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Case report

A Case of Symmetrical Acrokeratoderma in an Indian Patient: A Decade-Long Misdiagnosis

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KEYWORDS

Symmetrical acrokeratoderma, pigmented aqua-exacerbated symmetrical acral hyperkeratosis, aquagenic maceration

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ABSTRACT

Symmetrical acrokeratoderma (SA) is a rare dermatosis characterized by symmetric, hyperkeratotic plaques on the dorsum of the hands, wrists, feet, and ankles, often misdiagnosed due to its resemblance to other acral hyperkeratotic conditions. We report a case of a 20-year-old Indian male with a decade-long history of asymptomatic, bilaterally symmetrical plaques on the wrists and ankles, exhibiting aquagenic maceration and seasonal variation. Clinical findings, supported by histopathology showing hyperkeratosis, acanthosis, and mild perivascular inflammation, confirmed the diagnosis. Treatment with oral acitretin (25 mg) resulted in marked improvement, which was maintained at the third month of follow-up. This case highlights the importance of recognizing SA's distinct clinical features, such as the aquagenic response and seasonal aggravation, to avoid misdiagnosis. We emphasize the need for increased awareness and further research into its pathogenesis and effective therapies, particularly in Indian populations where reports are scarce.

1. Introduction

Symmetrical acrokeratoderma (SA), also known as pigmented aqua-exacerbated symmetrical acral hyperkeratosis, is a rare dermatosis first described in Han-Chinese populations in 2010 (1). It is characterized by symmetric, non-pruritic, brownish-black hyperkeratotic plaques on the dorsum of the hands, wrists, feet, and ankles, sparing the palms and soles. Lesions exhibit transient maceration in warm weather and improve in cooler weather (2).

Although the exact prevalence of SA remains unknown, most documented cases involve young males of Asian descent, particularly from China and India (3, 4). Its rarity, along with overlapping features with other dermatoses such as aquagenic acrokeratoderma, palmoplantar keratodermas, and acanthosis nigricans, contributes to underdiagnosis and misclassification. This case report describes a classic presentation of SA in a young Indian male and integrates histopathologic and immunologic findings with current literature to raise awareness and highlight potential diagnostic pitfalls.

2. Case Report

A 20-year-old male presented to our outpatient dermatology clinic with bilateral, symmetrical, asymptomatic brownish plaques over the dorsum of both wrists (Fig. 1, 2) and lateral ankles (Fig. 3). The lesions first appeared at 10 years of age and had gradually increased in size and pigmentation.

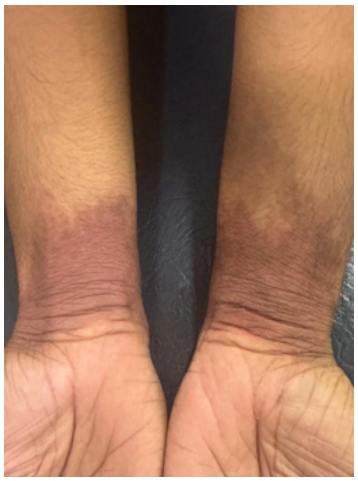

Fig. 1. *Hyperpigmented plaques noted over the flexors of both wrists before water immersion test.*

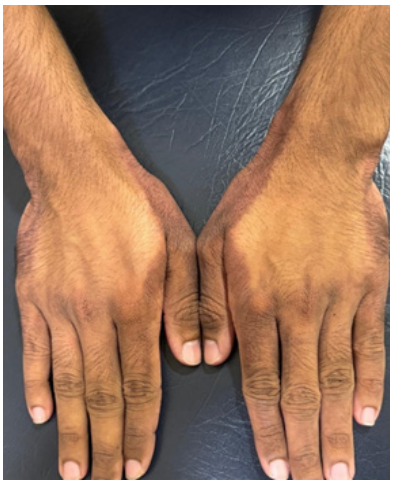

Fig. 2. *Hyperpigmented plaques noted over the lateral aspects of both hands.*

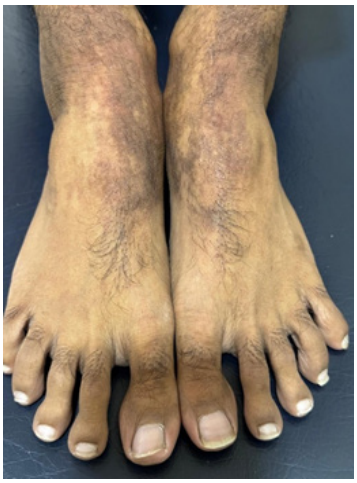

Fig. 3. *Hyperpigmented plaques noted over the dorsum of both feet.*

There was no family history of similar skin disorders. The patient denied pruritus, pain, or burning. He had no history of systemic comorbidities or history of prior drug intake. Notably, he reported that the lesions became whitish and macerated after bathing, returning to their original appearance once dry. He also noted worsening of the lesions during summer and spontaneous improvement during winter.

