A resistant case of subcorneal pustular dermatosis with an excellent long-lasting response to adalimumab

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Sneddon-Wilkinson disease, also known as subcorneal pustular dermatosis (SPD), is a relapsing pustular dermatosis of unknown etiology. The vesiculopustules typically present on the trunk and intertriginous areas. The mainstay of SPD treatment is dapsone; second-line therapies include corticosteroids and other immunosuppressive drugs. Here, we report a case of recalcitrant severe SPD that responded dramatically to adalimumab. The patient was a 30-year-old man who presented with generalized erythroderma and vesiculopustules on the trunk and extremities. In some pustules, the pus settled with gravity into the lower half of the blister. He responded dramatically to a combination of adalimumab (80 mg on day 1, 40 mg on day 7, and 40 mg biweekly afterward) and prednisolone 50 mg daily. During follow-up, prednisolone was tapered to 5 mg daily over 2 months, and adalimumab was maintained biweekly; he is still in remission after 8 months. Thus, adalimumab can be used as an effective and easy-to-use treatment in refractory cases of SPD.

Keywords: Sneddon-Wilkinson disease, subcorneal pustular dermatosis, adalimumab

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INTRODUCTION

Sneddon-Wilkinson disease, also known as subcorneal pustular dermatosis (SPD), is a relapsing pustular dermatosis of unknown etiology. The vesiculopostules typically present on the trunk and intertriginous areas. The mainstay of SPD treatment is dapsone; second-line therapies include corticosteroids and other immunosuppressive drugs [1].

Here, we report a case of SPD who was unresponsive to dapsone, corticosteroids, mycophenolate mofetil, and acitretin for 4 years. He responded dramatically to adalimumab.

CASE PRESENTATION

The patient was a 30-year-old man who presented with generalized erythroderma and vesiculopustules on the trunk and extremities (Figure 1). In some pustules, the pus settled with gravity into the lower half of the blister. He was on acitretin 25 mg daily with no improvement over the course of 2 months prior to admission. He also used dapsone 150 mg daily, prednisolone 50 mg daily, and mycophenolate mofetil 2 g daily during the 4 years of disease without complete improvement and with frequent exacerbations.

The previous biopsy showed subcorneal pustules

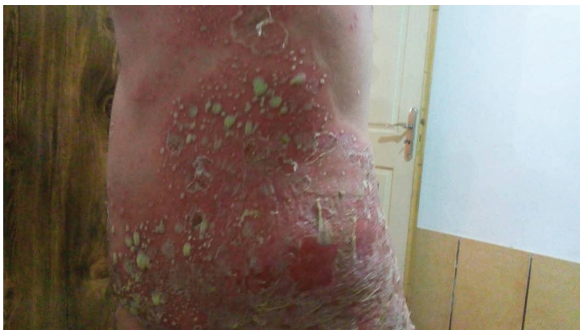

Figure 1. Severe vesiculopustule formation on the trunk before commencing adalimumab.

with acanthosis and exocytosis of neutrophils; the papillary dermis showed mixed neutrophils and lymphocytes around the vessels. Microscopic examination showed subcorneal neutrophilic pustules with mild acanthosis, exocytosis of neutrophils, and acantholytic changes at the granular layer. The upper dermis revealed mild mixed inflammatory cell infiltrate composed of lymphocytes, neutrophils, and few eosinophils (Figure 2). Direct immunofluorescent examination was negative in two biopsy specimens.

He was admitted to the dermatology ward. Acitretin was discontinued. Adalimumab 80 mg on day 1 and prednisolone 50 mg daily were commenced for him. The patient responded dramatically to the combination: within 2 days of treatment, all of the pustules dried off (Figure 3). Adalimumab was prescribed at the dose of 40 mg on day 7 and 40 mg biweekly afterward. During follow-up, prednisolone was tapered to 5 mg daily over 2 months, and adalimumab was maintained biweekly. The patient is still in remission 8 months after treatment on adalimumab.

