Case report

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Successful treatment of generalized subcorneal pustular dermatosis involving mucosa with cyclosporine: a case report

Mukozayı da etkileyen bir generalize subkorneal püstüler dermatozun siklosporin ile başarılı tedavisi: Bir olgu sunumu

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Abstract

Subcorneal pustular dermatosis (SPD), is rare, chronic, and relapsing pustuler eruption. Mostly, it affects middle-aged and elderly women. Mucosal involvement is extremely rare. Here we report a 31-year-old woman with generalized SPD with lip involvement. The patient had annular or serpiginous patterned well-demarcated plaques on the trunk, intertriginous areas and flexor aspects of the limbs, palmoplantar areas, face, and lip mucosa. The bullous lesions exhibited the typical half and half sign. The patient was treated successfully with systemic steroids and cyclosporine.

Key words: *subcorneal pustular dermatosis, Sneddon-Wilkinson disease, mucosa, lip, cyclosporine*

Öz

Subkorneal püstüler dermatoz (SPD), nadir görülen, kronik ve tekrarlayan püstüler erupsiyonlarla karakterize bir dermatozdur. Çoğunlukla orta yaş ve yaşlı kadınları etkilemektedir. Mukozal tutulum oldukça nadirdir. Burada dudak tutulumu da gelişen jeneralize SPD tanılı 31 yaşında bir kadın hasta bildirilmektedir. Hastanın gövdesinde, intertrijinöz alanlarda ve ekstremitelerin fleksör yüzlerinde, palmoplantar alanlarda, yüz ve dudak mukozasında annüler, serpijinöz desenli, iyi sınırlı plaklar mevcuttu. Büllöz lezyonlarda tipik hipopiyon işareti mevcuttu. Hasta sistemik steroid ve siklosporin ile başarılı bir şekilde tedavi edildi.

Anahtar kelimeler: *subkorneal püstüler dermatoz, Sneddon-Wilkinson hastalığı, mukoza, dudak, siklosporin*

Introduction

Subcorneal pustular dermatosis (SPD), also known as Sneddon-Wilkinson disease, is an uncommon, benign yet chronic, relapsing neutrophilic dermatosis.[1] The cutaneous lesions characterized by sterile pustules, often

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in annular and circinate configurations, are typically found on the trunk, intertriginous areas and flexor aspects of the limbs. The involvement of the face and mucosal surfaces is infrequently reported.[2-5] Herein, we present a case of a young woman diagnosed with generalized SPD, which resulted in erythroderma and involved the face and the lip treated with systemic corticosteroid and cyclosporine.

Case

A 31-year-old female presented to the outpatient clinic with a widespread, pruritic, erythematous rash and pustular eruption prominent on the trunk. She had malaise, and pain on the lesional skin. The patient admitted that her complaints started one month ago as circular 1-2 cm diameter lesions on the neck and the legs. She was prescribed topical isoconazole, systemic amoxicillin and clavulanic acid in another medical centre for ten days. Due to the lack of improvement, oral terbinafine was recommended, presumably with a tinea corporis diagnosis. She applied to our outpatient clinic for further medical attention due to the progression of the lesions.

Dermatological examination revealed widespread involvement of the trunk, intertriginous and flexural surfaces with numerous pustules.

Multiple scaly plaques forming annular patterns and pustules on an erythematous inflammatory background were seen (Fig. 1a). The pustules typically appeared as "half-half blisters", also known as "hypopyon sign" (Fig. 1c). The most severely affected areas on the first day were the trunk and proximal extremities. Gradually, involvement spread peripherally, including distal extremities, palmoplantar areas and the face, involving >75% body surface area. Personal medical history and family history were insignificant.

