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CASE REPORT

Transient effectiveness of dapsone for skin lesions in a patient with discoid lupus erythematosus

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ABSTRACT

Dapsone is a synthetic sulfone preparation and is second-line treatment for cutaneous lupus erythematosus (CLE) including discoid LE (DLE). This report describes a patient with DLE, which was improved by dapsone quickly but the recurrence was noted. Oral administration of prednisolone (5 mg/day) was required but the complete healing was very difficult. In this report, we will discuss the effectiveness and limitation of dapsone for patients with DLE and the mechanisms of action.

***Keywords**: dapsone; diaminodiphenyl sulfone; DDS; discoid lupus erythematosus*

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Introduction

For cutaneous lupus erythematosus (CLE) patients, topical treatments of corticosteroid or calcineurin inhibitor are recommended worldwide[1,2]. As the next step, hydroxychloroquine (HCQ) and/or chloroquine are often recommended as the first choice for systemic therapy, and dapsone (diaminodiphenyl sulfone, DDS), along with immunosuppressants such as methotrexate and corticosteroids belong to the second option group[3]. However, the effectiveness and limitation of dapsone for patients with discoid LE (DLE) and the mechanisms of action are not fully understood.

Case Report

A 46-year-old Japanese man had noticed erythematous lesions on his cheeks and nose twenty five years before the first visit. Then, the skin eruptions spread to the back and other regions after sunburn. He was referred to our department. Erythematous lesions with hyperkeratosis and a slight scaling were observed in his face, upper back, and forearms (**Figure 1**). Fever, joint pain, stomatitis or hair loss was not observed. Blood tests showed no abnormality in blood count and biochemical examinations, and complement components were within normal range. The anti-nuclear antibody was 80 times, but particular autoantibodies were not detected. Urine tests were also normal. Pathological examination of the erythematous lesion revealed thinning and flattening of the epidermal layer, vacuolar degenerations in the basal layer and lymphocytes infiltration at the dermoepidermal junction. Lymphocyte infiltration was also observed at perivascular area in the dermis, and hyperkeratotic pluggings were found in the hair follicle. Deposition of IgG, IgM and C3 was found in the basement membrane by direct immunofluorescent staining.

Based on the clinical appearance and pathological findings, he was diagnosed as chronic CLE (CCLE) or disseminated DLE[4]. He received oral administration of 75 mg/day dapsone and topical hydrocortisone butyrate. The erythematous lesions were improved quickly to those of slight erythema

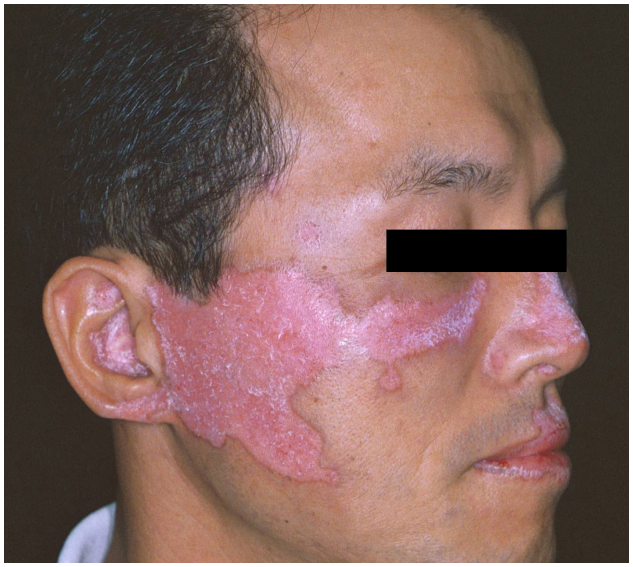

Figure 1. Clinical lesions. Erythematous lesions with hyperkeratosis and a slight scaling were observed in his face, upper back, and forearms

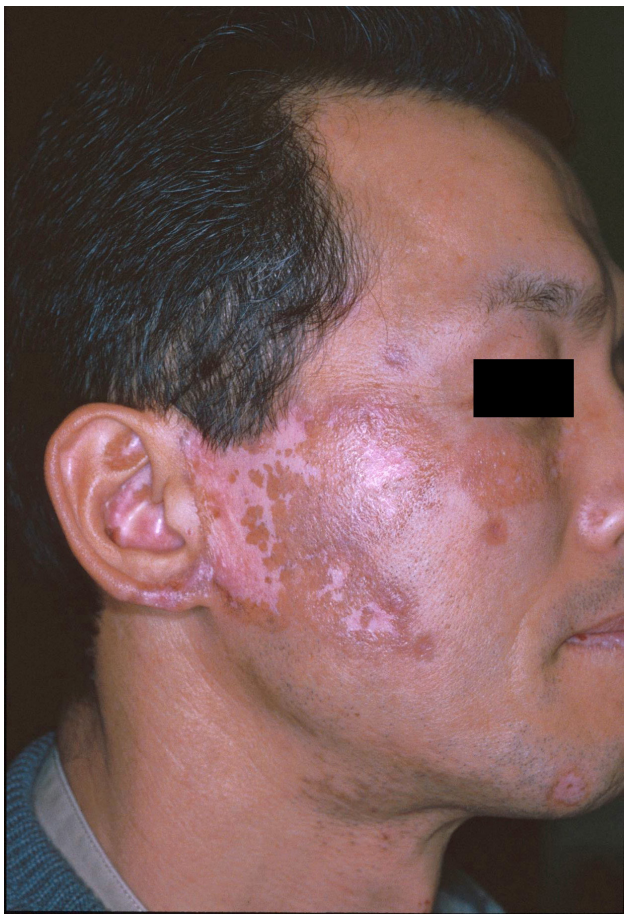

Figure 2. Improved clinical lesions. The erythematous lesions were improved quickly to those of slight erythema and pigmentation 4 months after treatment.

