IJD Case Report

Pityriasis lichenoides chronica successfully treated with combination of narrowband UVB phototherapy and cyclosporine: a case report

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Pityriasis lichenoides chronica (PLC) is a chronic skin disorder with unknown etiology. This disorder still poses difficulty in diagnosis and treatment. Currently, there is no guideline established for the treatment of PLC. Most of the proposed therapy show transient or limited effects. Combination therapy is usually the best approach. A 40-year-old man presented with erythematous papules appearing recurrently in crops in the last nine months localized mainly to the trunk and proximal extremities. A complete response was not achieved with topical and systemic corticosteroids. The combination of narrowband UVB phototherapy and cyclosporine showed satisfying results following a short duration of therapy. A complete response was seen after eight sessions of narrowband UVB phototherapy. No side effects were observed. The combination therapy of phototherapy and cyclosporine is a potential choice that needs to be considered in the management of PLC.

Keywords: cyclosporine, phototherapy, pityriasis lichenoides, therapeutics

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INTRODUCTION

Pityriasis lichenoides chronica (PLC) is an uncommon, chronic skin disorder of unknown cause that presents numerous challenges to clinicians. The prevalence of PLC is estimated at 0–0.5%, seen more frequently in males during late childhood or young adulthood. PLC usually manifests as recurrent crops of erythematous-to-brown scaly papules that spontaneously regress within several weeks to months. This disorder poses diagnostic and therapeutic challenges. Most available treatments have a transient or limited effect. The mainstay of therapy is a combination of topical corticosteroids and phototherapy. Other treatments include antibiotics, topical tacrolimus, methotrexate, and cyclosporine [1-4]. This paper reports our experience treating a patient with PLC using narrowband UVB plus cyclosporine.

CASE PRESENTATION

A 40-year-old man was referred to our dermatology and venereology clinic due to pruritic, erythematous papules that spread all over his body in the last nine months. The lesions initially appear on the abdomen, then spread to the chest, neck, head, back, and limbs. The lesions usually appeared recurrently in crops, sometimes disturbing the patient's daily activities with an average itch visual analog scale score of 4–5 out of 10. There was no history of atopy, drug intake prior to the lesions, or drug or food allergies. Five months ago, the patient was treated with methylprednisolone

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and clobetasol propionate ointment; the lesions resolved for a short period but then reappeared when methylprednisolone was stopped. The patient was then treated with cyclosporine 100 mg twice daily for four months. The clinical appearance improved, but new lesions still occurred recurrently, only achieving partial remission.

On physical examination, the patient's vital signs were within normal limits. Dermatological examination revealed multiple erythematous papules on the chest, abdomen, back, and thighs (Figure 1). The complete blood count, liver enzyme test, kidney function test, and total IgE were within normal limits. A skin biopsy demonstrated mild orthokeratosis, spongiosis, and a perivascular lymphocyte infiltrate, with the pathologist diagnosing chronic dermatitis.

The patient started narrowband UVB whole-body phototherapy twice weekly with an initial dose of 300 mJ/cm^2. The phototherapy dose was increased by 10% in each session. Cyclosporine was continued at 100 mg twice daily for four weeks, and clobetasol propionate ointment was applied to the erythematous papules. After eight phototherapy sessions, no new lesions appeared, and the previous lesions began to disappear. The cyclosporine dose was tapered down to 100 mg a day. No side effects were observed. Dermatological examination revealed multiple hyperpigmented macules and papules on the chest, abdomen, back, and thighs (Figure 2).

This patient underwent a total of 12 phototherapy sessions. The dose of cyclosporine given was tapered down to 1 x 50 mg daily. After three months of therapy, the patient's condition tended to be stable, with only 1–2 non-spreading lesions appearing.

Ethical consideration

The authors declare that they have obtained the patient consent form. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. Patient consent form was uploaded during manuscript submission.

DISCUSSION

Pityriasis lichenoides (PL) encompasses a spectrum of inflammatory diseases that consists

