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A D R ISSN 2994-1180 ANNALS OF DERMATOLOGICAL RESEARCH

Case Report

# Subcorneal pustular dermatosis (Sneddon-Wilkinson disease): a case report

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## Abstract

This article contains modern data on the etiology, clinical presentation and treatment as well as the differential diagnosis of a rare dermatosis, Sneddon-Wilkinson disease.

Clinical observation of subcorneal pustular dermatosis in a 68-year-old woman is reported. Histopathological examination played a key role in making the correct diagnosis. Treatment consisted of a physiotherapeutic approach (311 nm UVB phototherapy) in combination with systemic glucocorticosteroids (dexamethasone). This therapeutic approach (instead of dapsone therapy) can be used due to its relatively good safety profile.

**More Information**

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**Submitted:** March 15, 2023
**Approved:** April 04, 2023
**Published:** April 05, 2023

**How to cite this article:** Potekaev NN, Sysoeva VS, Bobko SI, Katunina OR, Borodulina KS. Subcorneal pustular dermatosis (Sneddon-Wilkinson disease): a case report. Case reports. Ann Dermatol Res. 2023; 7: 013-015.

**DOI:** 10.29328/journal.adr.1001024

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**Keywords:** Subcorneal pustular dermatosis; Sneddon-Wilkinson disease; Path morphology; Dapsone; Phototherapy



## Introduction

Subcorneal pustular dermatosis or Sneddon-Wilkinson disease (SPD) is a rare chronic benign relapsing vesiculo-pustular disease of unknown ethiology, that most commonly occurs in middle-aged and elderly women. At the same time, rare cases of the development of SPD in children are described in the literature [1].

Although the cause of occurrence remains unknown, the role of previous or concomitant infections cannot be ruled out [2]. Endocrine disorders, mental disturbances, and immunoallergic diseases have a certain value in the occurrence of the disease [3]. In addition, there is a number of diseases associated with subcorneal pustulosis, such as monoclonal gammopathy, multiple myeloma, rheumatoid arthritis, Crohn's disease, pyoderma gangrenosum, Sweet syndrome, Crocker-Williams persistently elevated erythema, pustular psoriasis, SAPHO syndrome (synovitis, acne, gangrenous pyoderma, osteitis). There are reports on the relationship of SPD with aplastic anemia, mycoplasmal pneumonia, cystic lesions of the liver in some helminthiasis, hyperthyroidism, and thymoma [4,5]. Pregnancy can be also a precipitating factor [6].

On normal or more often slightly erythematous skin primary focus of the disease appear in groups – small (up to 5 mm in diameter). Superficial pustules with a flabby tire contain neutrophils. The lesions are prone to confluence and peripheral growth can form annular, serpiginous patterns. After opening they dry out and form superficial scale crusts resembling impetigo. In their place, slight hyperpigmentation may remain. Lesions are usually symmetrical, located mainly in large folds (axillary, inguinalfenoral), in the abdomen, under the mammary glands, and on the extensor areas of the limbs. Rarely, the face, palms, and soles are affected. Mucous membranes and skin of the scalp are always free from lesions. Subjectively patients may complain of a feeling of itching and burning, while the general condition remains satisfactory. Clinical and biochemical blood tests – no pathology [2,7]. Although the protein electrophoresis is carried out abroad because of the association of paraproteinemia with SPD [3] According to G. Baeza-Hernández, 2021, the etiological factor Mycoplasma Pneumoniae was discussed [8].

Histologically the main symptom of the disease is subcorneal pustules with polymorphonuclear leukocytes. The

epidermal layer under the pustule has practical changes. In the dermis, a perivascular infiltrate containing neutrophils, single monocytes, and eosinophils is detected. Acantholysis is absent [2].

In a group of patients during a direct immunofluorescence reaction (RIF) sometimes deposits of IgA are found in the epidermis. In isolated cases, the method of indirect FIR reveals circulating antibodies to IgA directed against epidermal cells. According to the modern classification, such cases are described as the SPD type of pemphigus [9].

