Case Report
Olgu Sunumu

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Subcorneal pustular dermatosis "Sneddon-Wilkinson disease": A case report

Subkorneal püstüler dermatoz "Sneddon-Wilkinson hastalığı": Bir olgu sunumu

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Abstract

Subcorneal pustular dermatosis (SPD), first described by Sneddon and Wilkinson in 1956, is rare, chronic, and relapsing dermatosis. Mostly, it affects women aged 40-50-years. It is characterized by grouped, annular, circinate, serpiginous, and sterile pustules on normal or erythematous ground. Lesions tend to involve intertriginous areas, the trunk, and extremities symmetrically. The etiology is not known; it has been suggested that it is a reactive neutrophilic dermatosis. In this report, we present a 71-year-old female who presented with lesions involving mostly intertriginous areas and received the clinical and histopathological diagnosis of SPD based on direct immunofluorescence assay. The patient was treated successfully with topical mupirocin 2% pomade, 1% blue de metylene, and oral macrolid 1 gr/day while being investigated for gammopathy and myeloproliferative diseases and waiting for biopsy results. After the biopsy results were obtained, the treatment of the patient continued with clobetasol 17-propionate 0.05%, colchium dispert 2x0.5 mg and dapsone 100 mg/day. The case was evaluated in the light of the relevant literature.
Keywords: Subcorneal pustular dermatosis, Sneddon-Wilkinson disease, neutrophilic dermatosis

Öz

İlk kez 1956'da Sneddon ve Wilkinson tarafından tanımlanan subkorneal püstüler dermatoz (SPD), nadir, kronik, tekrarlayıcı seyirli bir dermatozdur. Sıklıkla kadınlarda ve 40-50 yaşlarda izlenir. Normal veya eritemli zeminde gruplar halinde ortaya çıkan, anüler, sirsine, serpijinöz, steril püstüllerle karakterizedir. Lezyonlar intertrijinöz alanlar, gövde ve ekstremitelerde simetrik yerleşme eğilimindedir. Etiyolojisi bilinmeyen hastalığın reaktif nötrofilik dermatoz olduğu ileri sürülmektedir. Bu çalışmada, daha çok intertrijinöz yerleşimli lezyonlar şikayetiyle polikliniğimize başvuran; klinik, histopatoloji ve direkt immünfloresan inceleme sonucunda SPD tanısı konan, gamapati ve miyeloproliferatif hastalık yönünden tetkik edilip, biyopsi sonuçları beklenirken topikal mupirosin %2 merhem, %1 metilen mavisi ile 2x1 haricen boyama ve ampirik olarak oral makrolid 1 gr/gün ile tedavi edilip, biyopsi sonucu sonrasında %0,05 klobetazol-17 propiyonat, kolşisin 2x0,5 mg, dapson 100 mg/gün ile tedaviye devam ettiğimiz 71 yaşındaki bir kadın olgu literatür bilgileri eşliğinde sunulmuştur.
Anahtar Kelimeler: Subkorneal püstüler dermatoz, Sneddon-Wilkinson hastalığı, nötrofilik dermatozlar

Introduction

Subcorneal pustular dermatosis (SPD) has a clinical course with annular and circinate sterile pustules appearing in groups on a normal or erythematous background. Lesions tend to locate in intertriginous areas, the torso, and extremities. This is a benign condition, but the prognosis worsens if associated with neoplastic diseases. The etiopathogenesis is unknown and has been suggested to involve reactive neutrophilic dermatoses.

Case Report

A 71-year-old woman presented to our outpatient clinic with a 6-month history of recurring lesions on her torso, beneath the breasts and inguinal area which healed spontaneously.

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There was no notable personal or family history. Physical examination revealed no other pathologies. Dermatological examination showed erythematous plaques with mostly intertriginous localization and in the mons pubis, with pustules on an erythematous background ranging from 3 to 8 cm in diameter, with annular, circinate lesions with yellow squama and crusts (Figures 1a, 1b, 1c).

No pathologies were found on routine blood tests. Immunoglobulin (Ig) levels and peripheral smear examination performed to determine the presence of any associated diseases were normal. Protein electrophoresis results were normal. While IgA and Lambda light-chain monoclonal gammopathy were found on serum immunofixation electrophoresis, no monoclonal gammopathy was found on urinary immunofixation electrophoresis. The serum free kappa, Lambda, and beta-2 microglobulin levels and urinary kappa and Lambda levels were normal. Direct microscopic examination of the active border of the lesions was negative for fungi. No growth was observed on microbiologic studies repeated for samples collected from the pustular lesions in the inguinal areas.

Biopsy material was collected from the patient for histopathological and direct immunofluorescence (DIF) assay, with a prediagnosis of Hailey-Hailey disease, pemphigus foliaceus, IgA pemphigus, or SPD. While waiting for the biopsy results, considering Hailey-Hailey disease clinically, topical mupirocin 2% ointment b.i.d., 1% methylene blue staining, and an oral macrolide at a dosage of 1 g/day were started empirically. Histopathological examination indicated stratified squamous epithelium with parakeratosis, subcorneal pustule formation containing neutrophils and eosinophils, and inflammatory infiltration in the dermis containing lymphocytes and eosinophil leukocytes. There was no spongiosis (Figure 2a, 2b). On DIF, no staining of IgG, IgA, IgM, or C3c was seen.

