Case Report

Orofacial Manifestations in a Child with Congenital Lamellar Ichthyosis: Practical Recommendations for Dental Management

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Abstract

Ichthyosis is a group of rare Mendelian disorders which affects cornification of the skin characterized by hyperkeratosis and/or scaling. The incidence of lamellar ichthyosis is estimated to be approximately 1 in 300,000 live births with no known sex predilection. The presenting features comprise a wide array of dermatological manifestations. This case illustrates findings in a 5-year-old child with the rare disorder, i.e., congenital lamellar ichthyosis with a history of rickets with interesting array of orofacial manifestations associated with the disease. The dental management may include a multispecialty approach for rehabilitation while using delicate approach to manipulate soft tissues and skin. We share a few practical recommendations which were helpful for allaying challenges we faced in this case.

Keywords: Autosomal recessive congenital ichthyosis, orofacial manifestations, practical recommendations for dental management, rare Mendelian disorder

Introduction

An ichthyosis is a group of rare Mendelian disorders affecting cornification of the skin characterized by hyperkeratosis and/or scaling.[1] Autosomal recessive congenital ichthyosis (ARCI) is a group of heterogeneous disorders that present at birth with generalized involvement of skin and lack manifestations in other organ systems.[2]

ARCI comprises a group of nonsyndromic ichthyosis that includes the phenotype spectrum of classic lamellar ichthyosis and nonbullous congenital ichthyosiform erythroderma, bathing suit ichthyosis, self-healing collodion baby, and harlequin ichthyosis.[3] The mutations in at least six different genes (transglutaminase I [TGM1], ABCA12, NIPAL4 or ichthyin, ALOXE3, CYP4F22, and ALOX12B) are reported to date, but the phenotype-genotype correlation is not fully proven.[4]

The incidence of lamellar ichthyosis is approximately 1 in 300,000 live births with equal sex predilection.[5] The severity of disease varies greatly from the mildest types which may be mistaken for normal, dry skin to life-threatening conditions such as harlequin ichthyosis.[6]

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This case report presents a case of lamellar ichthyosis with the orofacial manifestations and its management in the pediatric dental clinic along with simple experience-based practical recommendations for challenges faced during dental treatment.

Case Report

A 5-year-old boy reported to the Department of Pedodontics and Preventive Dentistry, with the chief complaint of multiple decayed teeth for 1 year. The child was born to a nonconsanguineous couple as a collodion baby. He had generalized scaling and dryness of skin since infancy and was diagnosed by the dermatologist as lamellar ichthyosis.

There was no similar familial history. The patients IQ and physical growth were normal. No ocular and otolaryngeal anomalies were detected. The patient was diagnosed with Rickets at the age of 1 year and is under treatment for the same.

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On extraoral examination, course dark scaly lesions were distributed generally but more prominent in the palmoplantar region. Thick, armor-like brown-gray scales were found covering the body and more prominent in the body flexures. The nails showed increased convexity and ridging [Figure 1]. The hair was sparse, dry, and gray in color. Patient reported less sweating and worsening of the condition during summer months.

The mouth opening was reduced with multiple fissures surrounding the oral cavity resembling a fish-like mouth. Angular cheilitis was present at the right corner of the mouth region [Figure 2].

Intraoral examination revealed multiple carious lesions with caries involving–54, 55, 64, 65, 73, 74, 75, 83, 84, and 85. High arched palate was found, no other soft-tissue anomalies were detected [Figure 3].

The management began with a preventive phase where diet counseling and education was done for the patient. The brushing technique was demonstrated, and oral hygiene education was done for the mother and child. To build rapport and gain confidence of the child, the 1st visit included a tour of the clinic, modeling, and tell-play-do approach. It was noted that, due to inability to sweat, the child had extensive hot flushes.

