

Taurodontism Associated with Ichthyosis Vulgaris in Two Siblings: A Case Report

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Abstract

Ichthyosis vulgaris is a common genetic keratinization disorder, clinically characterized by scaling, especially on the flexor limbs, and with palm plantar hyperlinearity. The prevalence of ichthyosis vulgaris is 1:250 and the prevalence of taurodontism varies from 2.5 to 3.2% in permanent teeth while in deciduous teeth, it is 0.54%. Most ichthyosis patients do not need any special dental care, but the dentist should be aware of any underlying medical issues. Oral and dental findings reported in ichthyosis patients included gingivitis, periodontitis, enamel hypoplasia, high caries incidence, delayed primary and permanent teeth eruption, bruxism, bifid teeth, the irregular morphology of teeth, fish mouth appearance, mouth breathing, xerostomia, and hyperkeratotic plaques on the tongue and very rarely taurodontism. This paper describes the clinical and radiographic features and endodontic management of taurodontism associated with ichthyosis vulgaris in two young siblings.

Keywords: Genetic, Ichthyosis vulgaris, Keratinization, Siblings, Taurodontism.

INTRODUCTION

Ichthyosis forms a large heterogeneous group of cornification disorders characterized by the accumulation of hyperkeratotic scales on the skin's surface.^[1] The term "ichthyosis" is derived from the Greek word "ichthys" meaning fish because this condition is characterized by fish-like dry, scaly, and thickened skin (hyperkeratosis). Ichthyosis vulgaris is most commonly manifested by xerosis, partial or total skin lesions, keratosis pilaris, and palmoplantar hyperlinearity along with generalized scaly dermatosis without the component of erythema and a varying degree of erythroderma. The prevalence of ichthyosis vulgaris is 1:250.^[2] Taurodontism is a dental anomaly characterized by enlargement of the pulp chamber, which may approximate the root surface. It can occur as an isolated entity or it can be associated with genetic disorders/syndromes.^[3] Taurodontism can be of three types, hypotaurodontism, mesotaurodontism, and hypertaurodontism. The prevalence varies from 2.5 to 3.2% in permanent teeth while in deciduous teeth, it is 0.54%.^[4] This is one of the rare cases of ichthyosis vulgaris associated with taurodontism involving all four permanent first molars. The patient's younger sibling also had similar features of ichthyosis vulgaris and taurodontism.

CASE REPORT

A 10-year-old boy reported to the Department of Pediatric Dentistry, with the chief complaint of pain in the lower right

back tooth region of the jaw for 1 week. He was born to a non-consanguineous couple. Generalized scaling was seen all over the body (Figure 1). He was diagnosed as a case of ichthyosis vulgaris by the dermatologist and was treated with different moisturizers. Family history revealed that his younger brother also had the same disorder (Figure 2) and the child's father also had taurodontic all four first permanent molars. The patient did not suffer from any other systemic abnormalities. The patient's physical growth and IQ were normal. Cutaneous examination reveals dry scales covering the entire body surface, that is, the face, the body trunk, the shoulder, and the upper and lower extremities (Figure 1). The patient's eyebrows were scanty and his hair was dry and sparse. No nail abnormalities were found. The patient had a normal mouth opening with fissures at the corner of the mouth bilaterally.

Intraoral examination revealed multiple carious teeth. Grossly decayed 55, 63, 64, over-retained 74, pre-shedding mobility with 53, deep dentinal caries with 26 and 46, space loss in

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Figure 1: Extraoral images of a 10-year-old boy with clinical features of ichthyosis vulgaris, (a) Scales on his face and scanty eyebrows, (b) Scales over the chest region, (c) Scales on back and shoulder region, (d) Scales on upper extremities, (e) Scales on lower extremities

relation to 35, presence of plaque and gingivitis, and poor oral hygiene. No other soft-tissue abnormalities were detected. Radiographic examination showed mixed dentition without any enamel defects. Space loss was seen in relation to 35 and taurodontism with 16, 26, 36, and 46 (Figures 3 and 4). Pediatricians' and dermatologist's opinions were taken. The treatment plan was explained to the parents and written consent was obtained from parents. Oral prophylaxis was done. Root canal therapy was performed with 26 and 36 followed by composite restoration. Extraction of grossly decayed 55, 63, 64, and retained 74, mobile 53 were done under local anesthesia. The brushing technique was demonstrated and oral hygiene instructions were given.

