

# Pemphigus presenting with prominent neutrophilic pustules mimicking two diseases

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Sir,

Pemphigus is a group of immunobullous disorders with intraepidermal blisters that may mimic some other conditions clinically and histologically. Hereby, we report a case of pemphigus masquerading as subcorneal pustular dermatoses (SCPD) and IgA pemphigus confirmed by immunofluorescence.

A 68-year-old female presented with a sudden onset of itchy, pus-filled lesions and redness over the face, neck, both upper limbs, lower limbs, and trunk for three days. On examination, multiple discrete to grouped pustules, several erosions, and crusting on a background of diffuse erythema over the face, neck, bilateral upper limbs, lower limbs, and trunk were noted. (Figs. 1a and 1b). The mucosa and scalp were unaffected. No history of drug intake prior to the onset of the lesion was present. The patient gave a history of itchy, fluid-filled lesions affecting similar areas four years previously. Histopathological (HPE) examination of the vesicle showed features of bullous pemphigoid, and the patient was initiated on dapsone, later switched to dexamethsone cyclophosphamide pulse (DCP) therapy i/v/o dapsone hypersensitivity reaction. She was lost to follow-up after eight cycles but continued taking oral cyclophosphamide 50 mg per day on her own for 2.5 years; meanwhile, the patient had two episodes of exacerbation and was treated with steroids.

Based on the current clinical presentation, the possibility of SCPD and IgA pemphigus was considered and investigated accordingly. Tzanck smear showed only neutrophils. HPE of the pustule showed a



**Figure 1:** (a and b) Multiple discrete to grouped pustules on a background of erythema (red arrow) present on the face, neck, bilateral upper limbs, lower limbs, and trunk. Some erythematous erosions (black arrow) and yellowish to brown crusts (yellow arrow) present on the trunk.

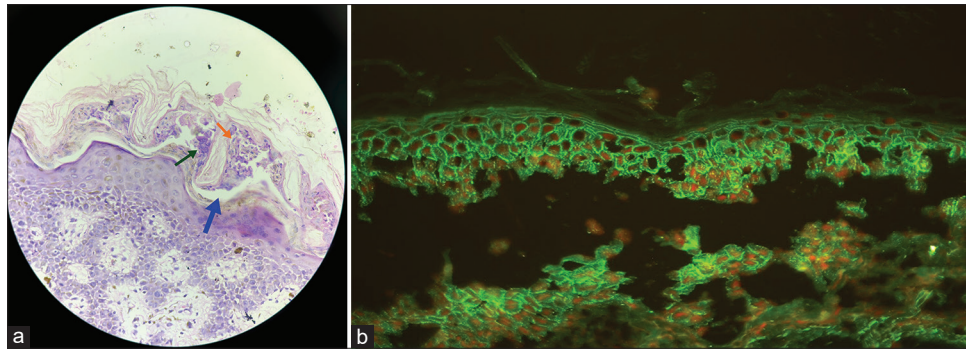
subcorneal split, neutrophils, and some acantholytic cells (Fig. 2a). Direct immunofluorescence (DIF) examination of perilesional skin showed intercellular staining of epidermis with IgG and C3 (Fig. 2b).

A typical case of pemphigus presents with widespread flaccid blisters, painful erosions, and prominent mucosal involvement. However, atypical presentations are not uncommon. A similar case of a young male patient, showing features of SCPD and IgA pemphigus clinically, was reported, showing DIF features of pemphigus. The patient showed a dramatic improvement with oral corticosteroids [1]. Similarly, a case of an elderly female who presented with features of sweet syndrome initially later developed a crusting similar to pemphigus foliaceus (PF). This

**How to cite this article:** Hegde P, Raviumar M, Rao R. Pemphigus presenting with prominent neutrophilic pustules mimicking two diseases. Our Dermatol Online. 2026;17(2):270-271.

**Submission:** 08.08.2025; **Acceptance:** 09.10.2025

**DOI:** 10.7241/ourd.20262.28



**Figure 2:** (a) Epidermis showing a subcorneal split (blue arrow) containing neutrophils (green arrow) and some acantholytic cells in the blister cavity (orange arrow) (H&E; 40x). (b) Intercellular staining of the epidermis with IgG and C3 with a fish net pattern (DIF; 10x).

was confirmed on DIF, and the patient responded well to DCP therapy [2]. Another case, of a young boy presenting with typical features of PF with prominent pustules progressing to erythroderma, is mentioned in the literature. The patient showed a dramatic improvement with colchicine after failing to respond to conventional treatment [3]. A middle-aged man with pre-existing psoriasis vulgaris lasting for 35 years presented with a new onset of erythematous, scaly lesions with prominent pustules. Interestingly, the patient did not respond to conventional psoriasis therapy but showed features of PF on DIF [4]. A case of PF in a young female presenting with predominant neutrophilic pustules confirmed on DIF was reported [5]. In the majority of such cases of an atypical presentation of pemphigus, DIF is of great importance, as in our case, to identify the correct diagnosis, as pemphigus requires to be treated with specific immunosuppressive therapy.

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**Source of Support:** This article has no funding source.

**Conflict of Interest:** The authors have no conflict of interest to declare.