A case of subcorneal pustular dermatosis (Sneddon-Wilkinson-disease) not responding to dapsone: therapeutic alternatives

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SUMMARY

We report a case of subcorneal pustular dermatosis (SPD) which did not respond to the medication with dapsone or retinoids. When the medication was changed to systemic corticosteroids we observed a dramatic improvement of the skin lesions. This case underlines the usefulness of systemic corticosteroids as a therapeutic alternative when standard therapy fails. A dose maintenance is necessary to avoid relapses.

KEY WORDS

subcorneal pustular dermatosis, Sneddon-Wilkinson-disease, systemic corticosteroids

Introduction

The international recommended therapy of subcorneal pustular dermatosis (SPD), after exclusion of G-6-PDH deficiency, is dapsone in a dosage of 50-150 mg/d (1). There are also reports of therapeutic alternatives i. e. with retinoids alone or in combination with PUVA (2,15). Additionally to the mentioned possibilities other therapeutic schemes may be applied, e.g. the administration of systemic corticosteroids (3).

We will highlight a case of SPD initially treated with dapsone and retinoids without obtaining a clinical improvement, that showed complete remission after treatment with corticosteroids.

Case report

A 65-year-old male patient suffering from a 4-year relapsing pustular eruption involving palms, soles and the trunk. Figure 1. The pustules tended to be grouped, they were flaccid and ruptured easily. Oral lesions were not seen.

Treatment with dapsone, PUVA and systemic retinoids was ineffective. Biopsies of lesional skin revealed subcorneal pustules filled with neutrophils. Figure 2. Direct Immunofluorescence was negative. Peripheral blood counts showed neutrophilia (80.4%) and lymphopenia (11.1%). The white blood count was $11.8x10^9$/l. The ESR was elevated 55/82 mm. Serum immuno-electrophoresis revealed an polyclonal eleva-

tion of immunoglobulins: IgA 1300 mg/dl and IgG 1820 mg/dl, while the IgM level was normal, 222 mg/dl. There was no evidence of a monoclonal gammopathy. CT-scan studies, PET-investigations and the peripheral blood ecell-count showed no signs of malignancy. A familiar IgA elevation was not detected.

The oral administration of dapsone during the 7-week hospitalization was ineffective. The patient received PUVA-therapy and oral retinoids but did not respond to this treatment.

Due to the severe course with worsening of the general condition we started a systemic corticosteroid regimen: fluocortolone 20 mg/d. A dramatic improvement of the skin lesions followed within 2 days. A complete remission was maintained with 12 mg fluocortolone/d orally over one year. Under this treatment IgA levels decreased to 625 mg/dl.

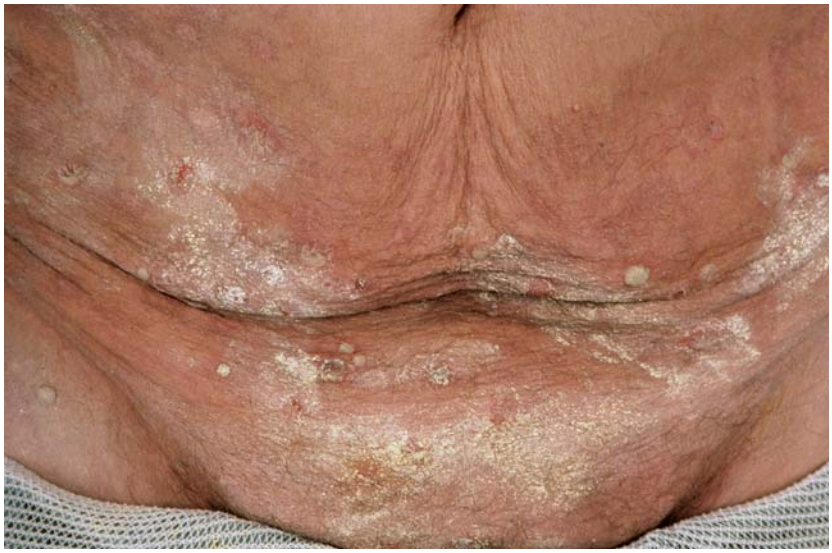

Figure 1. Patient reported,. Pustules on the skin.

Discussion

Subcorneal pustular dermatosis is a rare, chronic relatively benign relapsing pustular eruption which affects mainly patients between 30 and 70 years. It was first described as an entity in 1956 by Sneddon and Wilkinson (4).

It is a sterile pustular eruption and the vast majority of the reports describe elevated IgA levels, perhaps acting as a chemo-attractant for neutrophils (5). The exact pathophysiology is unknown. Hyperactivation of cutaneous neutrophils in SPD may be partly caused by excessive production of tumor necrosis factor-alfa (TNF-á) (3). SPD has been associated with a number of other conditions. A relationships with pyoderma gangrenosum (6), multiple myeloma (7), benign monoclonal gammopathy-IgA (8,9), rheumatoid arthritis (10), systemic lupus erythematosus (11), hyperthyroidism (12), Crohn disease (13) and multiple sclerosis (14) have been described.

Serum protein electrophoresis should be repeated in appropriate intervals because a paraproteinemia may develop several years later (15).

SPD cases refractory to first-line medications like dapsone and retinoids present a therapeutic challenge, and the usefulness of systemic corticosteroids in such patients has not often been described. In cases non-responding to the first-choice medications we suggest the use of systemic corticosteroids as a therapeutic option and a long-term monitoring of the patient's condition.

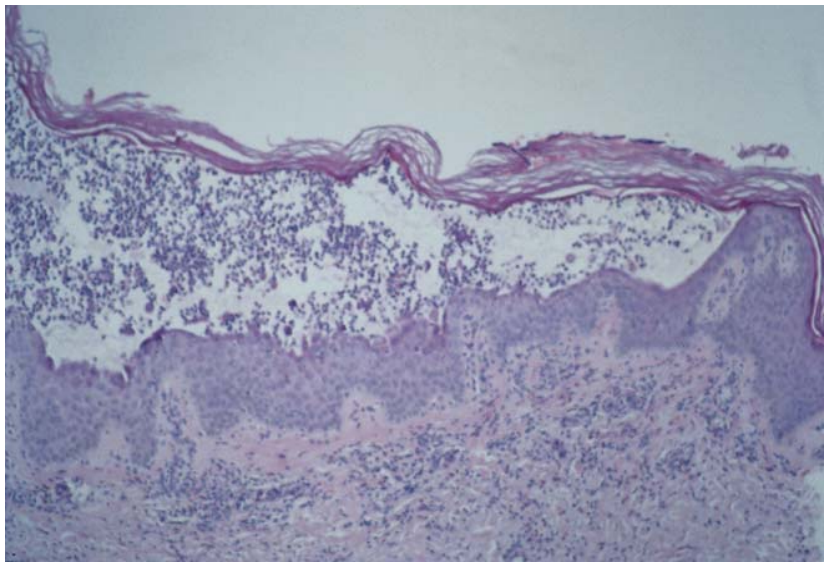

Figure 2. Same patient. Biopsy revealing subcorneal pustules filled with neutrophils.

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