NB-UVB is an expecting treatment for pityriasis rubra pilaris

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Summary

Pityriasis rubra pilaris (PRP) is an erythematous papulosquamous disorder characterized as a reddish or orange scaling dermatitis with normal skin islands, follicular hyperkeratosis, and keratoderma of the palms and soles. The pathogenesis of this disease and the best treatment remain unclear. Narrowband ultraviolet B (NB-UVB) therapy is one treatment for PRP, although there are few reports on it. Recently, we succeeded in controlling a case of PRP with NB-UVB and report this case with a review of the literature.

KEY WORDS: pityriasis rubra pilaris; adult; itching; NB-UVB; etretina.

Introduction

Pityriasis rubra pilaris (PRP) is a papulosquamous disorder of unknown etiology that often progresses to erythroderma and causes disabling keratoderma of the palms and soles (1). Treatments for this disease include retinoids, methotrexate, photochemotherapy [topical or systemic psoralen and ultraviolet A (PUVA)], topical emollients, keratolytics, and vitamin D3 ointment, although PRP has high spontaneous remission rates within 1-3 years. The effect of phototherapy is controversial. While ultraviolet B (UVB) does not seem to be helpful or can even exacerbate PRP (2-8). Here, we report a patient who was successfully controlled with NB-UVB therapy and review the literature on NB-UVB for treating PRP.

Case report

A 71-year-old Japanese man first developed scaly erythema on the upper half of his body 2 months before presentation. Subsequently, the erythema spread over his entire body. Oral cyclosporine (50 mg = 0.7 mg/kg/day) and topical steroid ointment were prescribed by a dermatologist under an initial diagnosis of psoriasis, but his eruption did not improve. He was referred to our Hospital in November 2013. His medical history included treatment for prostatomegaly, but no psoriasis. There was no family history of psoriasis. On physical examination, he had geographic scaly erythema on his entire body, including his face, which was very pruritic (Figure 1a, b). A biopsy from a scaly erythematous lesion on his waist revealed acanthosis, parakeratosis, and a perivascular lymphocytic infiltrate in the underlying dermis (Figure 1c, d). He was diagnosed with psoriasis vulgaris at this time, and we decided to increase the dose of oral cyclosporine to 150 mg (2.3 mg/kg/day). Despite continued oral cyclosporine and topical steroid ointment, his erythema spread. He was admitted for further treatment and to examine his status. We suspected a diagnosis of PRP based on the follicular hyperkeratosis, islands of normal skin on the trunk and limbs (Figure 2a, b), and hyperkeratosis with an orange hue on the palms and soles (Figure 2c, d). Laboratory tests showed increased levels of alanine aminotransferase of 60 U/L (normal 3-49 U/L) and blood urea nitrogen of 25 mg/dL (normal 7-21 mg/dL) and a reduced sodium level of 137 mmol/L (normal 139-149 mmol/L). Reexamination of his first biopsy specimen showed irregular parakeratosis, short, thick rete ridges, and a remaining granular layer. A second biopsy from a follicular hyperkeratosis lesion on the abdomen revealed dilating hair follicles and keratotic plugs but no perifollicular parakeratosis. Considering the physical examination, clinical course, and biopsy result, he was diagnosed with PRP. We discontinued the cyclosporine because of the widespread erythema and started etretinate 50 mg (0.75 mg/kg/day). Although his erythema improved gradually, the marked pruritus did not improve despite steroid ointment and several antihistamines. The dose was decreased to 30 mg at 2 months because paronychia developed after 1.5 months on etretinate, and it was discontinued after 2.5 months. No immediate exacerbation was observed with topical steroid ointment only. His erythema improved gradually after
Figure 1a-d - (a) Geographic scaly erythema on the patient’s face and (b) trunk. (c, d) A biopsy of the scaly erythema on the waist shows acanthosis, parakeratosis, and perivascular lymphocytic infiltrate in the underlying dermis [original magnification: (c) ×40, (d) ×400].

