

Pustular Palmoplantar Psoriasis Successfully Treated with Nb-UVB Monochromatic Excimer Light: A Case-Report

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Abstract

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Barber's palmoplantar pustulosis (PPP) is a form of localised pustular psoriasis, affecting the palmar and plantar surfaces. It is a chronic disease, with a deep impact on the patients' quality of life. The Authors discuss a case of Barber Psoriasis successfully treated with monochromatic excimer light.

Introduction

Pustular palmoplantar psoriasis is a peculiar form of localised pustular psoriasis occurring on palmar and plantar surfaces of the skin (type Barber or Barber-Königsbeck). It is a chronic and stubborn pathology, difficult to manage and with the tendency to recur over time [1]. In contrast to plaque-type psoriasis, pustular psoriasis shows homozygous or compound heterozygous interleukin-36 (IL36RN) gene mutations leading to aberrations in IL-36R antagonist function [2]. The immunological peculiarities of pustular psoriasis contribute to the less favourable outcome of this psoriasis type with tumor-necrosis factor-alfa antagonists. Narrow-band ultraviolet-B phototherapy (308-311 nm) has been recognised as a valid therapeutic option for the disease, leading to quick remissions and low recurrence rates. In recent years, devices able to deliver the radiation only to

affected areas of the skin have been developed, with the major advantages of sparing unaffected areas of the body surface and being more compliant for the patients [3].

Case Presentation

An otherwise healthy male subject, 38 years old, affected by a pustule palmar psoriasis (Fig. 1), showed up to our Clinic presenting, on his palms, numerous small flat pustules (2-3 mm in diameter), whitish to yellowish in colour, on an erythematous basis. He lamented a slight, continuous, burning sensation. He reported the appearance of these lesions about two months before, at first on his left palm, and then rapidly spreading to involve also the

right palm. He also described how new pustules appeared in a few hours, while the older ones were creating a yellowish crust which was falling spontaneously.

History for drug assumption was negative, and he didn't report any contact with local irritants. The patient didn't refer any revealing pathology, and he showed no familiarity for psoriasis or other skin diseases.



Figure 1: Our patient affected by pustular psoriasis on his palms

During the clinical evaluation, no other lesions were observed in any other part of the body, apart from small, hurting fissurations on the flexural surfaces of the wrists, while fingers were not affected. Nails showed no sign of psoriatic lesions. A rheumatologic evaluation showed no apparent joint involvement. Routine blood testing for inflammation and infections was negative.

A punch biopsy performed on a pustular area showed subcorneal unilocular pustulosis and a neutrophilic infiltration with neutrophilic exocytosis, configuring the classical aspect of "spongiform pustule of Kogoj". Moreover, the cultural test showed sterile pustules, confirming the initial diagnosis of palmoplantar psoriasis (Barber's palmoplantar psoriasis). Narrowband UVB (308-311 nm) is considered by dermatologists worldwide an efficient treatment for psoriasis [1]. For this reason (and according to our positive personal experience), after having discussed with the patient about the therapeutic possibilities, and after having received his informed consent, we decided to perform a focused narrowband UVB treatment, using a monochromatic excimer light device developed to deliver a 308 nm radiation only to the affected areas, thus sparing the unaffected surrounding skin. The device emits UVB light with the wavelength of 308 nm, with a power density of 50 mW/cm² at a distance of 15 cm from the skin, and a maximum radiation area of 512 cm².

The therapeutic procedure schedules two sessions in a week for the first four weeks of treatment, and then one session in a week for the rest of the treatment. During the first session, we

estimated the MED (Minimal Erythema Dose) of the patient in an uninjured part of the body (flexural part of the forearm), to be able to set the first dose of radiation, corresponding to the MED decreased by 10%. During the following sessions, we proceeded increasing the dose gradually depending on the clinical response. Regarding time (seconds of irradiation), our patient needed 15 seconds of illumination for the first session (0.75 J/cm²); the duration of the following treatments was increased by 5 seconds (0.25 J/cm²) each time.

At week 6 (10 treatments), the clinical picture was substantially improved, showing a clear reduction of inflammation, erythema, desquamation, and the complete disappearance of pustules. We decided to interrupt the treatment at week twelve (16 total sessions) since we evaluated the complete resolution of the disease (Fig. 2).



Figure 2: The same patient after the treatment with excimer light

During the entire treatment time, no side effects or adverse events were noted. Only a slight, transient erythema appeared after each of the first four sessions, however leading to no harms or discomfort for the patient. Since treatment discontinuation, we are monitoring the subject to evaluate the possible recurrence of psoriasis. However, after 12 weeks without any treatment, no lesions are present to date.

Discussion

Psoriasis is a chronic inflammatory disease involving the skin, and sometimes affecting joints, bones, tendons, ligaments, nails, and mucosal membranes [1]. About 3% of the Italian population is affected by psoriasis, and this percentage reflects the worldwide prevalence of the disease [4].

Psoriasis has many clinical variants, the most common of which is the vulgar type (plaque-type), guttate, inverse, mucosal, sebopsoriasis, pustular,

arthropathic, erythrodermic. Pustular psoriasis can be divided into two major clinical variants, localised and generalised, both characterised by the same elementary lesion, a sterile, non-follicular, superficial pustule, but differentiated, among the others, by the degree of body surface involvement. Its histopathology is characterised by the “spongiform pustule of Kogoj”, that is a suppurative pustule located in the upper part of spinosus layer and filled with neutrophils [5].

