canal therapy with 36 followed by preformed pre-contoured stainless steel crown placement was done. Extraction of 54, 55, 64, 65 root pieces and extraction of over-retained 75 and 85 was done under local anaesthesia with adrenaline. Radiographic investigations (Figure 9) showed mixed dentition without any enamel defects; however, multiple carious lesions were seen on radiograph, delayed eruption pattern of the teeth was also seen. Adequate oral hygiene instructions and demonstration of brushing technique was performed for the patient. Oral hygiene maintenance protocol was informed to the parents and they were also instructed to be on a regular follow-up. For the management of his skin problem, he was referred to dermatologist.
The follow-up after 2 months revealed good oral hygiene (Figures 10 and 11), correction of lingually placed 35 (Figure 11) and eruption of teeth (Figure 12).

Discussion

Ichthyosis Vulgaris is a common genetic keratinization disorder, clinically characterized by scaling, especially on the flexor limbs, and with palm plantar hyper linearity. Till date, six major distinct clinical subtypes of this disorder are known. The most severe form being Harlequin ichthyosis (HI,MIM/242500) followed by Lamellar ichthyosis (LI,MIM/242300); Congenital ichthosiform erythroderma (CIE,MIM/242100); Epidermolytic ichthyosis (EI,MIM/113800); recessive X linked ichthyosis (RXLI,MIM/308100); to the mildest form of Ichthyosis Vulgaris [4]. The epidermis of patient suffering from Ichthyosis vulgaris shows a reduction in their size and numbers or, complete absence of keratohyalin granules [5].The degradation products of keratohyalin granules occupy the cytoplasm of keratinized cells in the stratum corneum and play important roles in the skin barrier function. In the granular layer of the epidermis the keratohyalin granules are predominantly composed of large (>400-kDa) profilaggrin polyproteins [6,7]. After the terminal differentiation of keratinocytes, profilaggrin is cleaved into 10-12 essentially identical 37-kDa filaggrin peptides. The liberated filaggrin aggregates the keratin filaments, resulting in collapse of the granular cells into a flattened square-shape [6]. In addition, the degradation products of filaggrin contribute to moisture retention in the cornified layers. Thus, filaggrin, a major component of keratohyalin granules, is indispensable to the normal, intact, skin barrier function. In this context, a loss or reduction in filaggrin expression results in excessively dry skin and impaired barrier function, which leads to clinical features of Ichthyosis Vulgaris [4]. Similarly in the case which was reported, the patient had findings which included; scales over the upper and lower extremities, the trunk region, the shoulder region and the face. Also, there was thickening and exaggerated lines on the palm and the feet.

There is a very little knowledge about the dental manifestations and its management of a patient with Ichthyosis Vulgaris. There are reports in literature of patients with high risk caries, gingivitis,
periodontitis, and enamel hypoplasia, and delayed eruption of the teeth, irregular morphology of the teeth and presence of plaque on the surface of the tongue in Ichthyosis [8]. Few authors have correlated Ichthyosis and its dental manifestations; List et al. noticed abnormal deciduous and permanent teeth or notched and pitted teeth in three individuals with ichthyosis [9], Bolgul et al. reported missing teeth, carious teeth and persistent deciduous teeth in a 14-year-old boy [10]. Over-retained teeth and delayed eruption of permanent teeth could hamper the normal growth and development of the jaw leading to malocclusion. Similarly in our case, we reported multiple carious teeth, over retained deciduous teeth, delayed eruption of permanent teeth and lingually erupting premolar tooth.

The dental treatment was initiated with a preventive approach, required extractions were carried out to get prevent malocclusion and enhance proper growth and development of the maxilla and mandible.

"The treatment of ichthyosis is essentially removal of the scaliness externally and the maintenance of a soft and pliable condition of the skin" [11]. Moisturizers are the mainstay of therapy of the ichthyoses. They work to increase the flexibility of the epidermis, hydrating and restoring epidermal barrier function, and by covering and removing scale [12]. One approach is to offer general dry skin care along with a list of moisturizers and suggest that affected individuals compare several until the “best” product for that person is identified. Keratolytics and humectants are said to demonstrate efficacy. However, irritation is common, and high plasma urea levels have been reported. The retinoids and calcipotriene have been used as topical therapy for ichthyosis in a small number of trials and have proved to be beneficial [13]. Systemic therapy with oral retinoids is the most effective therapy available for most of the ichthyoses. The retinoids have significant guaranteed and potential side effects such as Angular chelitis [11]. The dentist should treat this by advising application of petroleum jelly or local anaesthetic gel to the affected areas. Most of the times, patients with ichthyosis do not require any modification in dental treatment; however, the dentist should be aware of the concurrent medical problem and its treatment, as there is possibility of hepatic toxicity with the use of retinoids, which can affect the choice of local anaesthetic agents during dental treatment. Regular follow-up should be carried out and the erupting permanent teeth should be taken into consideration to prevent any type of malocclusion. During dental treatment care must be taken to avoid manipulating the patient's skin, particularly in the perioral areas, since affected areas can be tender or friable [14].

Conclusion

Ichthyosis vulgaris is a dermatological disease whose early diagnosis and treatment planning as a dentist will enhance growth and development of the maxilla and mandible, furthermore will prevent dental malocclusion. Preventive oral hygiene methods and regular follow up will reduce further decay and improve the aesthetics and function of the teeth.

References