Congenital Teeth and Nail Changes with Palmar Keratoderma: A New Entity or a Rare Association?

Dear Editor,

A 12-year-old male child, born out of nonconsanguineous marriage, presented to our Dermatology clinic complaining of thickening of palmar skin for the past 3 months. Cutaneous examination revealed multiple hyperkeratotic plaques over bilateral palms, extending to the dorsum of hands [Figure 1]. He was asymptomatic, and no acquired cause for this could be determined. On further evaluation, he was found to have hypodontia. Only the deciduous maxillary second molar teeth was present [Figure 2]. Nail examination showed yellowish-brown discoloration of the nail plate and marked longitudinal ridges [Figure 3]. Toenails were affected more severely than fingernails and there was more significant involvement of the nails in the past. The nail and teeth findings were present since he was 5 years of age. He did not complain of intolerance to heat or abnormal perspiration. The hair, built, and facies were normal. None of his family members complained of similar problems. KOH examination of nail clippings and culture did not reveal any fungal hyphae or spores. Biopsy from the skin over the palm showed hyperkeratosis, acanthosis, a normal granular layer, and mild perivascular lymphohistiocytic infiltrate. Orthopantomogram revealed missing permanent teeth. Genetic testing could not be done due to limited resources.

To summarize, the patient had abnormal teeth and nail development with palmar hyperkeratosis. Since two ectodermal structures were affected, the patient was diagnosed with Ectodermal dysplasia (ED). However, it was difficult to arrive at a specific diagnosis as genetic confirmation could not be done. Besides genetic counselling, the patient was prescribed emollient and keratolytic for palmar keratoderma and referred to orthodontics for restorative surgery.

The EDs comprise a large, heterogeneous group of inherited disorders that are defined by primary defects in the development of two or more tissues derived from embryonic ectoderm. EDs related to teeth and nails are Witkop and Fried tooth and nail syndrome (TNS), Tricho-onychodental (TOD) syndrome, Curry--Hall syndrome, Deafness, and onycho-osteodystrophy with

Table 1: Differential diagnosis of congenital teeth and nail changes with or without palmoplantar keratoderma

Features	Witkop TNS	Fried TNS[1]	TOD syndrome[2]	Rapp-- Hodgkin Syndrome[3]	DOOR Syndrome[4]	Curry--Hall Syndrome[5]	Clouston Syndrome[6]
Gene	MSX1	MSX1		TP63	TBD1D24	EVC	GJB6
Inheritance	Autosomal dominant	Autosomal recessive	Autosomal dominant, described in the Japanese family	Autosomal dominant	Autosomal recessive	Autosomal dominant	Autosomal dominant
Tooth	Malformed/unerupted permanent teeth, widely spaced, conical shape, dental caries	Absent primary teeth, conical/ peg-shaped	Hypoplastic enamel, dysplastic dentine	Anodontia, hypodontia, conical teeth	Enamel hypoplasia, high arched palate	Conical teeth	Normal
Nail	Koilonychia, brittle, marked ridging, pitting	Thin & concave nails	Thin & dysplastic nails	Anonychia, hyponychia, dystrophic nails	Onychodystrophy, hypoplastic nails	Dysplastic nails	Dystrophic nails
Hair	Normal, mild brittleness	Significant sparseness of hair growth	Scanty, fine, curled hair	Sparse, friable, pili torti	Normal	Normal	Generalized hypotrichosis
Sweat glands	Normal	Normal	Hypohidrosis	Hypohidrosis	Normal	Normal	Normal
Facies	Normal	Everted lips	Normal	Small mouth, narrow nose	Antimongoloid slant, prominent epicanthal fold, asymmetric face	Normal	Normal
Other				Cleft palate, cleft lip	Mental retardation, epilepsy, SNHL, osteodystrophy	Polydactyly, short limbs	Palmoplantar keratoderma, Eccrine syringofibroadenoma, Epidermoid cyst

TOD: Tricho-onychodental, DOOR: Deafness and onycho osteodystrophy with mental retardation, Witkop TNS: Witkop tooth and nail syndrome, Fried TNS: Fried tooth and nail syndrome, SNHL: Sensorineural hearing loss

