Trachyonychia with Juvenile Pityriasis Rubra Pilaris

Pityriasis rubra pilaris (PRP), papulosquamous disorder of unknown etiology, is characterized by well-defined, orange-red scaly plaques showing keratotic follicular papules, palmo-plantar waxy keratoderma, erythroderma. Nail changes have been reported with Type 1 PRP (Griffiths Classification) as distal discoloration, yellow-brown subungual hyperkeratosis, nail-plate thickening, and splinter hemorrhages while distal onycholysis and onychogryphosis have been reported with Type 5 PRP.[1,2] There are no previous reports of nail matrix involvement or trachyonychia in patients with PRP.[3]

A three-and-a-half-year-old boy presented multiple well-defined. discrete with orange-red plaques, with prominent erythematous border, distributed over hands and feet, along with nail changes for the past 2 months. There was no preceding history of vaccination, sore throat, any other illness, or drug intake. There were characteristic islands of sparing, and erythematous follicular papules to plaques were present over the knees and dorsa of hands and feet as well [Figure 1]. Nail examination showed erythematous plaques extending onto proximal nail folds (PNF), ragged cuticles, and 'sand-papered' nail plates with increased longitudinal ridging. pitting and fine onychoschizia, suggestive of trachyonychia [Figure 2a]. There was increased longitudinal curvature of the thumb and index fingernails with minimal subungual hyperkeratosis present only in the toenails [Figure 3a]. There were no associated scalp lesions, areas of hair loss, pigmentary changes, eczematous lesions, mucosal lesions, or cutaneous infections in the child or any of the family members. The direct microscopic examination of nail clippings from the toenails (after incubating with potassium hydroxide solution) did not reveal any fungal elements.

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clinical diagnosis of PRP with trachyonychia was made. Skin biopsy showed mild acanthosis with orthokeratosis and parakeratosis, no dilated vessels, or neutrophilic infiltrate. Clinical-pathological correlation facilitated a diagnosis of circumscribed juvenile PRP (Type 4). Given the benign nail changes at a young age, a nail biopsy was not attempted. As both conditions tend to have a good prognosis, the child was started on topical calcipotriol with clobetasol cream, white soft paraffin cream, and oral biotin (5 mg per day). There was a marked improvement in both skin and nails [Figures 2 and 3] at 2 months, resolution over 6 months, and no relapse over further 2 months of follow-up.

Nails are involved in 13%–33% patients with PRP, with nail changes preceding skin lesions in 2%.[1,4] Predominantly nail bed involvement has been reported, more commonly with Type 1 (classic adult PRP), with concomitant involvement of the skin of the palms and soles. Another study had described nail involvement, ranging from thickening and discoloration to complete sloughing in 72% of the patients.^[5] Yellowish-brown discoloration, subungual hyperkeratosis, and splinter hemorrhages have been reported in the literature, signifying nail bed involvement, thus, suggesting that PRP involves the nail bed mainly, concurrent with the involvement of the palmo-plantar skin. This is in contrast to nail psoriasis which tends to involve the nail matrix and/or skin of proximal nail folds. Nail involvement in Type 1 PRP had shown parakeratosis, acanthosis, and focal basal liquefaction with intermittent collections of keratohyaline granules, with dermal mononuclear inflammatory infiltrate on histopathology.[1] In contrast, our patient showed trachyonychia involving 20 nails, indicating that the nail matrix was the predominant site of involvement, which has previously not been reported to the best of our knowledge.

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Chander Grover, Geetali Kharghoria¹

Department of Dermatology and Venereology, University College of Medical Sciences and Guru Teg Bahadur Hospital, 'Department of Dermatology and Venereology, All India Institute of Medical Sciences, Delhi. India

Address for correspondence:

Dr. Chander Grover, Professor, Dermatology and STD, UCMS and GTB Hospital, Delhi - 110 095, India. E-mail: chandergroverkubba@ rediffmail.com

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Figure 1: (a-c): Clinical photograph showing all 20-fingernail involvement with trachyonychia and ragged cuticles. Orange-red colored plaques are seen involving the palms, soles, with similar scaly papules and plaques over dorsa of hands extending onto the proximal nail folds

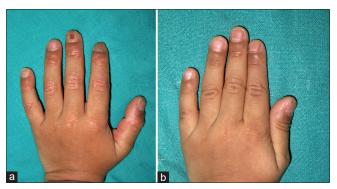


Figure 2: (a and b) Response to topical therapy and oral biotin in the skin lesions and nail lesions over the hand



Figure 3: (a and b) Response to topical therapy and oral biotin in the skin lesions and nail lesions over the feet

Trachyonychia is a manifestation of intermittent inflammation of the nail matrix of varying severity.

Mild trachyonychia seen in our case suggested an intermittent inflammation of the matrix, related to PRP involving the overlying skin. The risk of long-term or irreversible damage to the nail is low in such cases, andtherapy needs to be kept as simple as possible, like in our case.

To conclude, this case showed an unusual temporal correlation of the onset and resolution of trachyonychia with juvenile PRP. It serves to highlight the good prognosis with an acceptable response to topical therapy.

Author contributions

Both the authors have equally contributed to the design of the manuscript, writing of the manuscript, and are accountable for all aspects of the work along with ensuring accuracy or integrity of the manuscript.

Consent statement

The patient's guardian has given written informed consent to the publication of the details of the patient in the manuscript.

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Conflicts of interest

There are no conflicts of interest.

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