Endodontic management of taurodontic 26 and 46 teeth: The anesthetized tooth in relation to 46 was isolated under a rubber dam. Access opening was done and pulp tissue was extirpated using an H-file. The pulp was voluminous and to ensure its complete removal, 2.5% sodium hypochlorite was used as an irrigant initially to soften the pulp. After pulp extirpation, further irrigation was done with normal saline and 0.2% chlorhexidine. The floor of the huge pulp chamber could not be visualized, which on further exploration revealed two divisions in the buccal and one in the distal side. The huge pulp chamber walls were circumferentially shaped using "K" files of standard ISO taper. Finally, irrigation was performed using sterile saline. A calcium hydroxide dressing was placed for a week. The canals were prepared for obturation. The custom-made gutta-percha/roll-cone technique was used for obturation

and was prepared on a glass slab using a stainless-steel spatula. The Gutta-percha cone was trial fitted, intraoral periapical radiograph was taken, and obturation was done. Cold lateral compaction technique was used to obturate the remaining canal (Figure 5). After 1 week, the pulpally involved tooth in relation to 26 was endodontically treated with a similar custom-made roll-cone technique. The teeth 26 and 46 were restored with posterior composite resin (Figure 5). The patient will be followed up periodically for the development of occlusion.

DISCUSSION

Ichthyosis vulgaris is a common genetic keratinization disorder that is clinically characterized by scaling, especially on the flexor limbs, and with palm plantar hyper-linearity. It is caused by heterozygous loss-of-function mutations in the filaggrin gene (FLG) which is found in the epidermal differentiation complex on chromosome 1q21.^[2] Six major distinct clinical subtypes of this disorder are known, to till date. The most severe form is harlequin ichthyosis (HI, MIM/242500) followed by lamellar ichthyosis (LI, MIM/242300); congenital ichthyosiform erythroderma (CIE, MIM/242100); epidermolytic ichthyosis (EI, MIM/113800); recessive X linked ichthyosis (RXLI, MIM/308100) to the mildest form of ichthyosis vulgaris.^[1] Ichthyosis vulgaris is caused by heterozygous loss-of-function mutations in the FLG which results in absolute or complete deficiency of filaggrin resulting in the disruption of the epidermal barrier,^[5] which leads to clinical features of ichthyosis vulgaris. Similarly, findings



Figure 2: Extraoral image of the child's younger sibling showing (a) Scales on his face and scanty eyebrows, (b) Scales over the chest region, (c) Scales on back and shoulder region, (d) Scales on upper extremities, (e) Scales on lower extremities

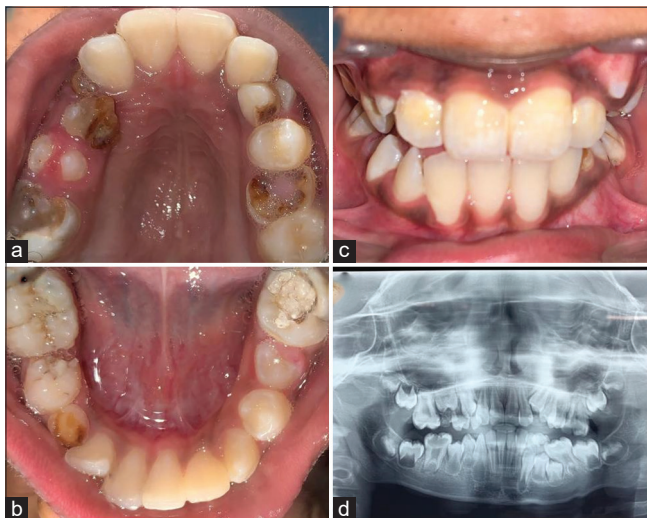


Figure 3: Pre-operative intraoral features seen in a 10-year-old boy. (a) Maxillary teeth, (b) Mandibular teeth, (c) Teeth in occlusion, (d) OPG Showing taurodontism in relation to 16, 26, 36, 46 teeth

were seen in scales over the upper and lower extremities, the trunk region, the shoulder region, and the face.

There is very little knowledge about the dental manifestations and their management in patients with ichthyosis vulgaris. The current case describes the association of taurodontism with ichthyosis vulgaris. Other dental manifestations of ichthyosis vulgaris as reported by various authors in the literature are a high risk of dental caries, gingivitis, periodontitis, enamel