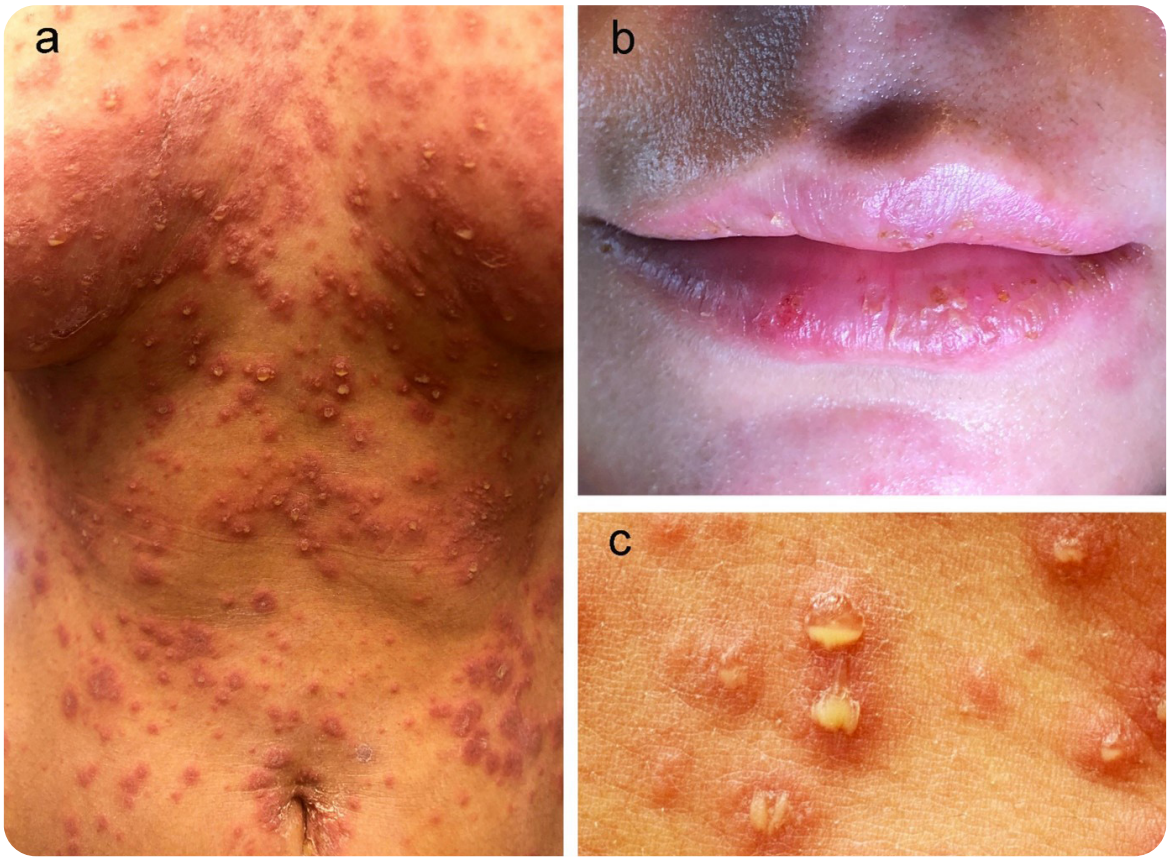

Fig. 1a. Multiple scaly plaques in annular configurations and sterile pustules on an erythematous base, **1b.** Pustular lesions on lip mucosa, **1c.** Half-half blisters on the trunk with both clear and yellow fluid

A complete blood cell count showed neutrophilic leukocytosis, which reached a level of 80% neutrophils. Erythrocyte sedimentation rate (ESR) (35 (0-20 mm/hour)) and C-reactive Protein (CRP) rate (15 (0-5 mg/L)) were elevated. Other serum biochemistry findings were unremarkable. Bacterial and fungal stains and cultures from pustules were negative. A complete urinalysis test showed leukocytes but without urinary tract infection.

A skin biopsy was performed for histopathological and direct immunofluorescence (DIF) assay. Histopathologic examination revealed subcorneal pustule, filled with neutrophils. There was neutrophilic spongiosis at the epidermis beneath the pustule. Superficial perivascular inflammatory cell infiltrate, containing neutrophils, lymphocytes and histiocytes, was present in the dermis (Fig. 2). Periodic acid-Schiff (PAS) stain was unremarkable, and direct immunofluorescence was negative.

The patient was diagnosed with SPD based on the clinical and histopathologic findings. Serum immunoelectrophoresis did not reveal paraproteinemia. Serum Immunoglobulin (Ig) levels were normal. Anti-HIV ELISA test and syphilis serology were negative. The antinuclear antibody panel was negative. Treatment was initiated with a dose of 2 mg/kg/day intravenous methylprednisolone for three consecutive days, followed by 1 mg/kg/day for five days. After admission to the hospital, the patient developed erythroderma with an instant generalization of the lesions to the extremities and palms. The face and palmoplantar area involvement were prominent. New onset of pustular lesions was also observed on the lip mucosa. (Fig. 1b). Other mucous membranes

