# Subcorneal Pustular Dermatosis Occuring in Association with Pyoderma Gangrenosum and Rheumatoid Arthritis: A Triple Whammy!


## Abstract

Neutrophilic dermatoses are a wide group of disorders encompassing indolent to severely disabling conditions. A co-existence of two such conditions, pyoderma gangrenosum (PG) and subcorneal pustular dermatosis, necessitates a thorough investigation for IgA dysglobulinemia. We report a middle-aged woman who developed PG following 18 years of (undiagnosed) subcorneal pustular dermatosis, along with rheumatoid arthritis, a known association of PG.


**Keywords:** *Neutrophilic dermatoses, pyoderma gangrenosum, subcorneal pustular dermatoses*


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## Introduction

Subcorneal pustular dermatosis (SCPD) belongs to a primarily epidermal infiltration type of neutrophilic dermatoses, whereas pyoderma gangrenosum (PG) belongs to a dermal infiltrate variety of neutrophilic dermatoses, with absence of vasculitis.[1] Co-existence of PG and SCPD, along with rheumatoid arthritis (RA), is a rare occurrence. Furthermore, the clinicians also need to be cognizant of the association of neutrophilic dermatoses with paraproteinemias and investigate the patients adequately.

## Case Report

A 38-year-old female presented with a history of extremely pruritic lesions all over the body since 18 years, along with painful ulcers over lower legs for 8 months. She had been treated with antibiotics and antiscabetics by various primary physicians with no significant relief. She also had morning stiffness and arthralgias in both knees and elbows and small joints of both hands for 5 years, which had been diagnosed as RA and maintained on nonsteroidal anti-inflammatory drugs. She had no history of gluten intolerance, recurrent diarrhoea or constipation. Past medical history was significant for hypertension controlled with antihypertensives, deranged lipid profile controlled with statins, and obesity with a body mass index of 33.6 kg/m$^2$.

On examination, she had small discrete flaccid pustules, along with crusted and eroded papules over the buttocks, abdomen, posterior aspect of thighs, and a few over ankles and upper limbs [Figure 1]. A large, well-defined ulcer of size 15 cm × 13 cm was present on the extensor aspect of right leg with bluish undermined edges and violaceous borders [Figure 2]. In addition, she had two smaller similar ulcers: over left foot in-step and on the same leg distally.

Tzanck smear from pustule revealed few acantholytic cells and neutrophils, while bacterial culture was negative. Routine hematological and biochemical profile was normal while antinuclear antibody and antineutrophilic cytoplasmic antibody were negative; C3/C4 levels were within normal limits and rheumatoid factor and Anti-cyclic citrullinated peptide antibody (anti-CCP) were both positive. Desmoglein 1 and 3 enzyme-linked immunosorbent assay was negative, Serum IgG Tissue Transglutaminase (TTG) was 2.59 U (normal <20), serum IgA TTG was 5.45 U (normal <20), and IgA endomysial antibodies were negative, thus ruling out dermatitis herpetiformis. Serum immunoglobulin profile for IgG, and IgM was normal and serum protein electrophoresis did not reveal any abnormal bands. Her urine protein electrophoresis



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was significant for elevated total protein of 1080 mg/24 hours but without any monoclonal band.

Skin biopsy from a pustule revealed a subcorneal blister containing neutrophils and occasional acantholytic cells, while superficial dermis showed mild edema and moderate perivascular mixed inflammatory infiltrate comprising of lymphocytes and many neutrophils [Figure 3]. The direct immunofluorescence was negative for IgG/IgA/C3 on two occasions.

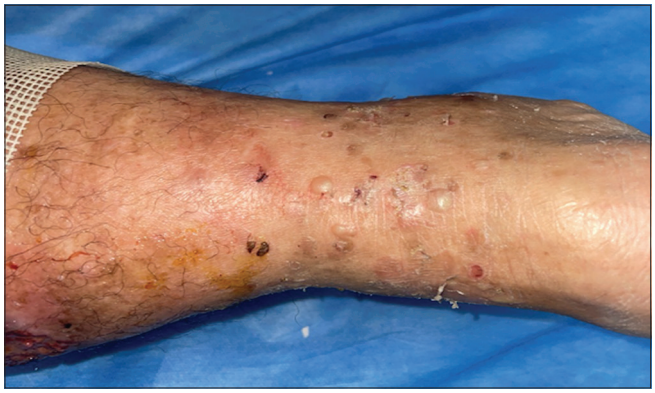

**Figure 1: Multiple flaccid pustules varying in size between 10 to 15 mm, with associated superficial crusts on skin of the anterior aspect of right leg**

