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Case Report

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Sub corneal Pustular Dermatosis in a Middle-Aged Woman: A Rare Presentation with Diagnostic Challenges and Successful Topical Corticosteroid Therapy

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Abstract

Sub corneal pustular dermatosis (SPD), also known as Sneddon–Wilkinson disease, is a rare, chronic, relapsing neutrophilic dermatosis characterized by the presence of superficial pustules. It predominantly affects middle-aged to elderly women and commonly presents with flaccid pustules over the trunk and intertriginous areas. Due to its clinical overlap, SPD is often mistaken for other dermatologic conditions such as pustular psoriasis, impetigo, and IgA pemphigus. Immunologic studies separate SPD-type IgA pemphigus from SPD and pustular psoriasis. Dapsone remains the first-line treatment for SPD, although dapsone-resistant cases have been increasingly reported. Other therapies have been used singly or as adjunctive therapy with success, such as corticosteroids, immunosuppressive agents, tumor necrosis factor inhibitors, and ultraviolet light therapy. We present the case of a 48-year-old woman with recurrent annular plaques localized to the right forearm. Histopathologic examination confirmed a diagnosis of SPD. The condition was effectively managed with topical corticosteroids alone. This case underscores the diagnostic challenges associated with SPD and demonstrates the potential effectiveness of topical therapy in mild presentations.

Introduction

Sub corneal pustular dermatosis (SPD), also known as Sneddon–Wilkinson disease, is a rare, chronic, and relapsing neutrophilic skin disorder first described by Sneddon and Wilkinson in 1956. It primarily affects women between the ages of 40 and 60, with a reported female-to-male ratio of approximately 4:1 (Kouskoukis et al., 2000; Larsabal et al., 2015). Although the exact cause remains unknown, SPD is categorized as a neutrophilic dermatosis (James, Berger, and Elston, 2015). Clinically, it is characterized by sterile, superficial pustules that coalesce into annular or circinate plaques (Kouskoukis et al., 2000). However, there is still an unclear designation as to whether SPD is its own entity

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distinct from pustular psoriasis, as the once thought characteristic histologic picture of psoriasis does not hold true for pustular psoriasis.

SPD has been reported to occur in association with several neoplastic, immunologic, and inflammatory conditions.

Despite its benign nature, SPD is often persistent and prone to recurrence. Histopathological examination reveals subcorneal pustules filled predominantly with neutrophils and lacking significant acantholysis—a feature that helps differentiate it from conditions such as IgA pemphigus and pustular psoriasis (Schmidt and Zillikens, 2013). SPD may occur idiopathically or in association with systemic disorders, including monoclonal gammopathies, autoimmune diseases, and malignancies (Sais et al., 1995).

While dapsone remains the first-line treatment due to its anti-neutrophilic properties, other therapeutic options—such as corticosteroids, immunosuppressive agents, biologics, and phototherapy—have been employed in cases resistant to standard therapy (Hashimoto et al., 2018). The overall incidence of SPD is estimated to be fewer than 1 case per 100,000 population per year, though no large-scale epidemiological studies have yet established its global prevalence.

Case Report

A 48-year-old Sudanese woman, Haneen Magdi Ahmed Abdelkarim, presented to the dermatology outpatient clinic on 15 October 2025 with a complaint of an itchy, red, annular rash on her right forearm. The lesion appeared two days prior to presentation and progressively enlarged. A similar episode occurred two months earlier and resolved spontaneously.

The dermatological examination revealed well-defined annular erythematous papules and plaques localized to the right forearm. The lesions were non-scaling and non-vesicular. There was no fever or systemic involvement.