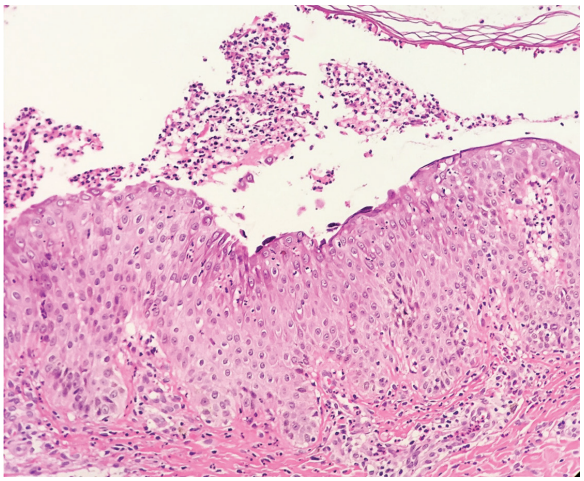

Figure 2. Subcorneal neutrophilic pustules, few acantholytic cells, mild acanthosis, exocytosis of neutrophils, and acantholytic changes in the granular layer. The upper dermis reveals a mild, mixed inflammatory cell infiltrate (H & E, 200×).

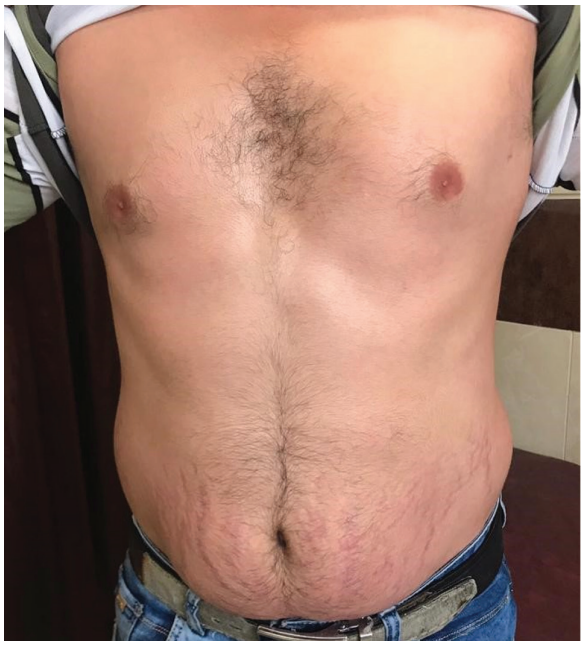

Figure 3. Dramatic response after 8 months of adalimumab therapy.

DISCUSSION

Although dapsone is known as the first-line treatment for SPD, not all cases of SPD respond well to this drug. There are a few case reports of the use of anti-tumor necrosis factor (TNF)-α biologics as a treatment for resistant cases of SPD. In SPD, sterile pustules containing neutrophils are noted as the hallmark of the disease, but the exact pathogenesis is still unknown. Some chemokines, including TNF-α, can lead to the accumulation of neutrophils [2]. Jean Jacques Grob *et al.* measured the levels of TNF-α in the serum, pustules content, and supernatant of monocytes of a resistant case of SPD. They concluded that the accumulation of neutrophils may at least partly be due to TNF-α excess [3].

Etanercept has also been used previously in cases of SPD with success [4,5]. In the report of Lorenz Kretschmer *et al.*, just one dose of infliximab resulted in the resolution of severe atypical SPD

in a 29-year-old patient [6]. But in another report, the improvement was only short lasting with infliximab [7]. There are only two previous case reports that used adalimumab in recalcitrant cases of SPD with success [2,8]. There is also a report of SPD in the palmoplantar area as a side effect of adalimumab therapy in a rheumatoid arthritis patient [9]. It resolved after adalimumab was discontinued.

This case report highlights the importance of adalimumab therapy in severe cases of SPD that remain unresponsive to first-line therapies. Adalimumab resulted in a dramatic, lasting improvement in our patient, who was resistant to standard therapies. Hitherto, the patient has been on this new drug regimen for 8 months and is still on adalimumab 40 mg biweekly. He is still in remission with satisfactory results and no side effects.

According to this case, adalimumab can be used as an effective and easy-to-use treatment for recalcitrant SPD.

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