By the localisation of the lesions, and depending on their different evolution, two main variants of localised pustular psoriasis may be distinguished, namely the Barber’s palmoplantar pustulosis (PPP) and the Hallopeau’s continuous acrodermatitis. PPP is the most common one, and it is characterised by the outbreak of little pustules on an erythematous and scaly basis, located on the palms of the hands and the soles of the feet. Pustules arise by rushes in a few hours, are 1 to 5 mm in diameter, and are surrounded by an erythematous ring. Usually, they are asymptomatic, but they can cause a burning sensation. They are flat-topped, whitish to yellowish with the tendency to become darker until they dry in a couple of days, leaving a crust and a brownish hyperpigmentation. Rushes are rapidly arising in the acute phase of the disease so that it is possible to observe in lesions in several stages of maturation. Going ahead with the disease, inflammation and erythema may spread to the fingers and the wrists, and painful rhagadiform lesions may appear.

PPP mainly occurs in middle-aged persons, more often females, even if we can’t rule out a more premature exordium in child’s age. Even if in around half of the patients, the disease can start with unilateral lesions, it is any way inclined to be symmetrically present on the extremities. The presence (contemporary or diachronic) in the same subject of pustular lesions together with lesions peculiar of other forms of psoriasis is not unusual, even if the real incidence of this association is still discussed [6, 7].

PPP can be associated with osteoarthritis, in a multifocal recurrent chronic form (CRMO) [8] or the sternocostoclavicular hyperostosis (SCCH), in this last case configuring the so-called SAPHO (Synovitis, Acne, Pustulosis, Hyperostosis, Osteomyelitis) syndrome [9].

The diagnosis of PPP is essentially clinical, possibly held up by laboratory studies; though these may be usually insignificant (there is the possibility of a transitory neutrophilic leucocytosis). Histopathology of the lesions shows different aspects according to the stage of the disease. In the initial phase, intraepidermal vesicles containing mononuclear cells may be seen, caused by focal spongiosis. With the evolution of the lesion, neutrophils can be seen inside the vesicles, leading to the development of the typical subcorneal pustules in the latter phase. Usually, a

cultural test of the pustulosis gives a negative result (sterile pustules). Many therapeutic options are nowadays available for PPP; however, none of them can be addressed to as the gold standard treatment [10].

Topical high potency corticosteroids, alone or in association with salicylic acid or vitamin D analogues, are still considered the best treatment for PPP [11]. This approach is effective in a large percentage of patients, yet doesn’t seem to possess the capability for a long-term control of the disease doesn’t prevent the recurrences, and the event of tachyphylaxis and side effects are major contraindications for its continuous use [12]. Better results can be obtained with systemic corticosteroids [13] and with oral acitretin [14], but the possible side effects, and the worsening of psoriasis that can be experienced early after their interruption makes them desirable only for particularly selected cases. Oral colchicine [15] leads to good results, but its use is limited by side effects like diarrhoea and nausea. Systemic cyclosporine A has proven efficacy in the control of the disease [16], but quick relapses after its interruption and the well-known collateral effects connected by the intrinsic characteristics of the drug (arterial hypertension, renal insufficiency, nausea and tiredness) limit its use [17]. A few studies quest for the possibility to recur to local administration of methotrexate-based gel [18], but the experience is still anecdotic.

In recent years, the new biological therapies (monoclonal antibodies, receptor fusion proteins and similar) have been developed to manage psoriasis in its inner mechanisms of immune regulation. In the clinical experiences with these new generation drugs, there are some reports of PPP successfully treated with efalizumab, alefacept, infliximab, adalimumab, and etanercept [10]. However, the legal (i.e. FDA and EMA-approved use of biologics in psoriasis) and scientific limitations (i.e. guidelines) for the use of biologics, together with the major possible medical involvements connected to this category of therapeutic agents, make of them a treatment dedicated to a few well-chosen patients and it is off-label.

Among the physical therapies, PUVA therapy, probably in association with oral retinoids (RePUVA) [19] has been used for pustular psoriasis.

The efficacy of UVB light in psoriasis has been largely demonstrated (10, 11, 19) so that nowadays UVB treatment may be considered the first-line treatment in many forms of psoriasis. UVB treatments can be safely used in pregnant women and children, and are related to less erythema in respect of UVA, no phototoxic effects and no epidermal thickening after long-term irradiation. No statistical differences have been shown between PUVA and UVB regarding success rates [19].

However, during the last years focused phototherapy with narrowband UVB (307-311 nm) showed a similar efficacy, but without a risk of secondary skin cancer development [21-24]. This therapeutical approach considers that psoriasis patients undergoing phototherapy usually receive high cumulative doses of radiation during their lives, thus leading to secondary cutaneous disorders, like photoaging, teleangectasias, excessive tanning, etc. On the contrary, a phototherapeutic device capable of delivering the UV-radiation only to affected areas could lower all these collateral effects dramatically decreasing the total dose of radiation. Moreover, the treatment may be tailored to each affected area with different doses of UV.

The monochromatic excimer light (MEL) device delivers a UVB wavelength at 308 nm only to the lesional skin, and seems particularly effective for variants of psoriasis where the involvement of the skin is lower than 20-25% of the total body surface. Its high potency (up to 4.5 J/cm²) and subsequently the need for short time of irradiation, together with the possibility to schedule just 1 session per week, makes MEL mostly appreciated to a large part of patients, thus increasing treatment compliance, which is particularly useful when we have to deal with long-lasting therapeutic protocols. Finally, the possibility to focus the radiations on skin lesions reduces the risks of acute and chronic side effects in the uninvolved safe skin.

In conclusion, our experience scores another point in favour of focused narrowband UVB treatments for localised psoriasis, showing once more its efficacy, relative rapidity of action and safety. Moreover, in our opinion, the efficacy of MEL on localised pustular psoriasis, a challenging clinical picture, which management is often difficult both for the patient and the dermatologist, makes it one of the best weapons we have to fight, and win, against this disease.

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