Thus, 2nd visit was performed where Casein phosphopeptide-amorphous calcium phosphate based varnish (GC Tooth Mousse, G C Dental, India) was applied using paint on method to all noncarious molar teeth. This was done to acquaint the child to the dental chair besides preventive value. A moist cloth dabbed in cold water was provided to the child underneath the drape which his mother applied to his forehead and neck region as required to allay the hot flushes. The dental chair chosen was the one next to the air-conditioner vent. Another problem we faced was the pull of the delicate skin around the oral muscosa, reduced mouth opening and chapping of lips. This was managed using repeated applications of cocoa butter every 5–7 min. A feather-touch like approach was used while retracting soft tissues and other oral manipulation.

At the third visit and fourth visits, restorations were done using Glass Ionomer Cement (GlasIonomer FX ULTRA, SHOFU) under moist cotton role isolation to cause minimal distress to the soft tissues. The appointment time was limited to 15 min. Over time the child showed tremendous understanding of the importance of dental care and remained positive and cheerful during the treatment period. Follow-ups are being held every month usually synced to the day and time he visits the Dermatologist for follow-ups.

Discussion

The term ichthyosis is derived from the Greek word "ichthys" meaning "fish" and refers to the similarity in appearance of the skin to fish scales.[7] Early reports of ichthyosis in the Indian and Chinese literature dating back to several 100 years.[8]

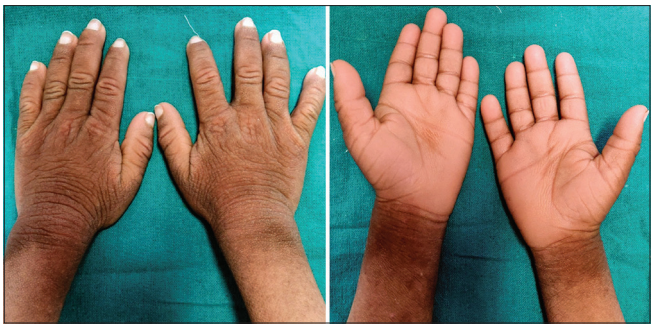

Figure 1: Brown scaly lesions over extensor (left) and flexor (right) aspect of arms with nail anomalies.

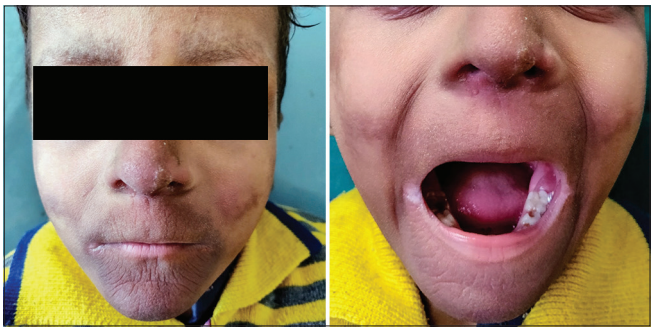

Figure 2: Extraoral view showing fish-like mouth with scales (left) and reduced mouth opening with angular cheilitis (right).

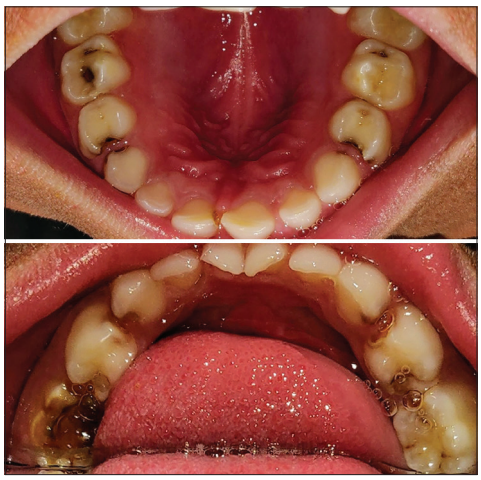

Figure 3: Intraoral occlusal view maxillary arch showing high-arched palate (top), mandibular occlusal view (bottom).