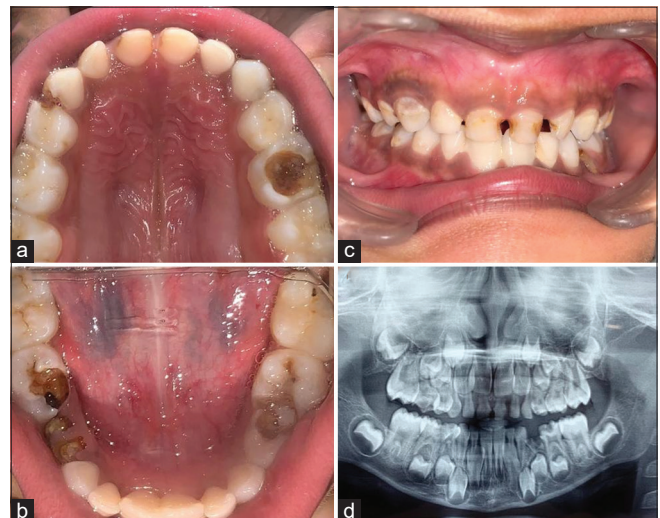


Figure 4: Pre-operative intra-oral features seen in the 7-year-old child (sibling) with ichthyosis vulgaris. (a) Maxillary teeth, (b) Mandibular teeth, (c) Teeth in occlusion, (d) OPG Showing taurodontism in relation to 16, 26, 36, 46 teeth

hypoplasia, delayed eruption of teeth, and presence of the plaque on the surface of the tongue.^[1] Bolgul *et al.* reported missing teeth, carious teeth, and retained deciduous teeth in a 14-year-old boy.^[6] Over-retained teeth and delayed eruption of permanent teeth could alter the normal growth and development of the jaw leading to malocclusion. Similarly, in our case, multiple carious teeth, taurodontism, and space loss in the lower left back tooth region were present.

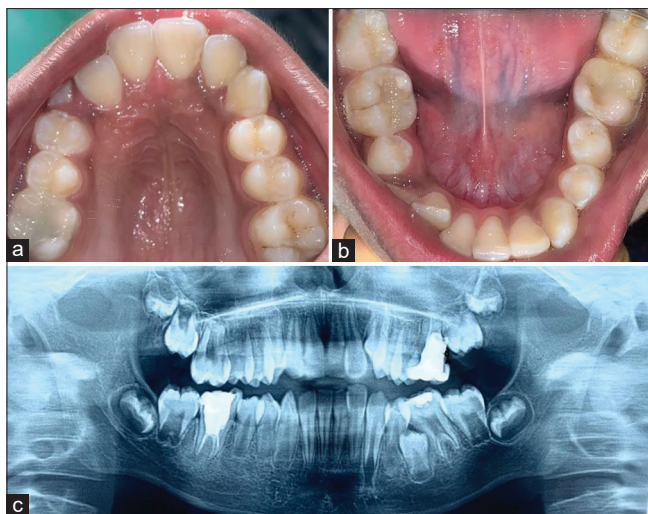


Figure 5: Post-operative intra-oral images of the 10-year-old boy with ichthyosis vulgaris. (a) Maxillary teeth, (b) Mandibular teeth, (c) OPG showing endodontically treated 26 and 46

Dental treatment was initiated with a preventive approach, and required extractions were carried out. Endodontic treatment in taurodont teeth has been described as complex and difficult.^[7] Durr *et al.* suggested that the morphology could hamper the location of the orifices, thus creating difficulty in instrumentation and obturation.^[8] Since the pulpal chamber was huge, mechanical debridement and shaping were achieved with circumferential filing using “K” files of standard ISO size and taper. Sodium hypochlorite irrigation was limited to the initial use followed by saline irrigation. Custom-made gutta-percha (roll-cone technique) was used for obturation along with the lateral compaction technique as described by Prakash *et al.*^[9] Thermoplasticized gutta-percha obturation techniques have been used by other clinicians.^[10]

The treatment of ichthyosis vulgaris includes the removal of the scalliness externally and the maintenance of a soft and pliable condition of the skin. The mainstay of therapy for ichthyoses is moisturizers. They work by increasing the flexibility of the epidermis, hydrating, and restoring epidermal barrier function. Systemic therapy with oral retinoids is one of the most effective therapies available for most of the ichthyoses. The retinoids have potential side effects such as angular cheilitis. The dentist should treat these affected areas by advising the application of petroleum jelly or local anesthetic gel. The erupting permanent

teeth should be monitored to prevent any type of malocclusion. Care must be taken to avoid manipulating the patient’s skin during dental treatment, particularly in the perioral areas, since affected areas can be tender or friable.^[1] Dentists should moisturize the oral and perioral tissues with petroleum jelly before commencing the treatment to avoid discomfort to the child.^[11]

CONCLUSION

Ichthyosis vulgaris is a dermatological disease that 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 reduce further decay and improve the esthetics and function of the teeth.

SOURCE OF SUPPORT

None.

CONFLICT OF INTEREST

None.

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