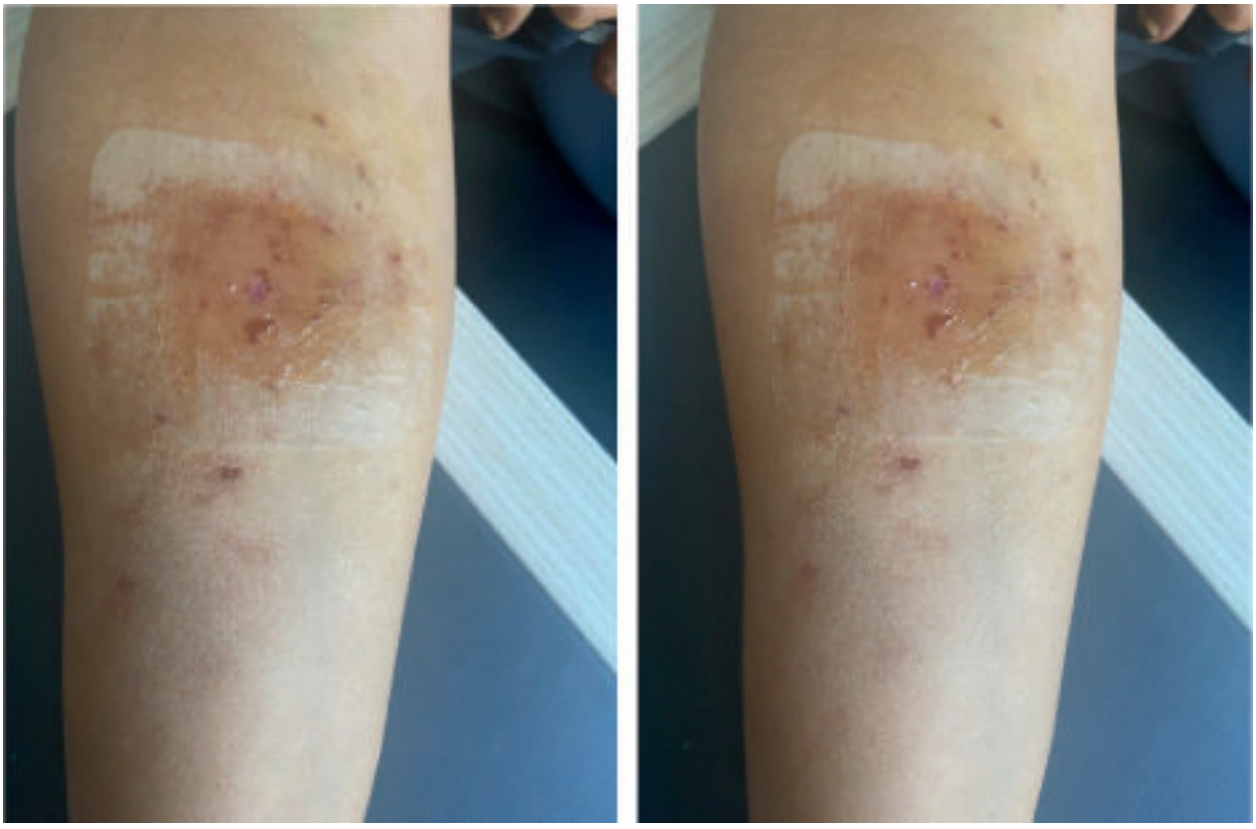
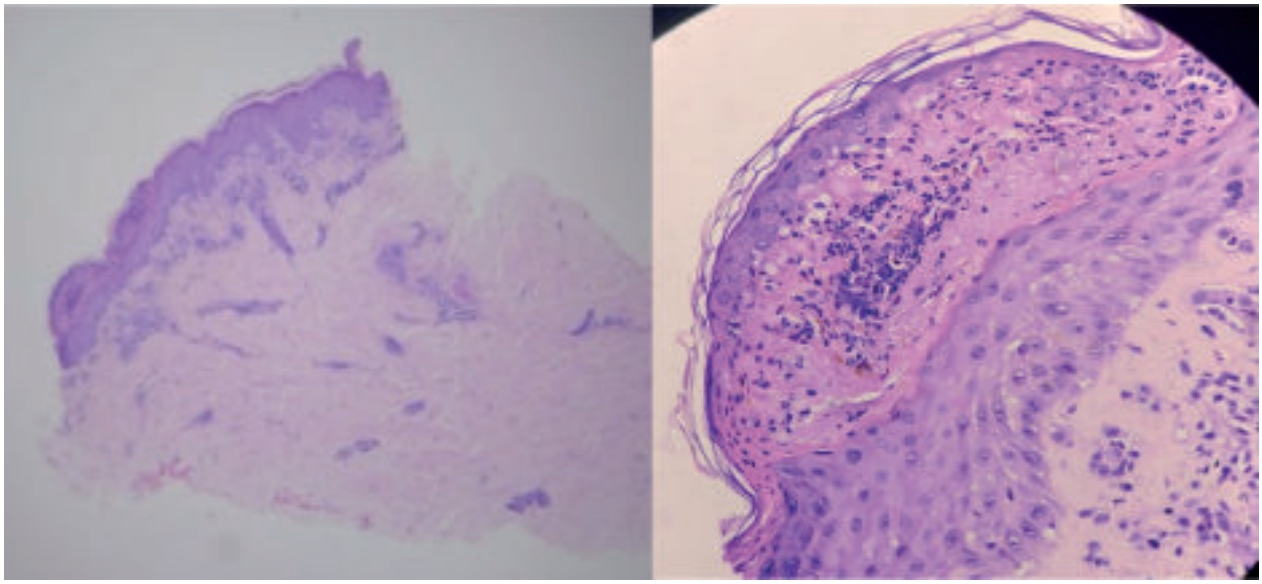


