Case Report

Subcorneal Pustular Dermatosis: A Case Report of a Patient with Diffuse Scleroderma

Abstract

Subcorneal pustular dermatosis (SPD) or Sneddon-Wilkinson disease is a rare, benign, chronic, sterile pustular eruption which is associated with various systemic diseases including immunoglobinopathies, neoplasms, and autoimmune disorders. This paper reports a case of SPD in a patient with diffuse scleroderma in a 37-year-old woman. The hypothesis that immune dysregulation may play a role in the pathogenesis of SPD was supposed by the coexistence of diffuse scleroderma and SPD in our patient.

Keywords: *Diffuse scleroderma, immune dysregulation, subcorneal pustular dermatosis*

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Introduction

Subcorneal pustular dermatosis (SPD) or Sneddon-Wilkinson is a rare, benign, chronic, sterile pustular eruption which is common in middle-aged or elderly women. It was originally described in 1956.[1] SPD is associated with various systemic diseases including immunoglobinopathies, neoplasms, and autoimmune disorders.[2-5] This paper reports a case of SPD in a patient with diffuse scleroderma in a 37-year-old woman.

Case Report

A 37-year-old female patient was admitted to our hospital with a 2-week history of recurrent generalized pruritic pustular eruption located mainly on the trunk and the extremities. No diagnosis and treatment procedures were made for the patient before her first visit in our unit. Her past medical history was notable for the presence of diffuse scleroderma for the past 7 years. Her medications included atenolol, valsartan, hydroxychloroquine, furosemide, and nifedipine. The dermatologic examination showed superficial vesicles and pustules located on normal skin or erythematous base of the trunk and the extremities. Gravity-induced demarcation could hardly be seen. The face, palms, soles, and mucous membranes were spared [Figures 1-3]. No lymphadenopathy or hepatosplenomegaly was presented. Physical examination showed a "bird-like" face with a beaked nose, telangiectasia, and radial furrowing around the lips. The fingers had a smooth, shiny, tapered appearance with the nails curving over the atrophic phalanges.

The differential diagnosis included SPD, IgA-pemphigus, pustular psoriasis, and tinea. Because there was no history of exposure to a new drug, acute generalized exanthematous pustulosis (AGEP) was not considered in differential diagnosis. Cultures of the pustules were sterile. Biopsies were taken for light microscopy and direct immunofluorescence examinations. Microscopic examinations demonstrated acanthosis, focal parakeratosis, subcorneal pustules, and crust in the epidermis. A few acantholytic cells were identified. Focal vacuolar degeneration of the basal layer, infiltration of lymphocytes, and a few eosinophils accompanied by focal edema of papillary dermis were observed [Figure 4]. Direct immunofluorescence examination was negative. The diagnosis of SPD of Sneddon and Wilkinson was made based on the clinical and histopathological findings.

Results of laboratory examinations including complete blood count, serum chemistries, serum protein electrophoresis, urine protein electrophoresis, and glucose-6-phosphate dehydrogenase (G6PD) were normal.

When the normal level of G6PD was confirmed, our patient was started on dapsone at a dosage of 50 mg daily, topical

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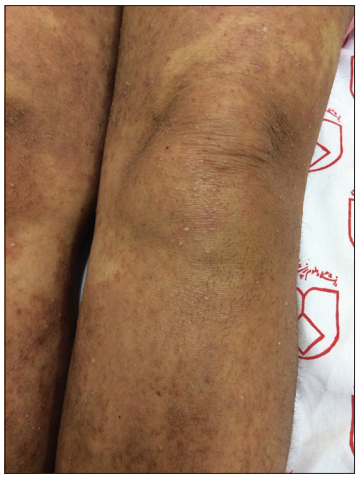