Differential diagnosis is presented in the (Table 1).

Without treatment periods of exacerbation repeat for several years, while the duration of remissions varies from several days to several weeks. The drugs of choice are sulfones (predominantly dapsone) [10], in the treatment of which most often a complete remission is achieved. Acitretinmaybeasecond-line drug. The literature describes isolated cases of successful use of drugs such as colchicine, cyclosporine, infliximab, etanercept, tumor necrosis factor alpha blockers, maxacalcitol, mizoribine, ketoconazole, tetracycline, minocycline, benzylpenicillin, vitamin E, azithromycin [11], doxycycline [8,12], adalimumab [13]. The efficacy of systemic and topical glucocorticosteroids in elderly patients has also been reported [14,15].

## Case report

Patient G., born in 1954, came to an outpatient appointment with complaints of lesions of the skin of the trunk, and upper and lower extremities, accompanied by periodic itching of moderate intensity. She was ill since 1988 when she first noted the appearance of lesions on the skin of the trunk after a seven-day fast. She independently used sulfuric ointment, according to the words, a skin biopsy was performed, but the diagnosis was not established (medical documents were not provided). Further notes of the undulating course of the process with periods of exacerbations and remissions, improvements against the background of insolation. Since 1992 she noted the spread of the process, hospitalization was carried out in a hospital, where the differential diagnosis was carried out between the diagnoses of Duhring dermatitis herpetiform and Subcorneal pustulosis, received treatment with a suspension for the external use of cindol and plasmapheresis. She was discharged with improvement. After suffering stress in 2017-2018 she noted another exacerbation of the process, phototherapy (UVB 311 nm) was carried out with a positive effect.

In 2022 due to another exacerbation, hospitalization in a daily stay hospital of the branch Clinic named after V.G. Korolenko for pathomorphological examination and in case of confirmation of the diagnosis of Sneddon-Wilkinson's subcorneal pustulosis, the start of dapsone therapy. During the additional examination, she underwent a course of phototherapy with UVB 311 nm (14 sessions) with a temporary positive effect. She also received 5 injections of Hemodez solution, received Phosphogliv, Mexidol, dexamethasone solution 4 and 8 mg intramuscularly according to the recommendation of a polyclinic doctor - with a temporary positive effect, and the course of phototherapy was interrupted. External treatment was not carried out. She refused to take dapsone due to the improvement of the skin process.

Concomitant diseases: duodenal ulcer, nodular goiter. Bad habits are denied. Allergy anamnesis: allergic reaction to novocaine. Heredity: parents – diabetes mellitus, father – Parkinson's disease, brother – cancer of the kidney, thyroid cancer. Sexually transmitted infections, HIV, viral hepatitis, and tuberculosis denies. No blood transfusions. The patient fulfilled the special information form that allowed them to perform all the necessary consultations and examinations.

### Objectively. The general condition is satisfactory

**Local status:** The pathological process on the skin is subacute, widespread, localized on the skin of the trunk, and symmetrically on the skin of the upper and lower extremities. Represented by papules, pustules with a flaccid tire, surrounded by an inflammatory corolla along the periphery, up to 0.5 cm in size, red, Nikolsky's symptom is negative. There are no dermoscopic findings for atypia. Mucous membranes, the skin of the scalp, palms, and soles are intact (Figure 1a-d).

**Table 1:** Differential diagnosis of SPD.

| Disease | Distinctive features |
|---|---|
| Streptococcal impetigo | Bacteria in pustules, the efficacy of topical and systemic antibiotic therapy |
| Duhring dermatitis herpetiformis | Severe pruritus, predominantly extensor areas, subepidermal IgA deposition in the basement membrane, and the presence of eosinophilic abscesses in the papillary dermis |
| Pemphigus foliaceus | Acantholysis IgG deposition in the upper parts of the spinous layer, acantholytic cells, positive Nilolsky's sign |
| Zumbusch generalized pustular psoriasis | High temperature, malaise, leukocytosis, spongiform Kogoi pustules in the spinous layer |
| Necrolytic migratory erythema (glucagonoma syndrome) | Excessive levels of glucagon in the biochemical analysis of blood the absence of forming pustules, erosion on the lip and mucous membranes of the oral cavity, and necrobiosis of the upper part of the epidermis. |
| Acute generalized exanthematous pustulosis | Lesions are located throughout the body, mainly on the distal parts of the extremities, leukocytoclastic vasculitis in histological examination |