Figure 1: Low-power photomicrograph showing itchy red rash over right forearm associated with sub corneal pustules. Annular erythematous papules, plaques on right forearm

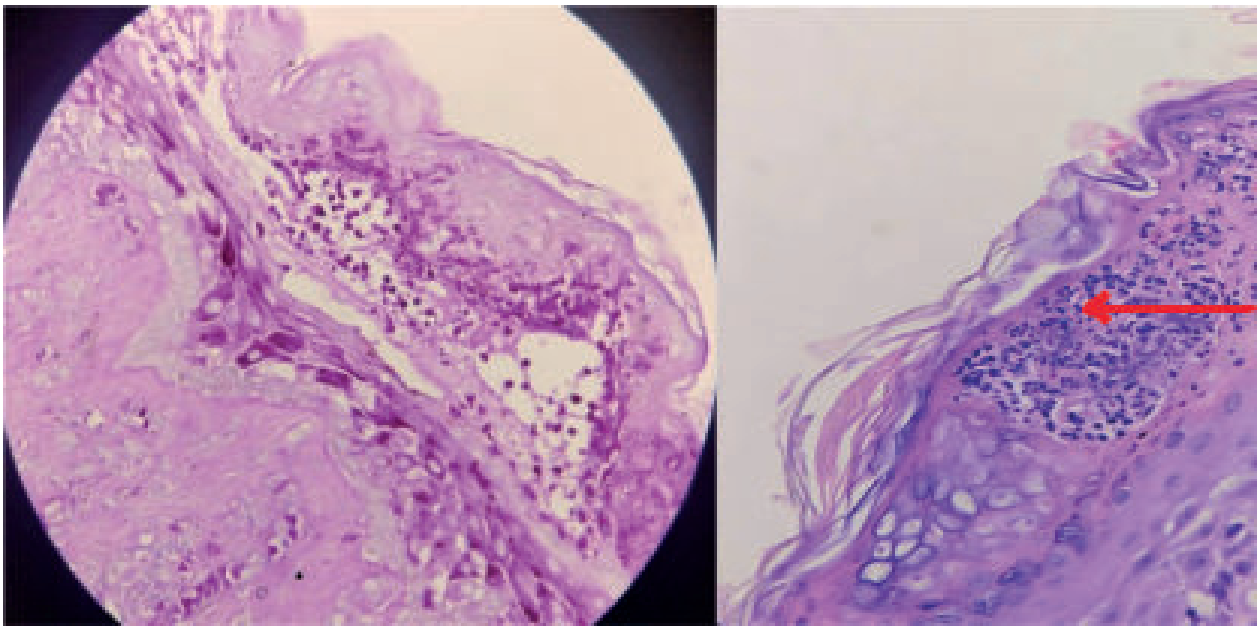
The patient was initially managed with *Resolve Plus* cream (combination of betamethasone and miconazole) by a general practitioner, without improvement. Differential diagnoses included: other specified, Psoriasis unspecified, Tinea incognito, granuloma annularae.

