

Case Report

Dyshydrosiform Immunoglobulin A Pemphigus: An Uncommon Yet Intriguing Case Report

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ABSTRACT

A 24-year-old female presented with relapsing and remitting, fluid-filled lesions on her palms and soles for 2 years. These lesions, which began as vesiculopustules, coalesced into larger plaques and eventually led to post-inflammatory hyperpigmentation. Histopathology revealed hyperkeratosis, subcorneal pustules and neutrophilic exudates, while direct immunofluorescence demonstrated intercellular immunoglobulin A (IgA) deposits in the upper epidermis. These findings were suggestive of dyshydrosiform subcorneal pustular dermatosis-type IgA pemphigus, a rare autoimmune blistering disorder. Treatment with dapsone led to significant clinical improvement within 2 months. IgA pemphigus is a chronic condition with a wide clinical spectrum. The involvement of acral sites, as seen in this case, is an uncommon presentation, emphasising the need for further investigation into the pathogenesis and variability of this rare entity.

Keywords: Autoimmune blistering disorder, Dapsone, Dyshydrosiform SCPD type immunoglobulin A pemphigus, Immunoglobulin A pemphigus, Subcorneal vesicle

INTRODUCTION

IgA pemphigus is a rare autoimmune blistering disease (AIBD) characterised by intercellular IgA deposition within the epidermis. Among its clinical variants, dyshydrosiform presentations limited to palms and soles are exceptionally uncommon.

CASE REPORT

A 24-year-old woman with no significant medical history presented with a 2-year history of recurring, painless, fluid-filled lesions on her palms and soles. These began as small vesicles or pustules and gradually merged into larger plaques. She denied any lesions on mucosal surfaces or other parts of the body. Past use of unspecified topical treatments provided no relief.

On examination, annular plaques with vesiculopustular margins were seen on the right middle finger, with similar lesions on the lateral sides of the left index and middle fingers. A larger plaque with intact vesicles, erosions and yellowish crusting was noted on the lateral aspect of the right toe. Post-inflammatory hyperpigmentation was present on the left foot [Figure 1a-e].

All routine investigations were within normal limits, and both 10% potassium hydroxide (KOH) smear and fungal culture yielded negative results. Haematology and gastroenterology evaluations, including serum studies, ruled out immunoglobulin A (IgA) gammopathy and inflammatory bowel

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Figure 1: (a) Right middle finger showing annular plaque with vesiculopustules at the margin of plaque. (b) Left middle and index finger show a few scattered vesiculopustules on the dorsal aspect with one on the middle finger coalescing to form an annular plaque. (c) Annular scaly plaque on the right middle finger seen on the ventral aspect. (d) Dorsum of the right toe shows plaque with crusting while there is evident post-inflammatory hyperpigmentation on the left foot. (e) Lateral aspect of the right toes shows a large plaque with interspersed vesicles and crusting.

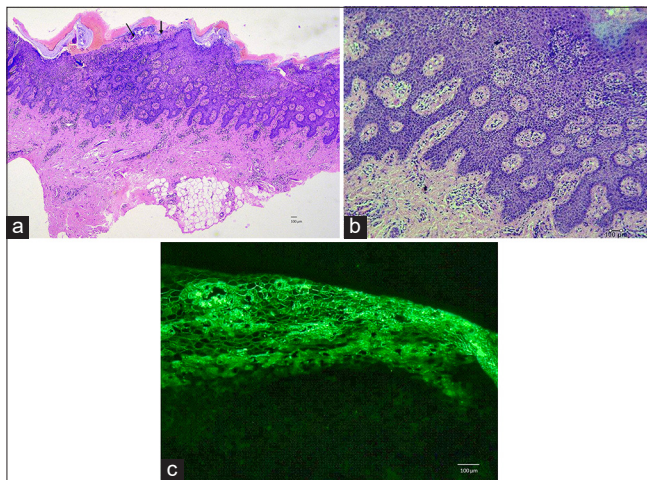


Figure 2: (a) Section shows skin with subcorneal abscess formation (arrows) and mild dermal perivascular lymphocytic infiltrate (Haematoxylin & Eosin [H&E] x50). (b) Section shows skin with epidermal acantholysis (black stars). H & E, x100. (c) Epidermis with fish net appearance with inter-epidermal deposition of IgA (fluorescein isothiocyanate, x200).

disease. The differential diagnosis for vesiculopustules on hands and feet included dyshydrosiform bullous pemphigoid, dyshydrosiform pemphigus vulgaris, pompholyx, bullous tinea pedis/manuum and palmoplantar pustulosis.