Figure 2a-d - At admission, the patient had expanded geographic scaly erythema, follicular hyperkeratosis, and islands of normal skin on the (a) trunk and (b) limbs and hyperkeratosis with an orange hue on the (c) soles and (d) palms.
etretinate was discontinued, but his pruritus did not improve. However, subsequently his eruption worsened again before phototherapy commenced. In addition to the topical steroid therapy, NB-UVB treatment three times a week was started; however, the palms and soles were excluded because few hyperkeratosis with an orange hue were noted on the palms and soles when NB-UVB was started. And there were no eruptions in the axilla and the intergluteal cleft from the start.

Some improvement in his erythema was seen 3 weeks later, and the pruritus improved rapidly after 2 months (Figure 3). The NB-UVB irradiation was decreased from three times a week to once a week after 2.5 months and to once every 2 weeks after another 6.5 months, without an exacerbation in the eruption or itching. When the NB-UVB treatment was reduced to once every 3 weeks, his eruption and itching worsened, so the NB-UVB therapy was resumed once every 2 weeks. The NB-UVB irradiation was discontinued at 17 months, because his eruption was minimal and was not exacerbated. The total UVB dose he received was 26.79 J/cm². Two months after discontinuing the UVB, some scaly erythema appeared spontaneously, but this was controlled with topical steroid ointment.

Discussion

Pityriasis rubra pilaris is an erythematous papulosquamous disorder characterized by follicular plugging, perifollicular erythema that becomes confluent, palmoplantar hyperkeratosis, and pityriasis capitis (9). PRP has been classified clinically into 5 types: types I and II are characterized by adult onset and types III to V by juvenile onset (10). Recently, a new category, type VI, was proposed, characterized by the presence of HIV infection (1, 9, 11). As the major type in adults, type I or classical adult-onset PRP affects over 50% of patients and is characterized by spontaneous remission rates of 80% within 1-3 years (11). Clinically, the erythroderma manifests as islands of normal skin and follicular hyperkeratosis and waxy diffuse palmoplantar keratoderma. Type II, atypical adult-onset PRP, affects about 5% of patients. It is characterized by a duration of 20 years or more. The clinical manifestations include ichthysisiform scaling on the legs, eczematous change, areas of alopecia, and palmoplantar keratoderma with lamellated scales (1, 10).

The difference between PRP and psoriasis, especially during the early phase of the disease, is a subject of discussion (1). This makes the diagnosis of PRP difficult, and the diagnosis needs to be based on physical examination, clinical course, and biopsy results. Pityriasis rubra pilaris is often resistant to both topical and systemic therapies (10). First-line therapy for PRP includes topical therapy with emollients, keratolytics and vitamin D3, physical therapy with phototherapy (topical and systemic PUVA), extracorporeal photopheresis, and systemic therapy with retinoids and methotrexate (1). Second-line therapy includes topical therapy with glucocorticoids and vitamin A analogs, physical therapy with UVA1, NB-UVB, or UVB phototherapy, and systemic therapy with azathioprine, cyclosporine A, fumaric acid esters, and tumor necrosis factor (TNF)-α antagonists (1).

We chose cyclosporine because our initial diagnosis was psoriasis. The cyclosporine had little effect, even though the dose was increased to 2.3 mg/kg. Consequently, we reevaluated his status and changed our diagnosis to PRP based on the clinical course, physical findings, and biopsy result. We tried etretinate next, and his eruption improved. Unfortunately, we...
Pityriasis rubra pilaris

Table 1 - Summary of reported phototherapies for PRP.