Histopathological Findings

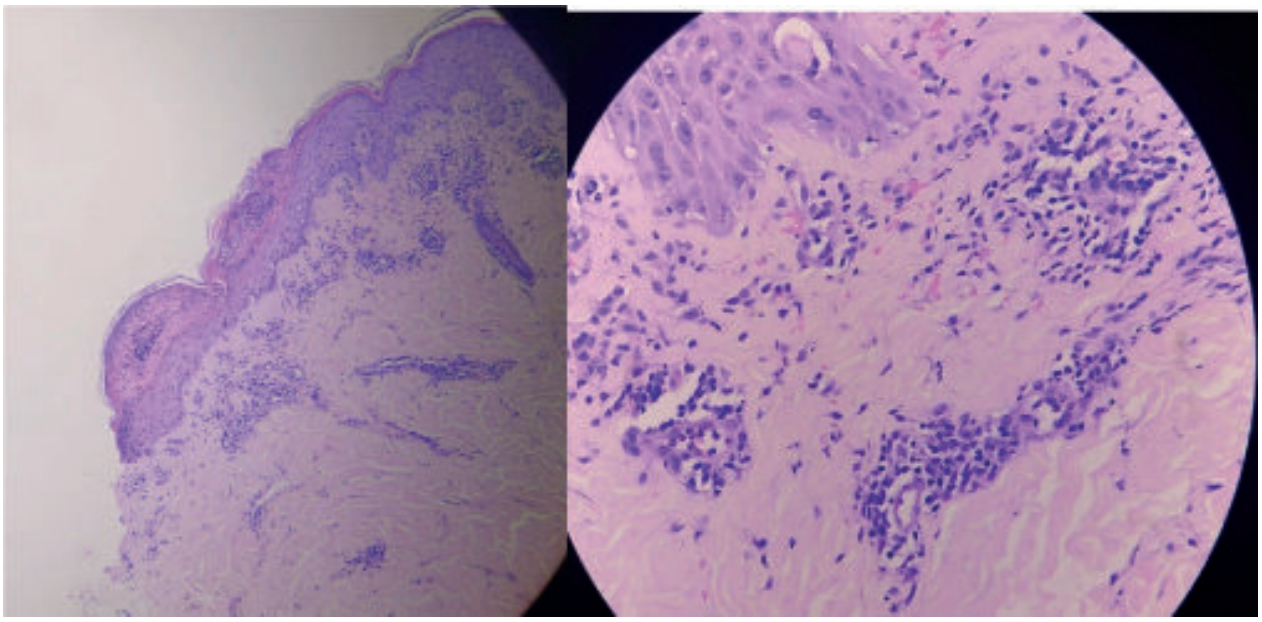
A punch biopsy was obtained on 21 February 2025 from an active lesion on the right forearm and sent for histopathological examination. Histopathology Lab received in formalin identified with patient's name and MRN is single skin tissue measuring 0.3 x 0.1 cm. Totally embedded in 1 block.



Figures 2: Microscopy at low power magnification. Sections show a sub corneal, superficial intraepidermal pustule containing plasma and neutrophils. No acantholytic granular keratinocytes noted or eosinophilic spongiosis in the adjacent intact epidermis. Superficial perivascular and interstitial inflammatory infiltrates composed predominantly of neutrophils with occasional eosinophils and lymphocytes.



Figures 3: Higher magnification reveal superficial perivascular and interstitial inflammatory infiltrates composed predominantly of neutrophils with occasional eosinophils and lymphocytes. PAS stain is negative for fungal organisms. Epidermis shows minimal acanthosis and spongiosis. Acantholysis is usually absent or very limited, helping differentiate from pemphigus foliaceus.



Figures 4: *Histopathological findings. Neutrophilic pustular dermatitis. Sub corneal and superficial intraepidermal pustules composed of neutrophils and plasma cells.*

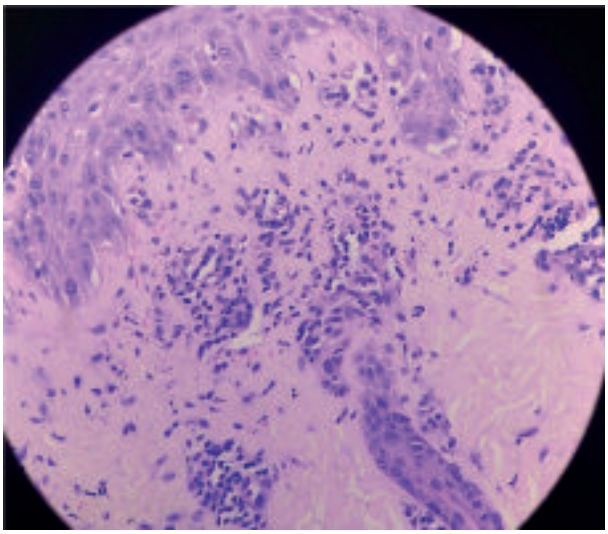


Figure 5: Histopathological findings at High power magnification. Dermis shows a mild superficial perivascular infiltrate, composed predominantly of lymphocytes and some neutrophils.