Discussion

SPD has a clinical course with annular and circinate sterile pustules appearing in groups on a normal or erythematous background. Pustules are classically half-pustular and half-clear fluid-filled blisters, are opened easily, are covered with superficial squama and crusts within a short time, and heal with slight hyperpigmentation[1]. Palmoplantar involvement has also been reported, however, the face and mucous membranes are often preserved[1]. Patients describe mild itching or irritation but no other symptoms. This is a benign condition, but the prognosis worsens if associated with neoplastic diseases[1]. SPD is rare; etiopathogenesis is unknown and has been suggested to involve reactive neutrophilic dermatoses. Although the mechanism has not been clarified, the presence of abundant quantities of neutrophil chemoattractants, including tumor necrosis factor-α, interleukin-8, C5a, and IgA in blood; pustules, and squama suggest that they play a role in the pathogenesis[1]. The associations of SPD with benign monoclonal IgA, IgG gammopathy, and multiple myeloma included in myeloproliferative diseases are well known. As these diseases can develop even many years after the SPD diagnosis, long-term follow-up of patients is required[2]. There have been anecdotal reports of the development of the disease with pyoderma gangrenosum, seronegative and seropositive rheumatoid arthritis, hypothyroidism and hyperthyroidism, systemic lupus erythematosus, Sjögren's syndrome, Crohn's disease, multiple sclerosis, IgA myeloma, Synovitis-Acne-Pustulosis-Hyperostosis-Osteitis syndrome, following echocardiography, granulocyte-macrophage colony-stimulating factor injection, mycoplasma infection, chronic lymphocytic leukemia, metastatic thymoma, apudoma, and lung epidermoid carcinoma[3-7]. Histopathologically, SPD is characterized by sterile pustules containing numerous neutrophils and rare eosinophils with subcorneal localization. Lack of spongiosis is important, and acantholysis is not expected in old lesions, except for acantholytic cells detected because of the proteolytic enzymes released by neutrophils. Tests based on direct and indirect immunofluorescence are frequently negative. However, the occasional presence of IgA deposits located in the granular layer of the epidermis or in the subcorneal layer has suggested this as a potential new variant of SPD named subcorneal IgA pemphigus[1].

Dapsone, the first choice treatment for SPD, is used at a dose of 50-200 mg[1]. Systemic or topical steroids are used alone or in combination with dapsone. While systemic retinoids, acitretin and etretinate have been used, isotretinoin has been shown to be ineffective[5]. Psorolen-ultraviolet-A and phototherapy with narrow-or wide-band ultraviolet-B, alone or in combination with dapsone and/or retinoids, are effective[8]. Good results have also been obtained with infliximab and etanercept[1,5]. There have been anecdotal reports on the treatment with topical tacalcitol, sulfapyridine, sulfamethoxypyridazine, ketoconazole,

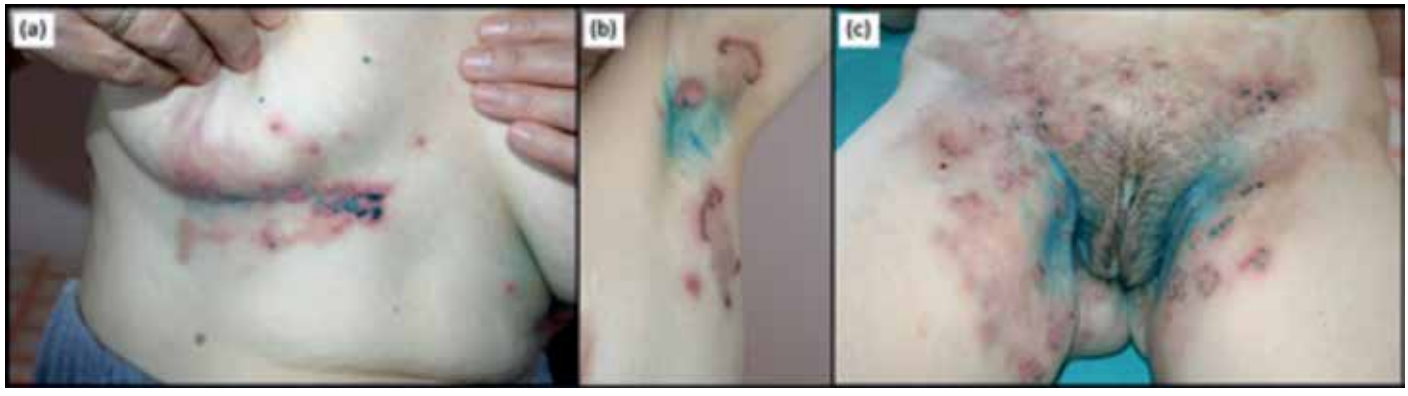

Figure 1. a) Annular, circinate erythematous plaques with pustules with yellow squama and crusts in right inframmamarian site with 1% methilen blue artifact, b) Annular, circinate erythematous plaques with pustules with yellow squama and crusts in left axillary site with 1% methilen blue artifact, c) Annular, circinate erythematous plaques with pustules with yellow squama and crusts in bilateral inguinal sites with 1% methilen blue artefact

tetracycline, minocycline, vitamin E, cyclosporine, colchicine, mizoribine, and mebhydrolin[1].