He had been previously prescribed moderate-potency topical corticosteroids, with no significant improvement.

Cutaneous examination revealed well-demarcated, bilaterally symmetrical, brownish hyperkeratotic plaques localized to the lateral aspect of wrists and ankles. Palms and soles were spared. All mucosae and nails were normal.

A water immersion test resulted in the lesions turning pale white and wrinkled within 2–3 minutes (Fig. 4). The changes reversed within 30 minutes of drying, confirming the aquagenic response.

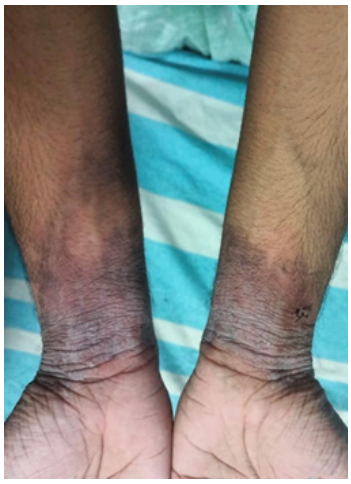

Fig. 4. *Macerated plaques noted over the flexors of both wrists after water immersion test.*

Systemic examination was unremarkable.

A Skin biopsy was done from the affected area and revealed hyperkeratosis, focal parakeratosis, acanthosis and normal granular layer. Dermis showed few dilated capillaries with mild perivascular and periadnexal chronic inflammatory infiltrate (Fig. 5).

No fungal elements were seen in the section.

These findings aligned with previous reports describing epidermal hyperplasia with chronic inflammatory changes (1, 2).

The patient was initiated on Oral Acitretin and showed significant improvement on 2nd month of follow up with complete resolution of lesions over the feet (Fig. 6, 7).

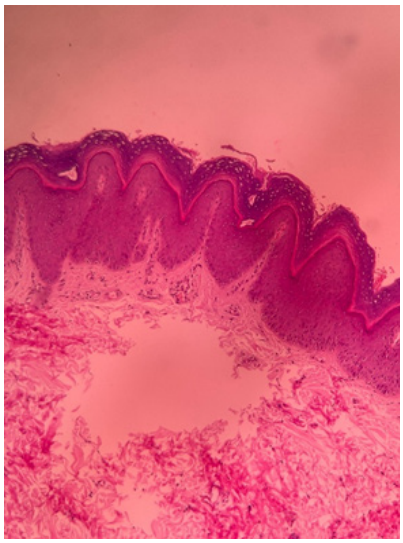

Fig. 5. *Histopathology of the section showing Hyperkeratosis, focal parakeratosis and mild perivascular inflammatory infiltrate.*

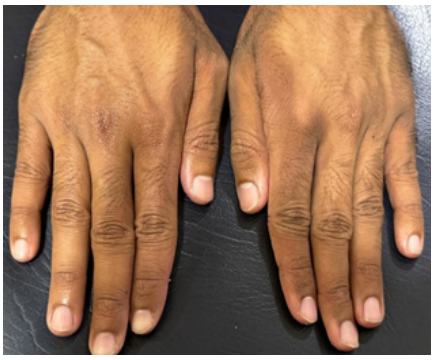

Fig. 6. *Follow up 2 months later shows marked improvement of lesions*

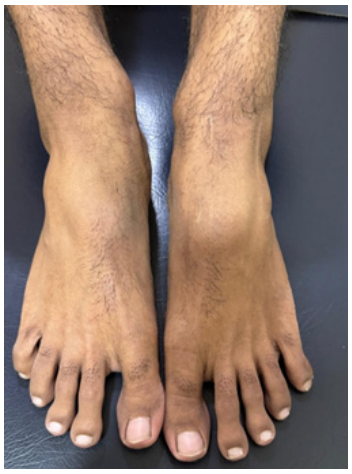

Fig. 7. *Complete resolution of lesions over feet after 2 months of Oral Acitretin.*

4. Discussion

Symmetrical acrokeratoderma has been predominantly reported in East Asian populations, particularly China. Indian case reports remain few, but suggest a similar presentation and course (2, 3).

The condition usually manifests in late childhood or adolescence, with a chronic, non-progressive course. Seasonal aggravation is commonly reported, with flare-ups during humid, warm months and resolution in colder climates. SAK presents as asymptomatic brownish plaques distributed symmetrically over the acral sites like wrists and ankles.

