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Case Report

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Refractory Pityriasis rubra pilaris treated with Adalimumab: a case report

Abstract

Pityriasis Rubra Pilaris (PRP) is a rare inflammatory dermatosis, clinically presenting as a keratinization disorder, with follicular and palmoplantar hyperkeratosis, in addition to orange-red scaly plaques, accompanied by characteristic islands of healthy skin. Sometimes, conventional treatment proves to be ineffective, and this work aims to present a case of a patient with failed first and second lines treatment options, responding well to adalimumab. Despite of all options available, more qualified and specific studies are needed to establish a standardized treatment based on evidence of a high scientific level, thus raising the degree of efficacy and therapeutic safety in PRP.

Keywords: pityriasis rubra pilaris, adalimumab, skin diseases, retinoids, dermatologic agents

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Abbreviations: PRP, pityriasis rubra pilaris

Introduction

Pityriasis Rubra Pilaris (PRP) is a rare inflammatory dermatosis, of still unknown etiology, which can present genetic inheritance or be acquired, equally affecting men and women. It can be classified, according to Griffith, into five types, with the classic adult form (type I) being its most common form. It presents clinically as a keratinization disorder, with follicular and palmoplantar hyperkeratosis, in addition to orange-red scaly plaques, accompanied by characteristic islands of healthy skin. Sometimes, conventional treatment proves to be ineffective, and this work aims to present a case with a new therapeutic option.[1–3]

Case report

Male patient, 42 years old, mechanic, previously healthy, with sudden appearance of diffuse erythematous scaly plaques on the scalp, with progression within 8 weeks to the face and trunk. To the clinical examination, presents with irregular, well-delimited erythematous-orange plaques, with furfuraceous and follicular desquamation, interspersed with healthy skin, affecting the scalp, face and trunk top. He also had involvement of palms and soles with yellowish hyperkeratotic plaques (Figure 1).

A biopsy was performed with findings compatible with the clinical hypothesis of PRP: parakeratosis alternating with orthokeratosis, acanthosis with short and wide epithelial cones, mild lymphocytic infiltrate perivascular, in addition to infundibular dilation. The patient did not respond to topical treatment with moisturizers and corticosteroids, then multiple attempts at systemic therapy with prednisone 40mg/day, acitretin 50mg/day, isotretinoin 40mg/day and methotrexate 25mg/week were performed, with no satisfactory response.

Therefore, an anti-TNF alpha immunobiological agent, adalimumab (80mg loading dose in week 0 and 40mg in week 1, followed by 40mg every 2 weeks), was prescribed in combination with methotrexate, resulting in significant clinical improvement after 20 days. The patient was maintained with the association of treatment for nine months, when the progressive withdrawal of methotrexate was chosen, without recurrence of the lesions. Today, 9 years after his diagnosis, he has no active lesions and has proposed de-escalation of the adalimumab. Figure 2

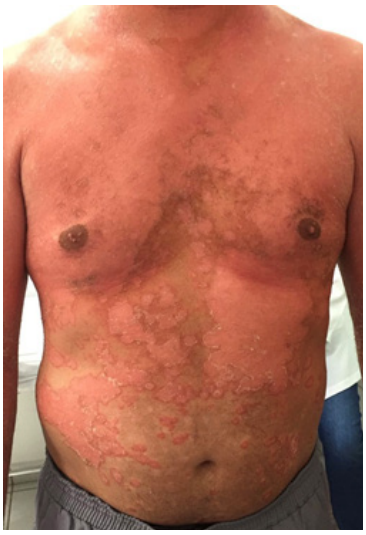

Figure 1 Before the treatment with adalimumab.

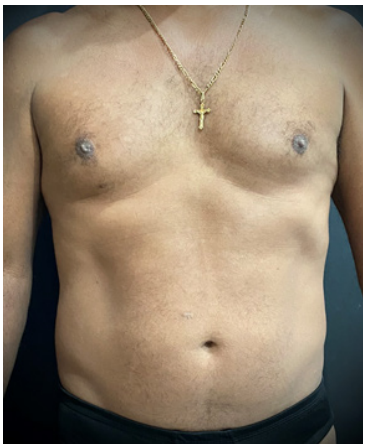

Figure 2 One year after the begnning of the treatment with adalimumab.

Results and discussion

PRP is an important cutaneous pathology that has a good prognosis in the vast majority of cases, with whitening of the lesions in up to 3 years. However, there are reports in literature that the disease may persist for longer than two decades. Its treatment involves, at first, topical medications, such as emollients, calcipotriene, retinoids, keratolytics and topical corticosteroids. In case of persistent and more severe disease, the systemic treatment options are retinoids (acitretin, isotretinoin, etretinate), methotrexate, cyclosporine and phototherapy. Immunobiological agents are currently being described as an option for the third line of treatment, with cases showing satisfactory response to TNF-alpha or IL-17 inhibitors.[4–6]

Conclusion

Despite the described treatments, more qualified and specific studies are needed to establish a standardized treatment based on evidence of a high scientific level, thus raising the degree of efficacy and therapeutic safety in PRP.

Acknowledgments

None.

Conflicts of interest

Authors declare there is no conflict of interest.

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