Figure 1: Patient's lesions 1

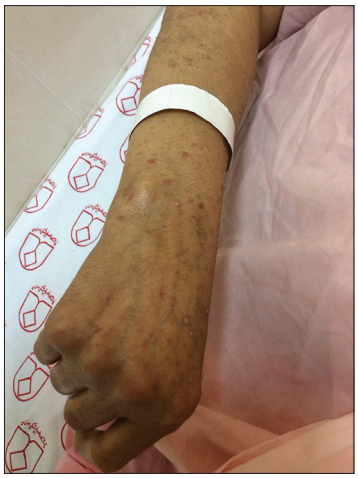

Figure 2: Patient's lesions 2

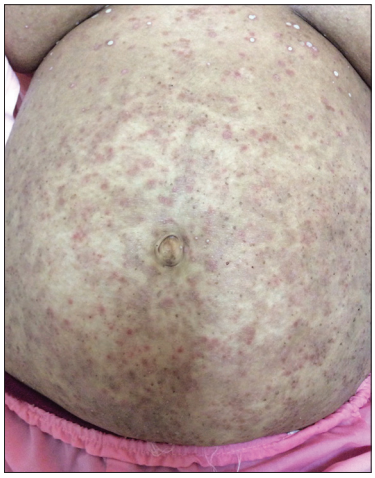

Figure 3: Patient's lesions 3

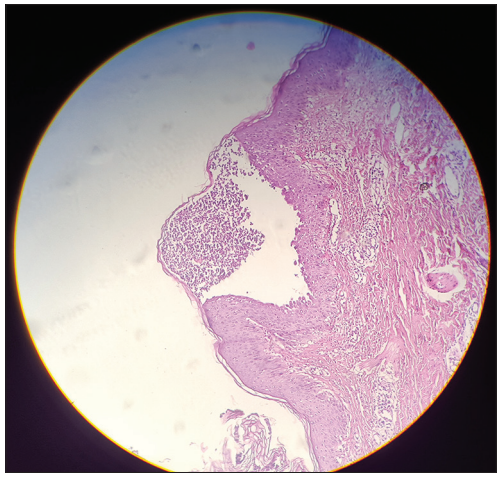

Figure 4: Histopathology

steroid twice daily, Burow's solution every 8 h, and oral hydroxyzine 25 mg daily. She refused to take dapsone and continued on topical medications for 2 weeks. Although the patient initially responded to topical therapy, the disease recurred two times during a 3-month follow-up. Dapsone was prescribed for the patient again, but she refused to take the drug and did not refer to our hospital.

Discussion

SPD or Sneddon-Wilkinson disease was originally described in 1956.[1] This rare, benign, chronic, sterile pustular eruption usually develops in middle-aged or elderly women.[2-5] The lesions coalesce into annular, circinate, or serpiginous patterns and involving more frequently the trunk, intertriginous areas, and flexor aspects of the limbs. The face, palms, soles and mucous membranes are usually unaffected in this disorder.[4,5]

The differential diagnosis includes pustular psoriasis, subcorneal-type IgA pemphigus, pemphigus foliaceus, dermatitis herpetiformis, dermatophyte infection, and acute AGEP. Moreover, some laboratory tests such as histopathological and immunofluorescence assays, culture of the pustules, and recent drug history are needed to rule out other diagnosis.[2,4,5]

SPD presents alone or with various systemic diseases including immunoglobinopathies, neoplasms, and autoimmune disorders such as benign monoclonal IgA, IgG, and IgM gammopathy, multiple myeloma, marginal zone lymphoma, rheumatoid arthritis, seronegative polyarthritis, Sjögren disease, and systemic lupus erythematosus.[2-9]

Diffuse scleroderma is a multisystem disease manifested by fibrosis, vasculopathy, and disordered immune system.[2,10-12] The hypothesis that immune dysregulation may play a role in the pathogenesis of SPD is supported by the coexistence of diffuse scleroderma and SPD in our patient. SPD is more frequent among women aged 40 years or older;[3,5] however, in our patient, it developed under the age of 40. The appearance of the disorder in our patient before her forties may be associated with the coexistence of SPD with diffuse scleroderma. A case report by Brantley and Sheth described a 37-year-old female patient with SPD who had past medical history of rheumatoid arthritis and

diffuse scleroderma.[2] In the study, underlying systemic immunologic defect was also noted as an important factor in the coexistence of SPD with rheumatoid arthritis and diffuse scleroderma. The patient was treated with dapsone and was stable after 9-month follow-up. The recurrence of disease in our patient with topical therapy suggesting the importance of dapsone in the treatment of SPD.

Conclusion

SPD may associate with underlying diffuse scleroderma and occur in the lower age in association with connective tissue diseases.

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.

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