The pustules are located just beneath the stratum corneum hence the term "sub corneal" and typically lack significant acantholysis. Occasionally, a small number of eosinophils or mononuclear cells may be present (Weedon, 2010). The dermis often shows a superficial perivascular infiltrate composed mainly of neutrophils, with scattered eosinophils and lymphocytes. Notably, there is an absence of acantholytic keratinocytes or eosinophilic spongiosis. Negative for dermatophytosis.

Direct immunofluorescence (DIF) is typically negative for immunoglobulin and complement deposition, which aids in differentiating SPD from IgA pemphigus—a condition characterized by intercellular IgA deposits (Wallach, 1999). Periodic acid–Schiff (PAS) staining is also negative, excluding fungal infections as a cause.

Diagnosis

Based on clinical and histological correlation, a diagnosis of sub corneal pustular dermatosis (Sneddon–Wilkinson disease) was made. Dermatophytosis and impetigo were ruled out.

Differential Histopathologic Diagnoses

IgA pemphigus:

Histologically similar to SPD, showing sub corneal pustules; however, direct immunofluorescence (DIF) reveals intercellular IgA deposition, especially in the upper epidermis (Hashimoto et al., 1997).

Pemphigus foliaceus:

Demonstrates prominent acantholysis in the upper epidermis (sub corneal level) and positive DIF with intercellular IgG and C3 deposition in a "chicken wire" pattern (Amagai and Stanley, 2012).

Pustular psoriasis:
Characterized by spongiform pustules of Kogoj, Munro micro abscesses, parakeratosis, and regular acanthosis. It also shows neutrophilic infiltration within the epidermis and stratum corneum (Griffiths and Barker, 2007).

Treatment and Outcome
While dapsone therapy was planned, the patient showed marked improvement with **topical corticosteroids** alone. The lesions resolved without recurrence during initial follow-up. The patient was advised regarding the chronic relapsing nature of SPD and the need for regular dermatological follow-up.

Discussion
Sub corneal pustular dermatosis (SPD) presents both diagnostic and therapeutic challenges due to its rarity and clinical resemblance to other pustular dermatoses. Accurate diagnosis necessitates a thorough clinicopathological correlation. SPD is frequently misdiagnosed as conditions such as pustular psoriasis—which typically shows deeper dermal neutrophilic infiltrates and parakeratosis—or other entities like impetigo and IgA pemphigus (Wollina, 2009; Schmidt and Zillikens, 2013). This diagnostic ambiguity likely contributes to underreporting and the absence of robust epidemiological data.

While a few case series have documented associations between SPD and systemic conditions such as paraproteinemia or autoimmune diseases, these remain isolated observations without confirmation through population-level studies (Sais et al., 1995). Histologically, SPD is characterized by sub corneal pustules predominantly composed of neutrophils, with little to no acantholysis—features that help distinguish it from IgA pemphigus (Hashimoto et al., 2018).

In our case, the diagnosis of SPD was supported by the absence of fungal elements on PAS staining and a lack of epidermal acantholysis. Although dapsone remains the first-line treatment due to its anti-neutrophilic properties, our patient achieved satisfactory clinical improvement with topical corticosteroids alone. This response aligns with previous reports suggesting that mild cases of SPD may respond well to topical therapy, especially when systemic treatment is not advisable (Larsabal et al., 2015).

Furthermore, this case highlights the importance of evaluating patients for potential underlying associations such as monoclonal gammopathy or autoimmune disorders. However, no such comorbidities were identified in our patient at the time of diagnosis.

Conclusion:
Sub corneal pustular dermatosis (SPD) is an important but rare cause of annular pustular eruptions in adults. The defining histologic feature is a sub corneal, sterile pustule rich in neutrophils, without acantholysis or immunoglobulin deposition—findings that help distinguish SPD from other pustular and autoimmune bullous disorders.

While dapsone is traditionally the first-line therapy due to its anti-neutrophilic properties, topical corticosteroids may offer effective symptom control in mild or early-stage cases. Ongoing follow-up is critical to detect recurrence and screen for possible systemic associations such as autoimmune conditions or hematologic abnormalities.

Patient Consent:
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflicts of Interest:
The authors declare no conflicts of interest.

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