

Subcorneal Pustular Dermatitis: A case report of a patient with diffuse scleroderma

Islam N¹, Shakil MIH²

Abstract

Subcorneal pustular dermatosis (SPD) or Sneddon-Wilkinson infection is an uncommon, harmless, constant, sterile pustular ejection which is related with different foundational sicknesses including immunoglobulinopathies, neoplasms, and immune system issues. This paper reports an instance of SPD in a patient with diffuse scleroderma in a 37-year-elderly person. The speculation that invulnerable dysregulation might assume a part in the pathogenesis of SPD was assumed by the concurrence of diffuse scleroderma and SPD in our patient.

CBMJ 2021 January: vol. 10 no. 01 P: 50-53

Key words: Diffuse scleroderma, resistant dysregulation, subcorneal pustular dermatosis

Introduction

Subcorneal pustular dermatosis (SPD) or Sneddon-Wilkinson is an uncommon, harmless, persistent, sterile pustular ejection which is normal in moderately aged or old ladies. It was initially portrayed in 1956.¹ SPD is related with different fundamental infections including immunoglobulinopathies, neoplasms, and immune system disorders.²⁻⁵ This paper reports an instance of SPD in a patient with diffuse scleroderma in a 37-year-elderly person.

Case Report

A 37-year-old female patient was conceded to our medical clinic with a 2-week history of repetitive summed up pruritic pustular ejection found primarily on the storage compartment and the limits. No determination and treatment techniques were made for the patient before her first visit in our unit. Her previous clinical history was remarkable for the presence of diffuse scleroderma for the beyond 7 years. Her drugs included atenolol, valsartan, hydroxychloroquine, furosemide, and nifedipine. The dermatologic assessment showed shallow vesicles and pustules situated on typical skin or erythematous base of the storage compartment and the limits. Gravity actuated outline could barely be seen. The face, palms, soles, and mucous layers were saved [Figures1-3].

No lymphadenopathy or hepatosplenomegaly was introduced. Actual assessment showed a "bird-like" face with a curved nose, telangiectasia, and spiral wrinkling around the lips. The fingers had a smooth, gleaming, tightened appearance with the nails bending over the atrophic phalanges.

Figure 1: Patient's lesions 1



1. Dr. Nahida Islam, Associate Professor, Head of Department, Department of Dermatology and Venereology, Community Based Medical College & Hospital, Bangladesh.
2. Dr. Mohammad Imdadul Hoque Shakil, Orthopedic Consultant, Department of Orthopedic, UHC sadar, Mymensingh, Bangladesh.

Address of correspondence:

Email: nislamcb5@gmail.com

Figure 2 : Patient's lesions 2**Figure 3:** Patient's lesions 3**Figure 4:** Patient's lesions 4

The differential determination included SPD, IgA-pemphigus, pustular psoriasis, and fungus. Since there was no set of experiences of openness to another medication, intense summed up exanthematous pustulosis (AGEP) was not considered in differential determination. Societies of the pustules were sterile. Biopsies were taken for light microscopy and direct immunofluorescence assessments. Minuscule assessments exhibited acanthosis, central parakeratosis, subcorneal pustules, and covering in the epidermis. A couple acantholytic cells were recognized. Central vacuolar degeneration of the basal layer, penetration of lymphocytes, and a couple of eosinophils joined by central edema of papillary dermis were noticed [Figure 4]. Direct immunofluorescence assessment was negative. The determination of SPD of Sneddon and Wilkinson was made dependent on the clinical and histopathological discoveries.

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 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 infection was initially depicted in 1956.¹ This uncommon, harmless, constant, sterile pustular ejection typically creates in moderately aged or old women.²⁻⁵ The sores mix into annular, circinate, or serpiginous designs and including all the more oftentimes the storage compartment, intertriginous regions, and flexor parts of the appendages. The face, palms, soles and mucous layers are generally unaffected in this disorder.^{4,5}

The differential conclusion incorporates pustular psoriasis, subcorneal-type IgA pemphigus, pemphigus foliaceus, dermatitis herpetiformis, dermatophyte disease, and intense AGEP. Additionally, some research center tests, for example, histopathological and immunofluorescence measures, culture of the pustules, and late medication history are expected to preclude other diagnosis.^{2,4,5}

SPD presents alone or with different fundamental infections including immunoglobulopathies, neoplasms, and immune system problems like harmless monoclonal IgA, IgG, and IgM gammopathy, numerous myeloma, minor zone lymphoma, rheumatoid joint inflammation, seronegative polyarthritis, Sjögren illness, and foundational lupus erythematosus.²⁻⁹