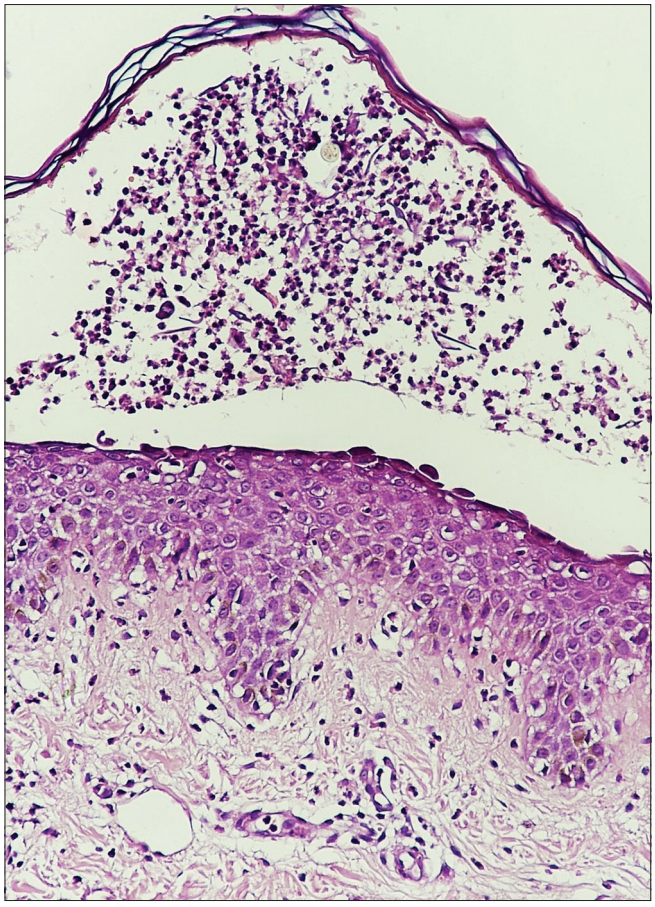

**Figure 3: Subcorneal pustule filled with neutrophils with evidence of acantholysis in underlying granular layer. (H and E, 200x)**

Skin biopsy from an ulcer showed a large focus of necrosis in the dermis rimmed by proliferating capillary channels and a dense inflammatory infiltrate comprising of neutrophils and lymphocytes [Figure 4]. There was no granuloma, fungal colonies, or Acid fast bacilli (AFB) identified in the smear.

Based on the clinical presentation and investigations, a diagnosis of SCPD with PG and RA was made. Her pustular

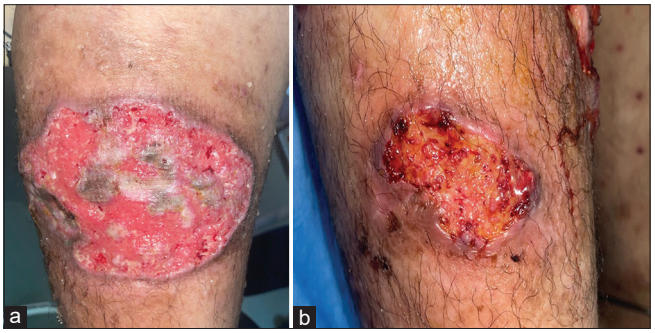


**Figure 2: (a) A large, well-defined ulcer of size 15 x 13 cm, present on the extensor aspect of right leg with irregular margins and bluish undermined edges. (b) Another ulcer of size 8x9 mm present on the lateral aspect of right leg**

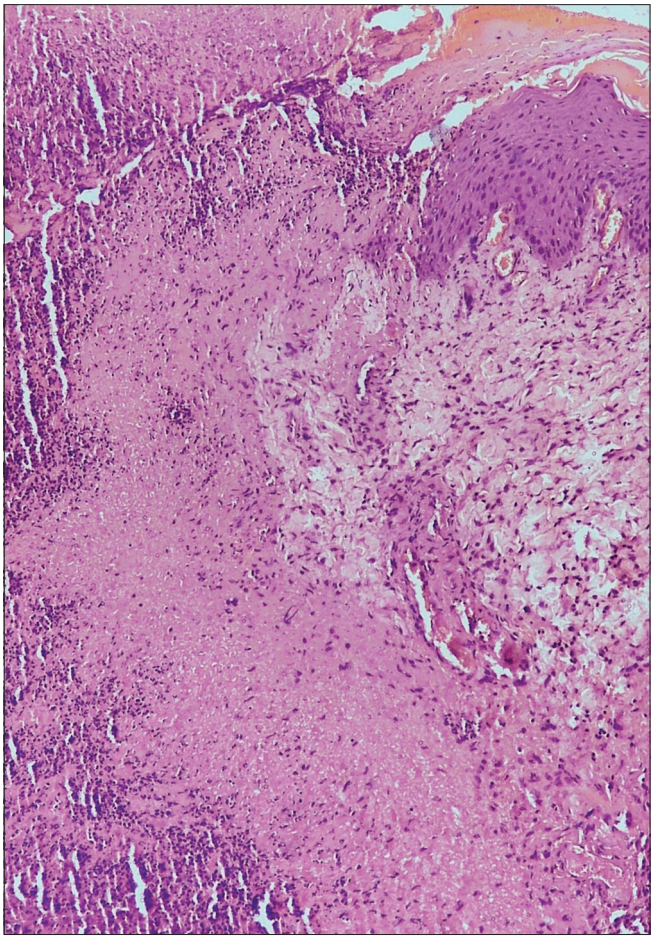

**Figure 4: Abrupt ulceration of epidermis with fibrinosuppurative exudates and extensive necrosis in underlying dermis. (H and E, 200x)**