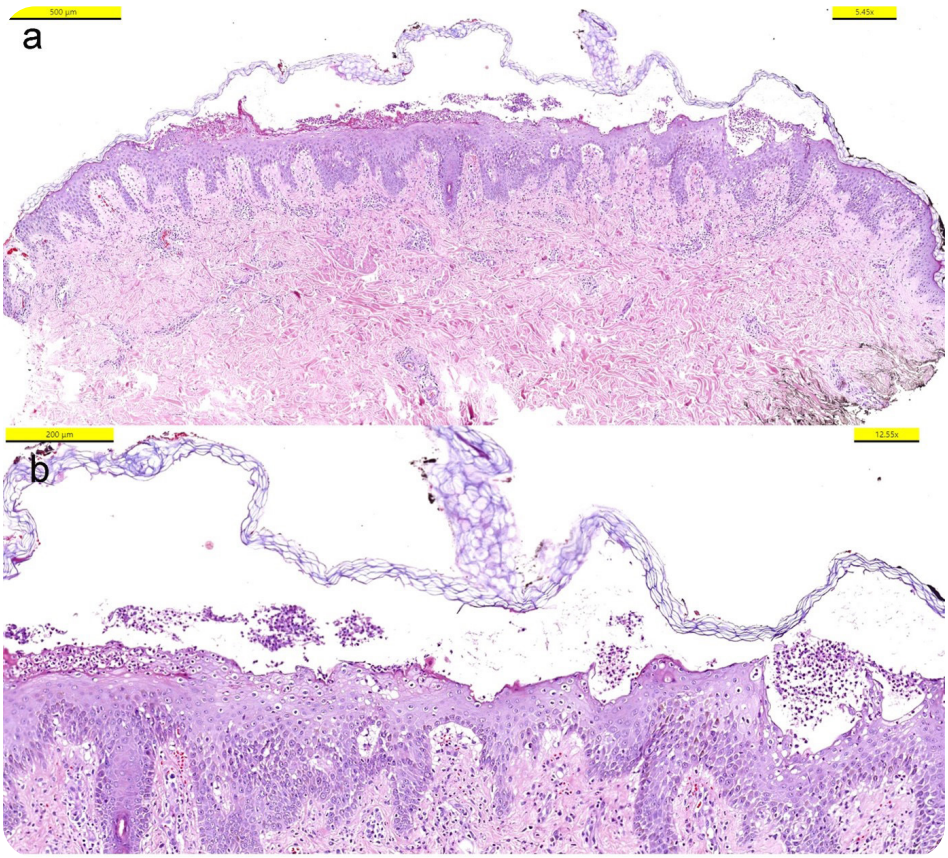

Fig. 2. Subcorneal pustular dermatosis: **2a.** Subcorneal pustule contains plenty of neutrophils (HEx5.45), **2b.** Neutrophils migrate through the epidermis underneath the pustule (HEx12.55).

were spared. Testing for the Nikolsky sign was negative. Oral cyclosporine 3 mg/kg/day was added to the treatment on the third day of hospitalisation.

The patient achieved significant improvement and the lesions resolved within a week. At maintenance, the corticosteroid was tapered off in 15 days, and cyclosporine was stopped within a month. No recurrence was observed during the 2-month follow-up period.

Discussion

SPD is a rare, chronic, recurrent pustular eruption that Sneddon and Wilkinson first described in 1956.[1] It is more common in women and middle-aged and elderly groups. Pustules are classically half-pustular, and half-clear fluid-filled blisters easily ruptured, covered with superficial squama and crusts within a short time and arise on normal skin or a slightly erythematous base. They merge to form annular or serpiginous patterned, well-demarcated plaques.[2] Lesions are classically localized on the trunk, proximal extremities, flexural surfaces such as the axilla, groin, inframammary folds and intertriginous areas. The palms, soles, nails, face and mucosal surfaces are generally spared.[6] Face involvement is described in a few cases.[3-5] On the other hand, SPD initially started as a few lesions, which rapidly generalized with face and mucosal involvement in our patient.

Generally, lesions progress over within a day or two to form a generalized distribution.[7]

The etiopathogenesis of SPD is not clear up to date. In addition, well-documented SPD cases have been associated with a broad spectrum of cutaneous and systemic disorders. These include pyoderma gangrenosum, multiple myeloma, IgA monoclonal gammopathy, lymphomas, solid organ malignancies, autoimmune connective tissue disorders such as Sjogren's syndrome, systemic lupus erythematosus, rheumatoid arthritis; thyroid disorders, drugs and infections.[6] We detected none of the associated diseases in our patient. The drugs associated with SPD are isoniazid, gefitinib, paclitaxel, cefazolin, and amoxicillin.[6] Although our patient had used amoxicillin, the treatment had initiated after the onset of lesions.

Dapsone is the first-line treatment of choice for SPD. Alternative treatments are required for patients with low G6PD levels, patients with anemia and in resistant disease. Treatment options other than dapsone include other anti-neutrophilic drugs such as colchicine, sulfapyridine, and sulfamethoxypyridazine; topical and systemic corticosteroids, cyclosporine, oral retinoids such as acitretin and etretinate; UVA and UVB light therapies, in various combinations.[8] For resistant cases, anti-TNF-α agents infliximab and etanercept have shown good responses.[6] Systemic corticosteroids are generally used for a short duration in the presentation of the disease. Thus, for our patient, systemic corticosteroid treatment has commenced initially. We started cyclosporine for a fast response, and stabilization was accomplished with cyclosporine added to therapy due to rapid and severe progress. The lesions faded significantly with cyclosporine treatment.

In conclusion, we present an atypical case of SPD. Although SPD is generally considered a self-limiting disease, in our case the patient developed erythroderma with lip mucosa involvement which required an immediate treatment. Our literature review did not reveal any similar cases in terms of face and lip involvement. We would like to highlight this rare presentation and the efficacy of cyclosporine as an alternative treatment.

Informed consent: Consent was gained from this patient for publication and presentation of her clinical information, case and images.

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