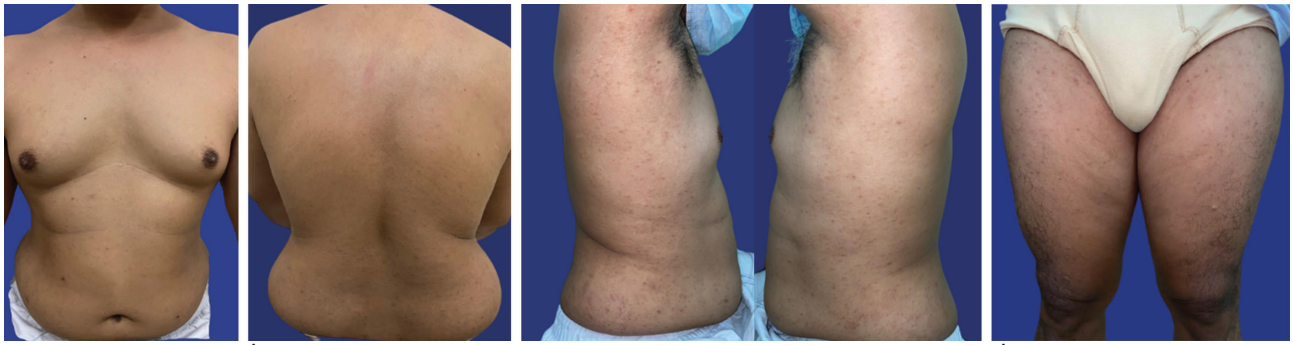

Figure 1. Erythematous papules on the trunk (a), back (b), lateral parts of the trunk (c) and thighs (d) on the first visit

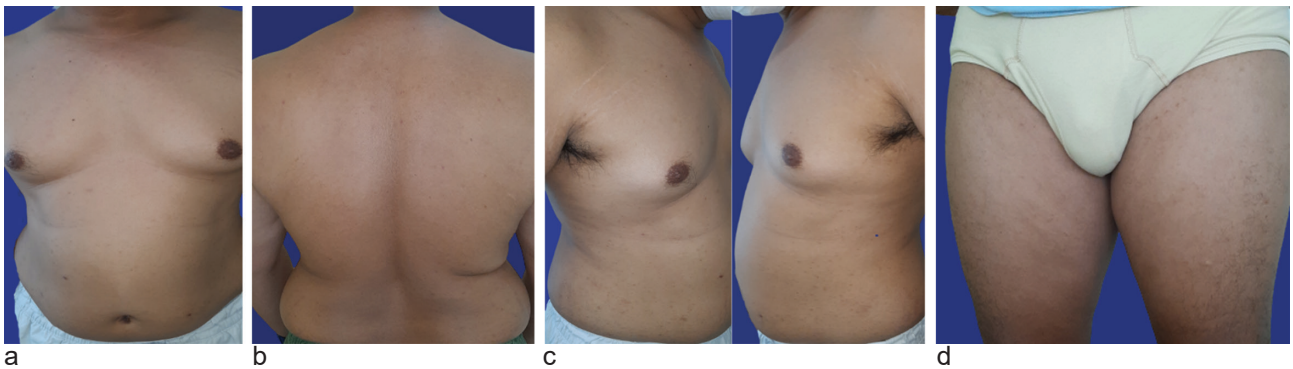

Figure 2. Hyperpigmented macules and papules on the trunk (a), back (b), lateral parts of the trunk (c) and thighs (d) after eight sessions of phototherapy

of pityriasis lichenoides chronica (PLC), pityriasis lichenoides et varioliformis acuta (PLEVA), and febrile ulceronecrotic Mucha-Habermann disease (FUMHD). PLC is relatively more benign than PLEVA and FUMHD. The lesions initially present as erythematous papules that develop a reddish-brown hue and centrally adherent scales. The papules can spontaneously flatten and regress over time, leaving hyperpigmented or hypopigmented macules. There are often periods of exacerbations and remissions that can take several years. PLC can be found mainly on the trunk and proximal extremities, but acral and segmental distributions have also been reported [5,6]. Our patient presented with erythematous papules that appeared recurrently in crops and were localized mainly to the trunk and proximal extremities. This clinical presentation suggested the diagnosis of PLC.

Besides the clinical presentation, the diagnosis of PLC can be confirmed by histopathology examination. Interface dermatitis with an infiltrate composed mainly of lymphocytes is usually observed in PLC. Exocytosis, focal parakeratosis, mild to moderate acanthosis, focal areas of spongiosis, necrotic keratinocytes, and extravasation of erythrocytes can also be found in the epidermis. In the dermis, edema and mild superficial perivascular lymphohistiocytic infiltrate can also be seen [2,6,7]. Histopathology examination of the patient's skin demonstrated a mild orthokeratosis with spongiosis and perivascular lymphocyte infiltrate may suggest the diagnosis of PLC.

The pathophysiology of PL remains uncertain. One hypothesis states that PL is a form of atypical immune response to a foreign agent in a genetically susceptible person. Various pathogens have been associated with PL, such as HIV, Epstein-Barr virus, varicella-zoster virus, *Streptococcus,* and *Staphylococcus*. Another theory states that PL is a T-cell premycotic disorder [1].