Diffuse scleroderma is a multisystem illness showed by fibrosis, vasculopathy, and cluttered resistant system.^{2,10-12} The theory that insusceptible dysregulation might assume a part in the pathogenesis of SPD is upheld by the concurrence of diffuse scleroderma and SPD in our patient. SPD is more successive among ladies matured 40 years or older;^{3,5} be that as it may, in our patient, it created younger than 40. The presence of the problem in our patient before her forties might be related with the concurrence of SPD with diffuse scleroderma. A case report by Brantley and Sheth described a 37-year-old female patient with SPD who had past clinical history of rheumatoid joint pain and diffuse scleroderma.² In the review, hidden fundamental immunologic deformity was additionally noted as a significant factor in the conjunction of SPD with rheumatoid joint pain and diffuse scleroderma. The patient was treated with dapson and was steady following 9-month follow-up. The repeat of illness in our patient with skin treatment proposing the significance of dapson in the treatment of SPD.

Conclusion

SPD might connect with basic diffuse scleroderma and happen in the lower age in relationship with connective tissue infections.

Affirmation of patient assent

The creators confirm that they have acquired all proper patient assent structures. In the structure the patient(s) has/have given his/her/their assent for his/her/their pictures and other clinical data to be accounted for in the diary. The patients comprehend that their names and initials won't be distributed and due endeavors will be made to disguise their character, yet obscurity can't be ensured.

Monetary help and sponsorship

Nil.

Irreconcilable circumstances

There are no irreconcilable circumstances.

References:

1. Sneddon IB, Wilkinson DS. Subcorneal pustular dermatosis. *Br J Dermatol.* 1956;68:385–94. [PubMed] [Google Scholar]
2. Brantley EI, Sheth P. Subcorneal pustular dermatosis in a patient with rheumatoid arthritis and diffuse scleroderma. *Dermatol Online J.* 2009;15:5. [PubMed] [Google Scholar]
3. Abreu Velez AM, Smith JG, Jr, Howard MS. Subcorneal pustular dermatosis an immunohistopathological perspective. *Int J Clin Exp Pathol.* 2011;4:526–9. [PMC free article] [PubMed] [Google Scholar]
4. Ratnarathorn M, Newman J. Subcorneal pustular dermatosis (Sneddon-Wilkinson disease) occurring in association with nodal marginal zone lymphoma: A case report. *Dermatol Online J.* 2008;14:6. [PubMed] [Google Scholar]
5. Scalvenzi M, Palmisano F, Annunziata MC, Mezza E, Cozzolino I, Costa C. Subcorneal pustular dermatosis in childhood: A case report and review of the literature. *Case Rep Dermatol Med.* 2013;2013:424797. [PMC free article] [PubMed] [Google Scholar]

6. Cartier H, Plantin P, Leroy JP, Larzul JJ. *Pyoderma gangrenosum, subcorneal IgA pustulosis and recurrent neutrophilic pleural and pulmonary diseases in a patient with IgA gammopathy.* *Ann Dermatol Venereol.* 1995;122:97–101. [PubMed] [Google Scholar]
7. Villasante De La Puente A, HormaecheaBeldarrain JA, García Aguinaga ML, Vera López E, Gilsanz Fernández C. *Pustulosis of Sneddon Wilkinson disease and multiple myeloma.* *An Med Interna.* 2001;18:373–5. [PubMed] [Google Scholar]
8. Tsuruta D, Matsumura-Oura A, Ishii M. *Subcorneal pustular dermatosis and Sjögren's syndrome.* *Int J Dermatol.* 2005;44:955–7. [PubMed] [Google Scholar]
9. Saulsbury FT, Kesler RW. *Subcorneal pustular dermatosis and systemic lupus erythematosus.* *Int J Dermatol.* 1984;23:63–4. [PubMed] [Google Scholar]
10. Sakkas LI, Chikanza IC, Platsoucas CD. *Mechanisms of Disease: The role of immune cells in the pathogenesis of systemic sclerosis.* *Nat Clin PractRheumatol.* 2006;2:679–85. [PubMed] [Google Scholar]
11. Medsger TA, Jr, Ivanco DE, Kardava L, Morel PA, Lucas MR, Fuschiotti P. *GATA-3 up-regulation in CD8+T cells as a biomarker of immune dysfunction in systemic sclerosis, resulting in excessive interleukin-13 production.* *Arthritis Rheum.* 2011;63:1738–47. [PubMed] [Google Scholar]
12. Hudson M, Fritzler MJ. *Diagnostic criteria of systemic sclerosis.* *J Autoimmun.* 2014;48-49:38–41. [PubMed] [Google Scholar]