Histopathologic examination of a palmar vesicle biopsy specimen showed hyperplastic and hyperkeratotic epidermis. Numerous subcorneal pustules containing neutrophilic exudates and proteinaceous material were present [Figure 2a and b]. Dermis showed mild perivascular lymphocytic infiltrate. Periodic acid-Schiff (PAS) staining showed no

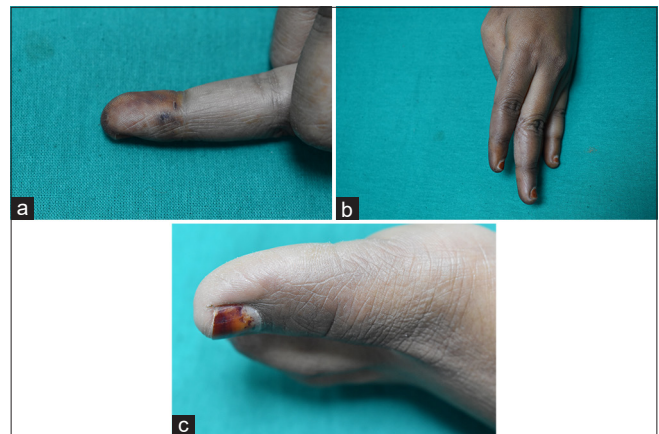


Figure 3: (a) Healing of plaque of right middle finger with crust formation. (b) Healing of lesions on the left middle and index finger. (c) Healing of lesion on the right toe (lateral view).

evidence of fungal hyphae. Direct immunofluorescence (DIF) of perilesional skin showed intercellular IgA deposits, in a fish-net pattern, predominantly in the upper layers of the epidermis [Figure 2c]. DIF for IgG, IgM and C3 was negative, and indirect immunofluorescence (IIF) also showed no detectable antibodies. Enzyme-linked immunosorbent assay (ELISA) was negative for antibodies against desmoglein 1 and 3 and testing for antibodies against desmocollins could not be performed due to unavailability. A final diagnosis of dyshydrosiform subcorneal pustular dermatosis (SPD)-type IgA pemphigus was rendered. The patient was initiated on 100 mg of dapsone, with improvement observed after 2 months [Figure 3a-c].

The plan involves continuing 100 mg daily for 1 more month, followed by a maintenance dose of 50 mg daily for an additional 3 months to sustain remission.

Table 1: Comparative analysis of acral vesicular/bullous dermatosis.

Condition	Clinical features	Histopathology (H&E)	Standard DIF profile	Rare/Atypical DIF patterns	Contrast with IgA pemphigus
Autoimmune conditions					
IgA pemphigus (SPD type) ^[1-3]	Annular plaques, vesiculopustules; rare acral-only	Subcorneal pustules, acantholysis	Intercellular IgA (fish-net, upper epidermis)	None	-
Dyshydrosiform bullous pemphigoid	Pruritic tense fluid filled vesicles/bullae on palms/soles	Subepidermal blister, eosinophils	Linear IgG/C3 at BMZ	Granular C3/IgM at DEJ (rare)	Linear BMZ deposits; eosinophil-rich
Dyshydrosiform pemphigus vulgaris	Acral vesicles, mucosal involvement	Suprabasal acantholysis	Intercellular IgG/C3 (lower epidermis)	Focal intercellular IgM (atypical)	IgG-driven; lower epidermal acantholysis
Lichen planus pemphigoides	Palmar/plantar bullae, lichenoid papules	Subepidermal blister, lichenoid interface	Linear IgG/C3 at BMZ	Shaggy fibrinogen at BMZ	Lichenoid pattern; no intercellular IgA
Epidermolysis bullosa acquisita	Acral bullae at trauma sites	Subepidermal blister, minimal inflammation	Linear IgG at BMZ (dermal side, salt-split skin)	None	Subepidermal, trauma-prone; dermal side IgG
Bullous lupus erythematosus	Photosensitive vesicles, bullae on normal or erythematous background at acral sites, mucosal lesions	Subepidermal blister, neutrophilic infiltrate	Linear/granular IgG, IgA, C3 at BMZ	Full-house pattern	Multisite systemic features, ANA+
Non-autoimmune conditions					
Bullous tinea pedis/manuum ^[5]	Vesiculopustules, scaling, +fungal studies	Subcorneal pustules, spongiosis, hyphae	Negative	Granular C3/IgM at DEJ or perivascular	Fungal elements present; non-specific DIF
Dyshidrotic eczema ^[1]	Small deep seated pruritic vesicles distributed symmetrically on acral areas, chronic recurrence	Spongiosis, vesiculation	Negative	Granular C3/IgM at DEJ (occasional)	Spongiotic features; no pustules
Palmoplantar pustulosis ^[1]	Sterile pustules on palms/soles	Intraepidermal neutrophilic pustules	Negative	Granular C3 at DEJ (rare)	Psoriasiform pattern; no intercellular IgA
Bullous fixed drug eruption	Recurrent acral bullae at same site	Interface dermatitis, necrotic keratinocytes	Granular C3/IgM at DEJ	Intercellular C3/IgA (very rare)	Drug history, interface pattern
Bullous scabies	Eczematous and vesicubullous lesions, burrows	Spongiotic vesicles with eosinophils, mites	Negative	Granular IgM/C3 at DEJ or perivascular	Scabies mite identifiable on H&E
Herpesvirus infections	Grouped vesicles, erosions	Ballooning degeneration, multinucleated cells	Negative	Granular IgM/C3 at DEJ	Cytopathic viral changes; PCR/culture positive
Erythema multiforme (bullous)	Target lesions, sometimes acral bullae, mucosal involvement	Interface dermatitis, necrotic keratinocytes	Negative	Granular IgM/C3 at DEJ	Acral targetoid lesions; drug or viral trigger

(Contd...)