<table>
<thead>
<tr>
<th>Case</th>
<th>sex</th>
<th>type</th>
<th>irradiation interval</th>
<th>effective</th>
<th>total irradiation period</th>
<th>total dose</th>
<th>with retinoid</th>
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<tr>
<td>1</td>
<td>F</td>
<td>III</td>
<td>three times a week</td>
<td>12</td>
<td>26 treatments</td>
<td>10 months</td>
<td>Yes, 0.5mg/kg/day</td>
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<td>2</td>
<td>F</td>
<td>III</td>
<td>9 treatments</td>
<td></td>
<td>45 treatments + or</td>
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<td>3</td>
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<td>3</td>
<td>F</td>
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<td>4</td>
<td>F</td>
<td>III</td>
<td>within 1 year</td>
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<tr>
<td>5</td>
<td>M</td>
<td>I</td>
<td>three times a week</td>
<td>2</td>
<td>4 months</td>
<td>5,650 J/cm²</td>
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<tr>
<td>6</td>
<td>F</td>
<td>IV</td>
<td>three times a week</td>
<td>2</td>
<td>2 months</td>
<td>1,720 J/cm²</td>
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<td>7</td>
<td>F</td>
<td>BB/IV</td>
<td>2 months, 19 sessions</td>
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<td>105 J/cm²</td>
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<tr>
<td>8</td>
<td>M</td>
<td>III</td>
<td>three times a week</td>
<td>3 weeks</td>
<td>17 months</td>
<td>26.79 J/cm²</td>
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<tr>
<td>1</td>
<td></td>
<td></td>
<td>every third day</td>
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<td>122.6 J/cm²</td>
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<td>2</td>
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<td>yes, 50mg/day</td>
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had to discontinue the etretinate shortly, because he developed severe paronychia. In addition, it did not improve his itching. His eruption improved gradually after etretinate was discontinued. However, his eruption worsened again soon after that, so we started to treat him with phototherapy. Therefore, we considered that NB-UVB therapy, not a delayed response to retinoid therapy, was effective in our patient.

The mechanism of phototherapy in psoriasis is the induction of T-cell apoptosis and regulatory T cells and a decrease in the production of IFN-gamma (12-14). The mechanism of UVB phototherapy in itching is a decrease in the number of nerves that release histamine from mast cells and interferon gamma (15). Therefore, we considered phototherapy to be effective for eruption and itching in PRP patients, too.

In PRP, phototherapy might act via a similar mechanism because the histopathology of PRP resembles that of psoriasis; however, the effect of phototherapy in PRP is still debated. Some cases have responded well to PUVA (1, 16-21), which is sometimes combined with retinoids or methotrexate (1, 17). One case responded successfully to bath PUVA therapy, despite his photosensitivity to UVB (20). Although ultraviolet B irradiation is effective for psoriasis, it has not been helpful or can even worsen PRP (1). A case provoked by UVB 4 days after irradiation was reported, and generalized PRP developed subsequently (20). In comparison, 7 patients were treated successfully with NB-UVB (2-8). Only one case of successful treatment with BB-UVB has been reported (23). However, in recent years, NB-UVB has replaced BB-UVB in most treatment facilities as it has been shown to be more effective for psoriasis (24). Table 1 summarizes the reported results of phototherapy. Few reports give the details of the phototherapy performed. In comparison, our report describes the phototherapy administered in the greatest detail.

In PRP, PUVA therapy is included as first-line therapy, while UVB therapy is regarded as second-line therapy because ultraviolet exposure reportedly aggravates PRP (1, 18, 25, 26). In addition, 26% of 57 PRP patients developed exacerbations in the summer (27). Considering these reports, we wanted to administer more phototherapy (bath PUVA), but the patient requested discharge because of his already long hospital stay. Therefore, we treated him with NB-UVB phototherapy as an outpatient three times a week. Our patient had no exacerbations with NB-UVB therapy. His pruritus disappeared after 2 months of NB-UVB irradiation, after steroid ointment and several antihistamines were ineffective. Although PRP has a high spontaneous remission rate within 1-3 years, we believe that our patient responded to NB-UVB and did not undergo spontaneous remission because his eruption and itching worsened when the NB-UVB treatment was reduced from once every 2 weeks to once every 3 weeks, and his eruption and itching improved after NB-UVB once every 2 weeks was resumed.

NB-UVB phototherapy can be administered to outpatients, making it an option for intractable PRP, but only for patients who have no exacerbation with ultraviolet rays. Similarly, NB-UVB might be an option for treating intractable pruritus, based on our experience with this case. Further reports on the treatment of PRP, particularly phototherapy, are needed to determine the optimum management of this disease.

References