lesions rapidly resolved on commencing treatment with dapsone 100 mg/day. Cyclosporine was started at a dose of 300 mg/day (about 3 mg/kg/day) in view of PG, along with daily dressing with Placentrex gel and paraffin gauze. The dose was tapered to 250 mg/day after 3 weeks, as islands of skin started appearing within the ulcer. Further tapering was done gradually and cyclosporine was discontinued after 3 months following complete resolution of the ulcer [Figure 5]. During this time, her SCPD remained in remission. However, the patient missed dapsone for a week in-between, leading to a new crop of pustular lesions, while still on cyclosporine 2 mg/kg. Reintroduction of Dapsone led to resolution again. RA has remained well-controlled with nonsteroidal anti-inflammatory drugs so far.

## Discussion

Our patient presented with relapsing and remitting recurrent crops of pustules for a duration of 18 years. A differential of dermatitis herpetiformis, IgA pemphigus, pemphigus foliaceous, and SCPD was kept clinically and the final diagnosis of SCPD was reached after obtaining the results of biopsy and Direct immunofluorescence (DIF).[2] IgA pemphigus was an important clinical differential, but a negative DIF ruled it out. Acantholytic cells, sparsely seen in our patient, are less commonly observed in SCPD; however, they may be seen in older lesions.[3-5] Exquisite response to Dapsone lent further support to the diagnosis.

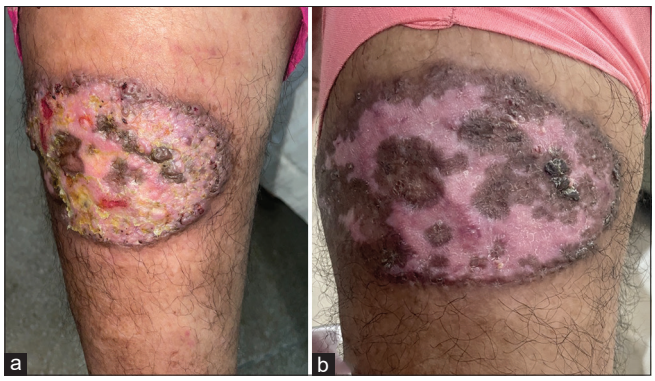


**Figure 5: (a) Ulcer on the anterior aspect of right leg with hyperpigmented margins and in stages of healing, with associated granulation tissue, (b) 10 weeks post-treatment: Ulcer with increased post-inflammatory hyperpigmentation in the center and periphery, without associated granulation tissue**

Co-association of two neutrophilic dermatoses as seen in this case (PG and SCPD) is possibly a pointer to an existent underlying immune dysregulation. The long interval between onset of SCPD and PG in the patient is unusual. Notably, most of the cases of co-association[6] of SCPD and PG in literature have had a concomitant paraproteinemia and thus must be thoroughly worked up and carefully followed up.

### *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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Nil.

### *Conflicts of interest*

There are no conflicts of interest.

## References

1. Neutrophilic Dermatoses | Plastic Surgery Key [Internet]. [cited 2023 Feb. 7]. https://plasticsurgerykey.com/neutrophilic-derma toses-3/.
2. Watts PJ, Khachemoune A. Subcorneal pustular dermatosis: A review of 30 years of progress. Am J Clin Dermatol 2016;17:653-71.
3. Weedon, D. 6—The vesiculobullous reaction pattern. In Weedon's Skin Pathology Weedon, 3rd ed.; Churchill Livingstone: London, UK, 2010; p. 123-168.e54, ISBN 978-0-7020-3485-5.
4. Naik HB, Cowen EW. Autoinflammatory pustular neutrophilic diseases. Dermatol Clin 2013;31:405-25.
5. Cheng S, Edmonds E, Ben-Gashir M, Yu RC. Subcorneal pustular dermatosis: 50 years on. Clin Exp Dermatol 2008;33:229-33.
6. Ferrillo M, Villani A, Fabbrocini G, Mascolo M, Megna M, Costa C, *et al.* A Case of the Co-Existence of Subcorneal Pustular Dermatosis and Pyoderma Gangrenosum and a Review of the Literature. Open Access Maced J Med Sci 2018;6:1271-4.