SUBCORNEAL PUSTULAR DERMATOSIS IN A 47-YEAR-OLD WOMAN

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Abstract: Subcorneal pustular dermatosis (SPD) is a pustular eruption which is rare, chronic, and recurrent. This condition is commonly found in women over the age of 40 years with unknown etiology. It is characterized with symmetrical pustules-vesicles that quickly develop to pustules on erythematous skin with peripherally spreading. The pustules undergo central healing leaving polycyclic, erythematous areas in which new pustules arise. The lesions typically involve the intertriginous areas, trunks, and flexor of limbs. We reported a case of a 47-year-old woman with blisters and pustules all of the body surface since a year ago. The blisters ruptured easily and became erosions. The dermatological status indicated generalized erythematous hyperpigmented macules which were multiple, discrete, lenticular-plaques in size, multiple vesicles with pustules on it, and ruptured pustules leaving erosions as well as crusted areas. Anamnesis, physical examination, laboratory examinations, and a skin biopsy was performed. The Gram staining of the blister showed only PMN leukocytes. Normal CBC, eosinophil 1.9%, lymphocyte 24.3%, elevated ESR 70 mm/hour, total IgE 831.000 IU/mL. The histopathological examination indicated spongiosis, subcorneal blister consisting of fibrin, neutrophils, and few eosinophils in the epidermis. The dermis revealed superficial perivascular inflammatory infiltration (neutrophils, lymphocytes, some eosinophils). These findings were typical for SPD. Conclusion: This case was diagnosed as subcorneal pustular dermatosis based on anamnesis, physical examination, and a histopathological examination.

Keywords: subcorneal pustular dermatosis, histopathological examination
Subcorneal pustular dermatosis (SPD), also known as Sneddon–Wilkinson disease, is an uncommon, chronic, relapsing pustular eruption with histopathologically characterized by subcorneal pustule that contains neutrophils. This disease was first described in 1956 by Sneddon and Wilkinson. Its etiology remains unknown. Typically, the patient has a history of a relapsing symmetrical sterile pustular eruption involving the intertriginous areas, trunks, and flexor aspects of the limbs. Scalp is almost never affected, nor are the mucous membranes, palms, and soles.

There is no racial predilection so far, however, many cases have been reported among the whites, Africans, Japanese, and Chinese. This disease is more frequently in females than in males (4:1), especially in the age group of 40 years or more. This disease occurs more rarely during childhood.

We reported a case of subcorneal pustular dermatosis in a 47-year-old woman. This was the first case reported from the Dermatovenereology Department of Prof. Dr. R. D. Kandou Hospital Manado.

**CASE REPORT**

A 47-year-old woman was admitted to the Dermatovenereology Department in May 5th, 2013 with a history of relapsing pustular eruption involving the trunk, flexoral proximal extremities, and groin since a year ago. She noted that the onset was on her axillary areas and then the eruption spread to the inframammary folds, sometimes with mild pruritus, but there were no other associated symptoms. The patient reported that some blisters had arisen within a few hours, ruptured in a few days, and left erosions covered by thin crust. She had a history of atopic dermatitis, while her mother suffered from asthma. There was no history of drug allergy.

Physical examination revealed multiple pustules that tended to coalesce forming circinate or serpiginous pattern, and flaccid bullae filled with clear fluid on the erythematous skin of the trunk, groin, as well as on the upper and lower extremities. Within few days the pustules ruptured easily leaving erosions and superficial crust. The eruption spreaded peripherally with central healing. Healed lesions presented residual hyperpigmentation without any trophic alteration. New pustules arised across the previously affected areas (Figure 1).

The laboratory examination showed normal complete blood count, eosinophil count 1.9%, lymphocyte count 24.3%, elevated erythrocyte sedimentation rate 70 mm/hour, and elevated total IgE 831,000 IU/mL. Blood ureum, serum creatinine, liver function test, fasting blood sugar, and HbA1c were within normal limits. The differential diagnoses of this patient were subcorneal pustular dermatosis and pemphigus foliaceus. A punch biopsy of a lesion on the upper arm was carried out.

The histopathological examination showed spongiosis, subcorneal blister consisting of fibrin, neutrophils, and few eosinophils in the epidermis. The dermis revealed superficial perivascular inflammatory infiltration consisting of neutrophils, lymphocytes, and some eosinophils (Figure 2). The Gram staining of the blister revealed PMNs, but none of positive/negative Gram cocci, spores, or budding cells. Moreover, the Giemsa staining showed only PMNs.

**DISCUSSION**

Subcorneal pustular dermatosis is a benign inflammatory skin disease and characterized by remission and exacerbation that may last for 5-8 years. This disease is classified as neutrophilic dermatoses.