Table 1: (Continued).

Condition	Clinical features	Histopathology (H&E)	Standard DIF profile	Rare/Atypical DIF patterns	Contrast with IgA pemphigus
Vesiculobullous darier disease	Flaccid vesicles or crusted papules on palms (Acral haemorrhagic variant); flexures and seborrheic areas; rare acral	Focal suprabasal acantholysis, corps ronds and grains	Negative	Focal intercellular IgG/IgA (very rare)	Genetic; acantholysis with dyskeratosis; lacks intercellular IgA pattern of SPD-type IgA pemphigus
Hand foot mouth disease	vesicles on palms, soles, oral mucosa	Intraepidermal vesicles with ballooning degeneration, necrotic keratinocytes, mild perivascular infiltrate	Negative	Non-specific granular IgM or C3	Viral, DIF negative, Spongiosis, ballooning degeneration, necrotic keratinocytes on histopathology

H&E: Haematoxylin and eosin, DIF: Direct immunofluorescence, SPD type: Subcorneal pustular dermatosis type, BMZ: Basement membrane zone, DEJ: Dermoepidermal junction, ANA: Antinuclear antibody, PCR: Polymerase chain reaction, IgA: Immunoglobulin A

DISCUSSION

IgA pemphigus is a rare autoimmune bullous disorder, typically beginning around age 45, with no gender bias. It has two primary subtypes: SPD and intraepidermal neutrophilic (IEN), along with less common variants such as IgA pemphigus vegetans, vulgaris and foliaceus. Clinically, IgA pemphigus often presents with vesicles, pustules, annular or circinate erythematous plaques and herpetiform lesions. Itching is reported in about half of the patients, and some may experience pain. Commonly affected areas include the trunk, flexures, scalp and intertriginous regions, while mucosal involvement is rare.^[1] A systematic review by Kridin *et al.* reported palmoplantar involvement in 3.3% of IgA pemphigus cases, highlighting its potential, though uncommon, acral presentation.^[1]

Histologically, SPD shows subcorneal pustules and acantholysis with intercellular IgA deposits in the upper epidermis, as seen in the present case. In contrast, IEN has deeper epidermal pustules with more widespread IgA deposition.^[2] Desmocollin 1 is the main target antigen in SPD, while desmogleins 1 and 3 have been implicated as the antigen in IEN.^[3] IgA titres are usually low, and IIF is positive in only about 50% of cases, making DIF the more sensitive test.^[4] This might explain the negative IIF in the present case.

IgA pemphigus has been linked to systemic conditions, notably ulcerative colitis (except in SPD), multiple myeloma and B-cell lymphoma. Treatment primarily involves dapsone, often with low-dose steroids. Although specific long-term data on the dyshydrosiform variants are lacking, available literature suggests that IgA pemphigus overall follows a

relatively benign course with good response to dapsone and a more favourable prognosis than classical pemphigus.^[1,4]

Dyshydrosiform variants of bullous pemphigoid and pemphigus vulgaris are rare, and eccrine gland antigens may play a role in their pathogenesis. IgA pemphigus shows subcorneal pustules with acantholysis on haematoxylin and eosin (H&E) and a specific “fishnet” intercellular IgA pattern on DIF, while tinea pedis/manuum exhibits subcorneal pustules with spongiosis and fungal hyphae on H&E, rarely non-specific linear/granular C3 at the dermo-epidermal junction/perivascular location on DIF and positive KOH, PAS and culture, which were negative here.^[5] While non-specific immunoreactants in non-autoimmune conditions like eczema rarely show linear/granular pattern at BMZ, or perivascular DIF patterns, a few autoimmune and rare non-autoimmune disorders may exhibit intercellular deposition. Table 1 has been added to illustrate these distinctions. Unlike such mimickers, our case showed consistent clinical, histopathological and intercellular DIF findings with a favourable response to dapsone, supporting the diagnosis of IgA pemphigus.

CONCLUSION

To summarize, this case highlights a rare presentation of IgA pemphigus affecting only the palms and soles. With few studies that have been done on IgA pemphigus till date, a deeper exploration is required to look into the varied spectra of this relatively uncommon autoimmune blistering disease.

Ethical approval: Institutional Review Board approval is not required.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for 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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Use of artificial intelligence (AI)-assisted technology for manuscript preparation: The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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