Pigmented Aqua-Exacerbated Symmetrical Acral Hyperkeratosis: Washing Shows Its True Colour

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Dear Editor,

Pigmented aqua-exacerbated symmetrical acral hyperkeratosis is a rare dermatosis, typically occurring in young or middle-aged men.[1] The disease is also known as *acquired symmetrical acrokeratoderma* in older texts, and the majority of the cases were reported in Han-Chinese ethnicity since 2010 except an Indian case series comprising of five cases.[2] We also encountered an Indian patient with similar clinical presentation.

An 18-year-old male presented with multiple asymptomatic hyperpigmented lesions over the dorsum of hands and feet for a 2-year duration with a history of summer exacerbation and spontaneous resolution during winters. Family history was noncontributory except the presence of atopic dermatitis in a younger sibling. Clinical examination revealed, multiple well-defined hyperpigmented plaques present bilaterally over the volar aspect of distal forearms, dorsae of hands, and feet without palmoplantar involvement. The lesions developed greyish-white maceration immediately after hand washing, which rapidly reversed upon drying [Figure 1a and b]. Clinical differentials considered were acral acanthosis nigricans, pigmented aqua-exacerbated symmetrical acral hyperkeratosis, and aquagenic syringeal acrokeratoderma. Histopathological features were non-specific. The findings were basket-weave hyperkeratosis, normal granular layer, irregular acanthosis, and mild perivascular lympho-mononuclear infiltrates [Figure 2a and b]. The patient was clinically diagnosed as *pigmented aqua-exacerbated symmetrical acral hyperkeratosis* since our clinical findings were consistent with the previous studies.[2-5] The patient was counseled about the disease course and potential treatment options. Since he was not willing for long-term oral retinoid therapy, the patient was treated with emollients without much improvement. The lesions resolved gradually, but completely after 3-4 months with the change of weather to winter [Figure 3a and b]. In the past two years of followup, the patient experienced similar summer relapses and spontaneous winter remissions.

Pigmented aqua-exacerbated symmetrical acral hyperkeratosis is a rare disease, characterized by multiple, symmetrically distributed, hyperpigmented hyperkeratotic plaques over acral sites sparing palms and soles.[2] The proposed pathogenic mechanisms are hyperproliferation of malassezia yeasts, increased trans-epidermal water loss due to aquaporin-3 down expression, and recently, missense mutation of transcription factor-4 (TCF-4) gene.[3,4] TCF-4 is a part of Wnt/β-catenin pathway, involved in the regulation and differentiation of epithelial stem cells, and its mutation may cause defective expression of loricrin, involucrin, and keratin-1, leading to hyperkeratotic presentation.[4]

Histopathological features of this disease are nonspecific.[2,4] Various immunohistochemistry studies showed higher expression of keratin-14, keratin-16, Melan-A, and filaggrin and down expression of aquaporin-3.[3,5]

The clinical differentials are acral acanthosis nigricans, aquagenic syringeal acrokeratoderma, addisonian pigmentation, psoriasis, and frictional keratosis. *Pigmented aqua-exacerbated symmetrical acral hyperkeratosis* differs from these

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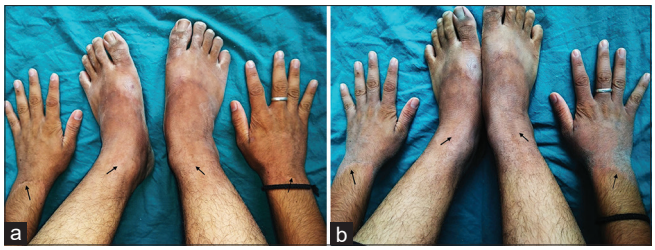

Figure 1: Baseline clinical images. (a) Multiple well-defined brownish black hyperkeratotic plaques over dorsal aspect of both hands and feet (Black arrows). (b) Greyish white maceration of the pre-existing plaques (Black arrows) after immersion of hands and feet in water

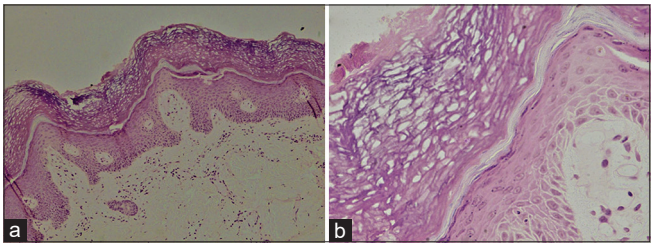

Figure 2: (a) Photomicrograph showing basket weave hyperkeratosis, mild acanthosis, mild perivascular lymphomononuclear infiltrate (H and E; 40×) (b) Photomicrograph showing basket weave hyperkeratosis, underlying confluent parakeratosis, preservation of granular layer, and irregular acanthosis (H and E; 100×)

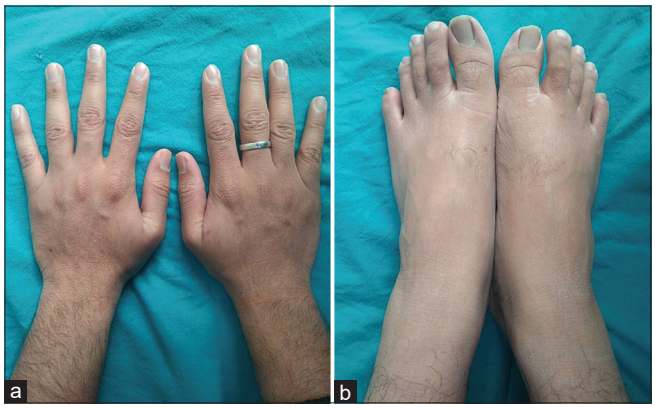

Figure 3: Post recovery clinical images. (a and b) Apparently normal hands and feet in winter without any pigmentation or textural change

by its asymptomatic nature, palmo-plantar sparing, post-immersion maceration, and spontaneous winter remission.[2] This entity is very resistant to treatment, but in a few cases, topical keratolytics and oral retinoids have shown some efficacy.[2]

Postimmersion maceration is a nonspecific phenomenon, characterized by the whitish discoloration of lesions with textural change on exposure to water, which returns back to original state immediately after drying.[2] The exact pathophysiology behind this unique phenomenon is unclear and warrants further detailed research. Although this phenomenon is not specific to any disease, seasonal occurrence of this phenomenon exclusively points towards Pigmented Aqua-Exacerbated Symmetrical Acral Hyperkeratosis.

Besides its predominant Chinese distribution, emerging reports from other countries question its exclusive ethnic predisposition and the possibility of under diagnosis in other ethnicities.[2,3] Thus, vigilant clinical examination and observation of this peculiar phenomenon may be useful to diagnose this relatively new entity.

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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