Our patient's clinical course—onset at 10 years, bilaterally symmetric acral involvement, aquagenic maceration, and seasonal variation (Fig. 1-4), is consistent with classical presentations.

The pathogenesis remains incompletely understood. Proposed mechanisms include:

- **Genetic predisposition:** Mutations in the Transcription Factor 4 (TCF4) gene may lead to overexpression of epidermal differentiation markers such as KRT1 and KRT14, loricrin, involucrin which results in epidermal hyperkeratosis (4).
- **Aquaporin-3 dysregulation:** Downregulation in lesional and perilesional skin causes increased trans-epidermal water loss which increases dryness and structural fragility (4).

Table I summarizes the differential diagnoses of symmetrical acrokeratoderma based on clinical features and distinguishing factors (5).

Table I. *Comparison of differential diagnoses for SA.*

Differential Diagnosis	Typical Sites Affected	Common Age/ Sex Profile	Key Clinical Features	Symptoms During Water Immersion	Other Associated Features
Symmetrical Acrokeratoderma	Dorsum of hands and feet; spares palms and soles	Young males, especially Asians	Brownish-black symmetric hyperkeratotic plaques; exacerbates in summer	Whitish maceration after water, reverting on drying	Seasonal variation; asymptomatic
Aquagenic Syringeal Acrokeratoderma	Palms and soles	More common in adolescent females	Burning, tingling sensation, translucent whitish papules after water exposure	Prominent symptoms; "hand-in-the-bucket sign"	Often associated with cystic fibrosis

Differential Diagnosis	Typical Sites Affected	Common Age/ Sex Profile	Key Clinical Features	Symptoms During Water Immersion	Other Associated Features
Acral Acanthosis Nigricans	Acral areas, commonly around joints	Middle-aged to elderly adults	Velvety, hyperpigmented plaques, usually non-palpable thickening	No effect on water immersion	Associated with obesity, insulin resistance
Palmoplantar Keratoderma	Palms and soles	Common in adolescents	Diffuse or focal hyperkeratosis; itching, burning, or pain	No change or effect with water immersion	May have genetic background; sometimes painful

Histopathologically epidermal hyperkeratosis, irregular acanthosis, orthokeratosis of stratum corneum and perivascular lymphocytic infiltrate in papillary dermis is noted, consistent with the findings observed in our case (Fig. 6).

Currently, there is no definitive treatment. Topical corticosteroids, are typically ineffective. Oral Acitretin 25mg was used in our case following which patient showed marked symptomatic improvement. Other topicals like Retinoids, Salicylic acid and Urea provide symptomatic relief. Botulinum toxin has been used recently which showed improvement in skin texture, thickness, pigmentation and extent of lesions (6, 7). A study done by Keshavamurthy et al. (2016) on Indian case reports on symmetrical acrokeratoderma described the most common sites of presentations of Symmetrical acrokeratoderma in Indian patients and their response to treatment (8) (Table II).

Table II. *Clinical presentation and treatment response in indian patients with SA.*

Patient No	Age at Onset (Years)	Duration of Disease (Years)	Sites Involved	Treatment Received	Response
1	11/M	4	Knuckles, interphalangeal joint, wrist, elbow	Topical retinoic acid + 10% urea, Acitretin 10 mg/day 16 weeks	Complete clearance
2	21/M	2	Knuckles, wrist, ankle	Acitretin 25 mg/day 12 weeks, topical 10% urea	Complete clearance; seasonal recurrence
3	24/M	6	Dorsum of hand and feet, wrist, flexural forearm, ankle, shins, knee	Topical retinoic acid 0.1% gel, 10% urea	Mild improvement; relapse in summer
4	28/M	5	Dorsum of hand and feet, forearm	Acitretin 25 mg 4 weeks, topical 10% urea	Complete clearance; seasonal recurrence
5	31/M	4	Dorsum of hand and feet, forearm, shin	Acitretin 25 mg 8 weeks then 10 mg 16 weeks, topical 10% urea	Complete clearance; seasonal recurrence

Patient education remains vital, particularly regarding avoidance of prolonged water exposure and managing expectations about the chronic course.

5. Conclusion

Symmetrical acrokeratoderma is an underrecognized, benign dermatological entity. Characterized by symmetric acral plaques with water-induced maceration and seasonal variability, it is often misdiagnosed or given ineffective treatment due to lack of existing literature.

Recognition of its unique clinical features, particularly the aquagenic maceration, seasonal pattern and its association with atopy is crucial for dermatologists. Histopathology supports the diagnosis, though is not pathognomonic.

This case adds to the growing evidence of symmetrical acrokeratoderma in Indian populations and underscores the need for awareness and further study into its pathogenesis and treatment.

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