The symptomatic and cosmetic problems of PL have directed the elaboration of several treatment choices despite its benign course. Each treatment has different success rates and adverse effects. No standard treatment is currently established; combination therapy is usually the best approach. The major therapies consist of topical treatment, systemic agents, and phototherapy [1,4,5]. Topical treatments comprise topical corticosteroids and calcineurin inhibitors. Topical corticosteroids are the most commonly reported treatment, often used as the first-line therapy. Topical corticosteroids can reduce inflammation and pruritus but fail to alter the course of the disease. Complete remission using topical corticosteroids occurs in around 4% of patients, with partial remission in 80%. Complete remission is defined if over 90% of lesions disappear, and partial remission is defined if over 50% (but less than 90%) of lesions subside. No study reported the adverse effects or relapse rate of topical corticosteroids used in isolation as an intervention [1,4,5]. Our patient applied clobetasol propionate ointment to the erythematous papules. No side effect was reported during the treatment. Topical corticosteroids, in combination with other therapies, seem to give satisfying results.

Several systemic agents were proposed to manage PL, such as antibiotics (tetracycline, erythromycin, azithromycin), methotrexate, cyclosporine, pentoxifylline, acyclovir, pyrimethamine, and bromelain. The data on cyclosporine use in PL are limited to case reports. Lis-Swiety *et al.* reported a successful therapy of PLEVA preceded by hand, foot, and mouth disease using cyclosporine (3 mg/kg/day). Cyclosporine inhibits the activity of calcineurin, acting selectively on T cells. It reduces lymphocytes and macrophages in the epidermis and dermis and inhibits the activation of natural killer, antigen-presenting, and T cells. Cyclosporine can also prevent the proliferation of keratinocytes and the release of histamine from mast cells. In the case of PL preceded by a viral infection, cyclosporine may also play a role in antiviral activity. The use of cyclosporine with a maximum dose of 5 mg/kg should be given for up to one year only. The side effects of cyclosporine are mostly dose-dependent and related to the duration of treatment. Renal dysfunction, hypertension, gastrointestinal disturbances, headache, tremor, and hypercholesterolemia are among the reported side effects [5,8-10].

Phototherapy is the most successful therapy for PL, especially in cases of PLC. The phototherapy that can be used for PL consists of narrowband UVB (NB-UVB), broadband UVB (BB-UVB), PUVA, and UVA1. Phototherapy is considered a safe and effective treatment for PLC, and some authors recommend it as the first-line therapy for diffuse and recalcitrant forms. Complete remission using UVB, BB-UVB, NB-UVB, and PUVA was reported in 53%, 91%,

75%, and 69% of patients, and partial remission was reported in 32%, 0%, 20.6%, and 22%, respectively. NB-UVB seems to be the preferred phototherapy because of its safety profile. Fernandez-Guarino *et al.* reported a complete response rate of 88% in patients with diffuse PLC who showed no response to topical therapy. The mean sessions needed was 23 with a cumulative dose of 16.99 J/cm^2. Le Huu *et al.* also reported successful treatment of PLC with NB-UVB. A complete response was obtained in 82.8% of patients after a mean treatment period of 4.6 weeks (cumulative dose 9.8 J/cm^2). Short-term adverse effects included minimal erythema, pruritus, burning, tingling, folliculitis, headaches, and dryness. The most common adverse effect was erythema, ranging from mild to severe. No long-term adverse effects were reported. The relapse rate was reported at 25.7% [4,5,11,12]. Our patient obtained complete remission after eight sessions (four weeks) with a cumulative dose of 3.42 J/cm^2 NB-UVB phototherapy combined with cyclosporine. No side effect of phototherapy was observed in our patient. This combination therapy seems to hasten the duration of treatment.

No case of PLC treated with a combination of cyclosporine and NB-UVB was previously reported. In dermatology, phototherapy UVB has been used as a treatment in association with other therapy to limit the toxicity of systemic drugs and to reduce the cumulative dose of UVB. Franchi *et al.* successfully treated psoriasis using a combination of cyclosporine and UVB phototherapy. This therapeutic approach proves to shorten the use of full-dose cyclosporine and reduce the frequency of UVB phototherapy administration. The side effect reported was a mild increase of blood pressure in 12.5% of patients [13]. Current guidelines advise NB-UVB should be chosen as a first-line modality when considering phototherapy so that cyclosporine can be an option in future treatment. The use of cyclosporine should be avoided in patients with a high cumulative dose of previous UVA and psoralen. In such patients, cyclosporine may increase the likelihood of cutaneous cancers like squamous cell carcinoma [10].

CONCLUSION

Currently, there is no sufficient evidence to establish an algorithm for the treatment of PLC, and further studies are needed to assess treatment outcomes with an extended follow-up period. In our case, the combination therapy of NB-UVB phototherapy and cyclosporine may be a potential treatment for PLC that proves effective. Further research with longer follow-ups is necessary to confirm this combination therapy's efficacy and safety.

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None.

Authors contributions

Kara Adistri and Rhida Sarly Amalia proposed the case to be reported. Kara Adistri and Rhida Sarly Amalia drafted the manuscript. Windy Keumala Budianti critically revised the manuscript. All authors gave final approval and agreed to be accountable for all aspects of the work ensuring integrity and accuracy

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