In the patient presented here, the recurring features of the lesions, tendency for location in intertriginous areas, fragility of pustules and being replaced by superficial squama and crusts, annular and circinate patterns, and negative results on repeated direct microscopic examinations, and microbiological studies suggested SPD. Observations of subcorneal pustules rich in neutrophils on histopathological examination and lack of spongiosis and negative staining on DIF supported the diagnosis. The Ig levels and peripheral smear results, performed to investigate any associated diseases, were normal. Serum immunofixation electrophoresis revealed IgA and Lambda light chain monoclonal gammopathy. The hematology department was consulted for this patient and did not suggest any pathologies to be present. Oral macrolide 2×500 mg and mupirocin 2% ointment, started before obtaining the biopsy results, were continued for 2 weeks. After obtaining biopsy results, topical 0.05% clobetasol 17-propionate treatment was started based on new lesions. While waiting for dapsone that was outsourced colchium dispert 2x0.5 mg; then, dapsone 100 mg/day were begun. Colchium decreases neutrophilic chemotaxis by inhibiting intracellular mycrotubulus systems. Dapsone also inhibits neutrophilic activation and chemotaxis by different pathways. During the third month of follow-up, the lesions had mostly regressed (Figures 3a, 3b, 3c).

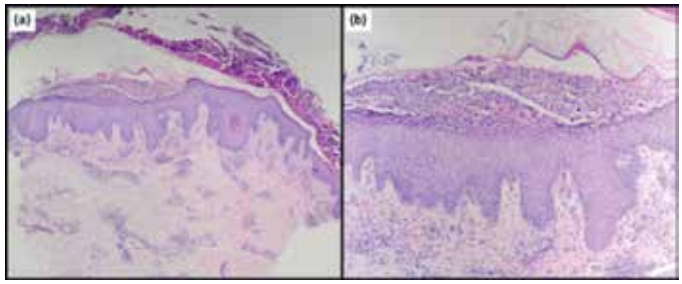

Figure 2. a) Parakeratosis in the stratified squamous epithelium, subcorneal pustule formation containing neutrophils and eosinophils (hematoxylin&eosin x100), b) Inflammatory infiltration in the dermis containing lymphocytes and eosinophil leukocytes. There is no spongiosis (hematoxylin&eosin x200)

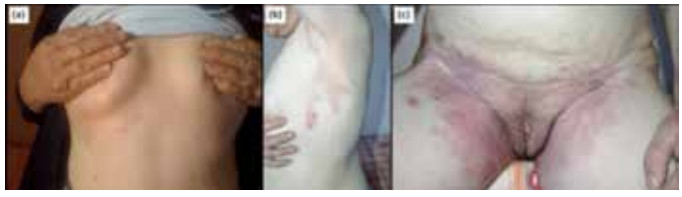

Figure 3. a) After the third month topical 0.05% clobetasol 17-propionate, colchium dispert 2x0.5 mg, dapsone 100 mg/day treatment on right inframmamarian lesions, b) After the third month topical 0.05% clobetasol 17-propionate, colchium dispert 2x0.5 mg, dapsone 100 mg/day treatment on left axillary lesions, c) After the third month topical 0.05% clobetasol 17-propionate, colchium dispert 2x0.5 mg, dapsone 100 mg/day treatment on bilateral inguinal lesions

In this paper, we present a 71-year-old female with lesions involving mostly the intertriginous areas who received the clinical and histopatological diagnosis of SPD based on DIF assay. The patient was treated successfully with topical mupirocin 2% pomade, 1% blue de methylene and oral macrolide 1 gr/day while being investigated and waiting for biopsy results. After the biopsy results were obtained, the treatment of the patient continued with clobetasol-17 propionate 0.05%, colchium dispert 2x0.5 mg, and dapsone 100 mg/day. The case was evaluated in the light of the relevant literature. This case represents a unique clinical picture, which highlights the requirement of long-term follow-up of patients with gammopathy and myeloproliferative disease.

Ethics

Informed Consent: Consent form was filled out by all participants.
Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Aslı Şahin, Hatice Meral Ekşioğlu, Emine Yalçın Edgüer, Concept: Aslı Şahin, Data Collection or Processing: Aslı Şahin, Analysis or Interpretation: Aslı Şahin, Literature Search: Aslı Şahin, Hesna Müzeyyen Astarcı, Writing: Aslı Şahin.

Conflict of Interest: No conflict of interest was declared by the authors.

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