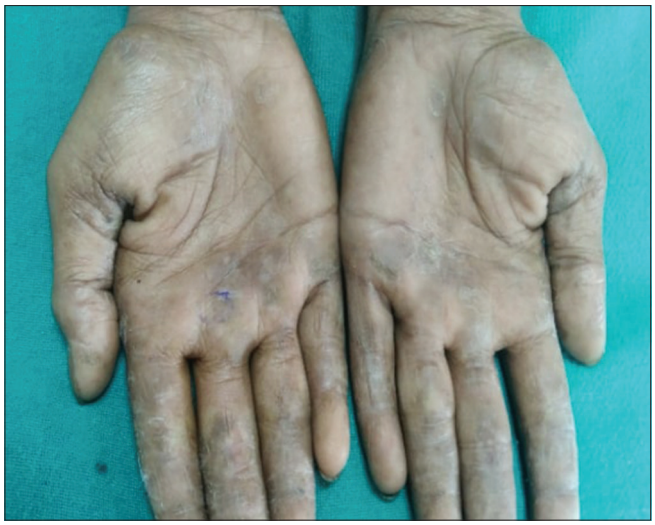

Figure 1: Multiple discrete to coalescent hyperkeratotic plaques over bilateral palms with mild scaling

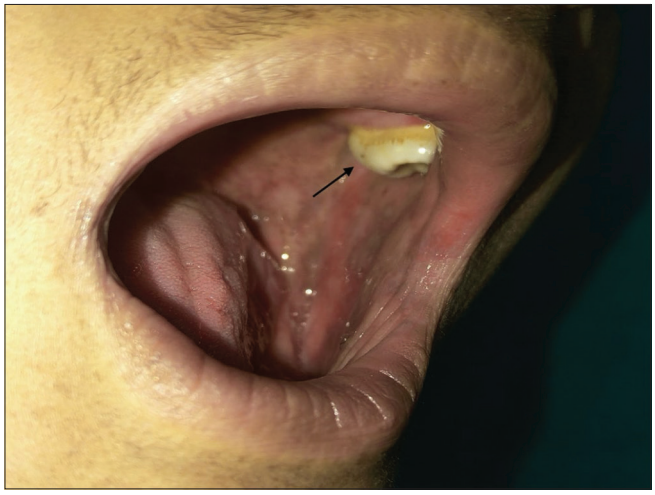

Figure 2: Oligodontia with only second maxillary molar with dental caries

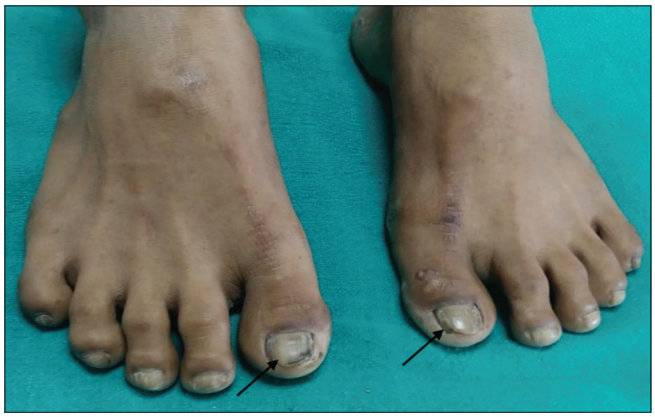

Figure 3: Yellowish-brown discoloration of nail plates, coarse pitting, and increased longitudinal ridges

mental retardation (DOOR) syndrome and Rapp Hodgkin disease, which are discussed in Table 1. Both Witkop and Fried TNS involve teeth and nail structures but the additional involvement of hairs and abnormal facial development in Fried TNS differentiate it from Witkop TNS.[1] Other similar conditions like TOD syndrome[2] and Rapp Hodgkin disease[3] present with hypohidrosis, DOOR syndrome[4] patients' have abnormal facies, mental retardation, and hearing loss, while Curry--Hall syndrome[5] presents with polydactyly and short limbs, thus ruled out. Clouston syndrome[6] is one of the ED which presents with palmar keratoderma, but normal hair and teeth abnormality rules out its possibility in the present case. Hence, the closest differential diagnosis for the present case is Witkop TNS, with a rare association with palmar keratoderma. Although the possibility of an associated new mutation and new clinical condition or variant of existing ectodermal dysplasia could not be ruled out here.

The treatment for TNS is generally symptomatic and supportive.[7] It primarily involves dental restoration and prevention of dental caries. Lubrication of the nails and trimming to keep them short and smooth to prevent fungal infections is essential. Understanding the psychosocial condition of these patients is crucial, as the unaesthetic appearance that accompanies them often has adverse psychological effects.

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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Conflicts of interest

There are no conflicts of interest.

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