ARCI usually presents at birth and can progress into any one of the spectra of disorders.[1] Lamellar Ichthyosis is a type of ARCI. Mutation of six genes plays a role in lamellar ichthyosis. Of these, TGM1 is thought to be one of the major causes of lamellar ichthyosis. Since this enzyme plays a vital role in saliva secretion, case reports suggest xerostomia and delayed speech as dental manifestations of the disease.[2] Other dental findings include enamel hypoplasia, gingivitis, periodontitis, high caries incidence delayed eruption of primary and secondary dentition, bruxism, fish mouth appearance, bifid teeth and hyperkeratotic plaques on the tongue.[2,6,7,9]

The present case too showed high caries prevalence, high arched palate, decreased mouth opening and fish like mouth appearance.

The condition has been reported to be associated with bilateral ectropion (33%), diminished or absent sweating (10%), seasonal recurrence of dermatosis during summer (15%) and nail dystrophies (<5%).[10] In the present case, diminished sweating, worsening of dermatosis during summer months, and nail dystrophies were reported. Recently, it has been reported that children with congenital ichthyosis, especially those with pigmented skin types, are more prone to develop Vitamin D deficiency and rickets[11] as was seen in the presented case.

Topical emollients remain the cornerstone for the treatment of lamellar ichthyosis. Topical retinoids and Vitamin D3 are helpful along with other antioxidants. Angular cheilitis and facial dermatitis may occur as a side effect of oral retinoid therapy as was found in the presented case.

Among pharmacological agents in dental use, local anesthesia and antiobiotics are safe, but titration of the dose should be done to prevent hepatotoxicity. The dental surgeon should also be careful during manipulation of the fragile and tender perioral skin as abovementioned alongside other recommendations based on our experience.[1,6]

Conclusion

Despite the rarity of ARCI, various cases show oral manifestations associated to the condition. The dental rehabilitation plays a crucial role in the functional, physical, and psycho-social rehabilitation of the patient and the practical recommendations which increase patient comfort and cooperation should be borne in mind.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Nair KK, Kodhandram GS. Oral manifestations of lamellar ichthyosis: A rare case report. Indian J Paediatr Dermatol 2016;17:283-6.
2. Ramar K, Annamalai S, Hariharavel VP, Aravindhan R, Ganesh C, Ieshwaryah K. Oral manifestation of autosomal recessive congenital ichthyosis in a 2-year-old patient. Case Rep Dent 2014;2014:483293.
3. Oji V, Tadini G, Akiyama M, Blanchet Bardon C, Bodemer C, Bourrat E, *et al*. Revised nomenclature and classification of inherited ichthyoses: Results of the First Ichthyosis Consensus Conference in Sorèze 2009. J Am Acad Dermatol 2010;63:607-41.
4. Fischer J. Autosomal recessive congenital ichthyosis. J Invest Dermatol 2009;129:1319-21.
5. Akiyama M, Sawamura D, Shimizu H. The clinical spectrum of nonbullous congenital ichthyosiform erythroderma and lamellar ichthyosis. Clin Exp Dermatol 2003;28:235-40.
6. Rathi NV, Rawlani SM, Hotwani KR. Oral manifestations of lamellar ichthyosis: A rare case report and review. J Pakistan Assoc Dermatol 2013;23:99-102.
7. Pranitha V, Reddy BV, Daneswari V, Deshmukh SN. Lamellar Icthyosis - A case Report. J Clin Diagn Res 2014;8:ZD01-2.
8. Hernández-Martín A, Cuadrado-Corrales N, Ciria-Abad S, Arias-Palomo D, Mascaró-Galy JM, Escámez MJ, *et al*. X-linked ichthyosis along with recessive dystrophic epidermolysis bullosa in the same patient. Dermatology 2010;221:113-6.
9. Vergotine RJ, de Lobos MR, Montero-Fayad M. Harlequin ichthyosis: A case report. Pediatr Dent 2013;35:497-9.
10. Bloom D, Goodfried MS. Lamellar ichthyosis of the newborn. The "collodion baby": A clinical and genetic entity; report of a case and review of the literature with special consideration of pathogenesis and classification. Arch Dermatol 1962;86:336-42.
11. Sethuraman G, Marwaha RK. Vitamin D, bone health and congenital ichthyosis. Indian J Dermatol Venereol Leprol 2016;82:249-51.