and pigmentation 4 months after treatment (**Figure 2**). However the recurrence was noted soon (**Figure 3**). Additional oral prednisolone (5mg/day) was administered, but the complete healing was very difficult. There were no side effects by dapsone during the course. HCQ could not be prescribed in the period of clinical course.

Discussion

In the treatment of CLE, the ultraviolet protection is important for prevention and topical treatment of corticosteroid or calcineurin inhibitor is recommended[1,2,5]. Systemic treatments are used for cases with extensive skin rash and/or intractable cases[1,2]. Antimalarial drugs such as HCQ and chloroquine (unapproved in Japan) are often recommended as the first choice for systemic therapy[1,2,6,7,8]. Dapsone, immunosuppressants such as methotrexate and corticosteroids are the second option[1,2,3].

Dapsone is a synthetic sulfone preparation and is used for several dermatological disorders such as leprosy, cutaneous vasculitis, systemic vasculopathy, autoimmune bullous disease and prurigo pigmentosum[9,10]. The mechanisms of action of dapsone are not fully understood, but it is thought to be central to the suppression of neutrophil function by neutrophil migration or inhibition of myeloperoxidase[9,11]. Additionally, neutrophil extracellular traps have attracted attention in recent years as the involvement of neutrophils in autoimmune diseases[11].

We treated this case of widespread DLE with dapsone and a low dose systemic corticosteroid. Tips for dapsone use in this case are as follows; 1) initial effectiveness, 2) requirement of a small dose of oral corticosteroid in the tapering of dapsone, 3) a careful attention for side effects.

As side effects, liver dysfunctions and hemolytic anemia are relatively frequent; we should pay attentions to severe side effects such as DDS (diaminodiphenyl sulfone) syndrome, drug-induced hypersensitivity syndrome, methemoglobinemia, leukopenia etc.[3,9] The careful follow-up is essentially important.

Today, CLE treatment in Japan is approached to overseas standard treatment due to approval of HCQ[6,7]. However, recent clinical study reports that dapsone was effective in more than half cases as treatments for second choice of CLE of various

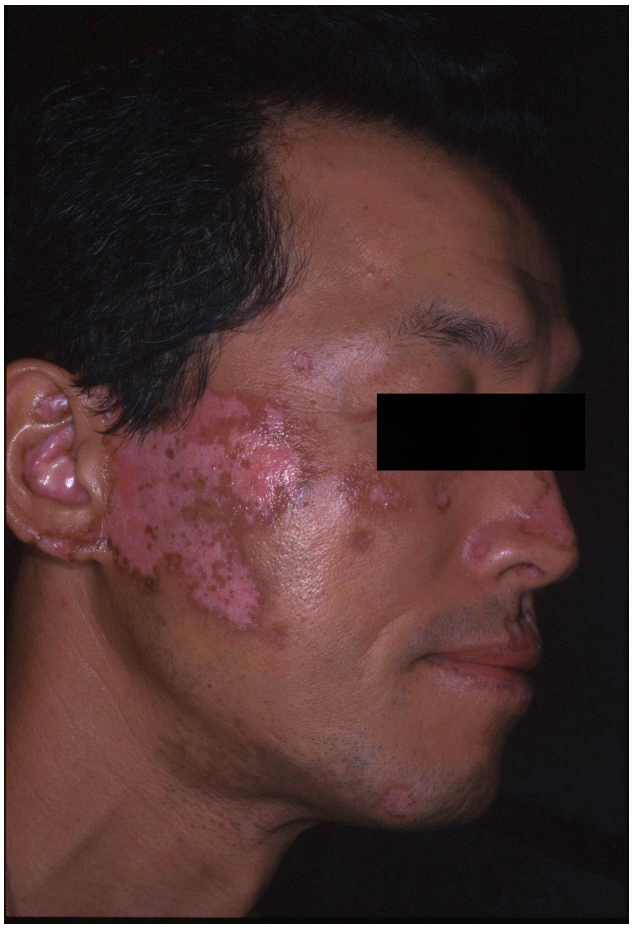

Figure 3. Recurrence lesions

disease types[3]. There are also cases in which HCQ cannot be used because of retinopathy; dapsone is still considered as one of the useful options.

Conflict of interest

We have no conflicts in this report. This case was published in Japanese (Visual Dermatology 16: 118-120, 2017) as an invitation article.

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