The dermatological feature of SPD is a small flaccid pustule or vesicle that becomes pustular and forms crops over a few hours either on normal or erythematous skin. The pustules are superficial and often merge, forming annular, circinate, or serpiginous patterns. The eruption often
spread peripherally with central healing and fades leaving post inflammatory pigmen-
tary changes without atrophy or scar.1,2,8 The interval between flare-ups alternating
with quiescent phases varies from a few
days to several weeks. It has a tendency to
locate in the flexural areas of the trunk and
limbs, and rarely on the palms and soles, albeit, it never affect the face and scalp.
There is no systemic symptom.1,2

The pathogenesis of this condition is
still obscure1,2,5 and no infectious agents
have been identified.1,6 Immunological
mechanisms have also been implicated.9
Several diseases have been described in
association with SPD, including IgA
paraproteinemia (up tp 40% cases)3 and
myeloproliferative disorders especially
multiple myeloma.1,3,10 The onset of this
disease may occur after years, therefore, prolonged follow-up is needed.11,12

Other various systemic disorders that
can be found with SPD were pyoderma
gangrenosum, ulcerative colitis, Crohn’s
disease, Ig G paraproteinemia, CD30+
anaplastic large–cell lymphoma, non small
cell lung cancer, apudoma, rheumatoid
arthritis, hyperthyroidism, and mycoplasma
pneumonia infection. Further associated
reported cases are multiple sclerosis,
Sjögren syndrome, chronic lymphocytic
leukemia, bullous pemphigoid, morphea,
and marginal zone lymphoma.1,3,10 None of
the previously known associations was
present in this patient.

The histopathological examination
reveals subcorneal pustule with neutrophils
and few eosinophils. The epidermis
beneath the pustule shows minimal changes
and perhaps slight intracellular edema.3,11
Older lesions may contains acantholytic
cells and superficial blood vessels with
non-specific mixed inflammatory cell
infiltration consisting of polymorph
neutrophils and mononuclear cells in the
dermis.3 In this patient, the histo-
pathological findings showed spongiosis
and subcorneal blister consisting of fibrin,
neutrophils and few eosinophils in the
epidermis. The dermis revealed superficial
perivasular inflammatory infiltration
consisting of neutrophils, lymphocytes, and
some eosinophilis. These findings were
consistent with SPD.

Direct immunofluorescent (DIF)
studies in SPD are negative. There were
some reported cases with clinical features
resembled SPD and intraepidermal deposit
of Ig A. The target of these IgA
autoantibodies was desmocollin 11,10 and
this disease was classified as SPD type IgA
pemphigus. However, we did not perform
immunoflorescence.

Sulfones are the primary treatment for
SPD. Dapson is the most effective in dose
of 50-150 mg daily.1 Although dapson has
a slower response than in dermatitis
herpetiformis, it leads usually in complete
remission.11,12 Some patients have
responded better to sulfapyridine therapy
(1-3 g daily). Systemic corticosteroids are
less effective but they have been used
successfully. Additional treatments include
retinoids (etretinate or acitretin) and PUVA
or narrow band UVB. Colchicine and
minocycline have also been useful in
anecdotal reports. The use of
immunobiological drugs such as infliximab
and etanercept have been reported with
good result.13-15 This patient was treated
with oral diamindophenylsulfone (dapson)
100 mg daily and improved fairly.

SPD is a benign condition but it may
recur over many years with remissions
varying over many years from days to
months. The general condition of the
patient is not affected unless there is an
associated immune dysfunction disorder.

CONCLUSION

We reported a case of a 47-year-old
woman with blisters and pustules all of the
body surface since a year ago. The
dermatological status indicated generalized
erythematous hyperpigmented macules
which were multiple, discrete, lenticular-
plaques in size, multiple vesicles with
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logical examination indicated spongiosis, subcorneal blister consisting of fibrin, neutrophils, and few eosinophils in the epidermis. The dermis revealed superficial perivascular inflammatory infiltration (neutrophils, lymphocytes, some eosinophils). These findings were typical for SPD. The patient was treated with oral diaminodiphenylsulfone (dapson) 100 mg daily and improved fairly.

**Figure 1.** Multiple pustules that tended to coalesce forming a circinate or serpiginous pattern, flaccid bullae filled with clear fluid on the erythematous skin of the trunk, groin and upper and lower extremities. Pustules ruptured easily in a few days leaving erosions and superficial crust.

**Figure 2.** Histopathological images of subcorneal pustules. A, Subcorneal pustule below the stratum corneum (arrow) (HE, 4x). B, Subcorneal pustule (arrow) (HE, 10x). C, Many neutrophils (arrow) below the stratum corneum (HE, 40x).

**REFERENCES**