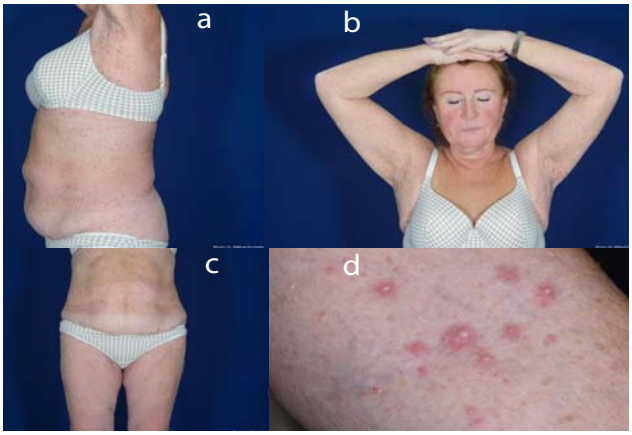


**Figure 1:** Rashes on the skin of the chest (1a), abdomen areas (1c), and arms (1b,d).

**Data from laboratory examinations:** Complete blood counts blood sedimentation rate 63 mm/h. Biochemical blood test: glucose 5.86, Rapid plasma reagin - negative, HBsAg negative, D-dimer more than 5000, glycosylated hemoglobin 6.2 Histological examination of a skin biopsy sample mild acanthosis of the epidermis with exocytosis of neutrophilic granulocytes into the epidermis, the presence of *subcorneal pustules containing neutrophilic leukocytes*, in some places single foci of spongiosis, single acantholytic cells are found. Around the vessels of the superficial plexus, there is a moderate lymphomonocytic infiltration with an admixture of neutrophilic granulocytes, leukocyte stasis is present in the lumen of the vessels (Figure 2a,b).

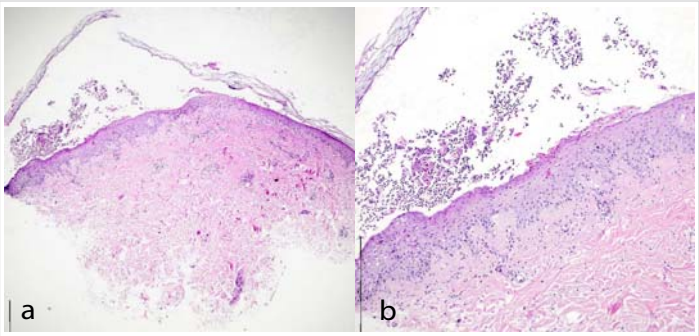


**Figure 2:** The pathomorphological pattern of the skin biopsy specimen.

**Conclusion:** Pathomorphological changes can be observed in Sneddon-Wilkinson subcorneal pustulosis, clinically differentiated from superficial pyoderma (bullous impetigo). Based on the clinical picture and histological examination data, the diagnosis was made Sneddon-Wilkinson subcorneal dermatosis.

H&E staining with different magnifications.

## Discussion and conclusion

The presented clinical case has high interest from the point of view of the complexity of the differential diagnosis of the rare Sneddon-Wilkinson dermatosis. To prescribe adequate therapy, only a pathomorphological study will help dermtovenereologists to confirm the correct diagnosis. In addition, this case demonstrates the effectiveness of physiotherapy (311 nm UVB phototherapy) in combination with systemic glucocorticosteroids (dexamethasone).

This approach may be advantageous in patients with contraindications to dapsone therapy or with severe comorbidities in the treatment of subcorneal pustulosis due to a lower likelihood of side effects.

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