SunKrist Publishing
Accelerate Scientific Knowledge in Open Access

SunKrist Journal of Dermatology and Skin Diseases

Case Report *Volume: 2, Issue: 1* *Scientific Knowledge*

Treatment Approach to Erythrodermic Pityriasis Rubra Pilaris: Case Report

Mori A[1], Sacco DM[1], Sandoval JD[1] and Oporto JI[2*]

[1]Escuela de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile

[2]Fundación Oftalmológica Los Andes, Universidad de los Andes, Santiago, Chile

1. Abstract

Pityriasis rubra pilaris is a rare inflammatory dermatosis that tends to progressively evolve to erythroderma. Finding the appropriate treatment is challenging because there are no large scale studies or international guidelines available. Traditionally, the hydration of the patient is essential, as are various topical and systemic therapeutic alternatives that can be used, with highly heterogeneous results. The objective of this report is to present a clinical case of erythroderma due to pityriasis rubra pilaris in an adult and to illustrate the process of choosing the most appropriate treatment according to the patient's response.

2. Keywords: Pityriasis rubra pilaris, Erythroderma, Treatment

Abbreviations:

PRP Pityriasis rubra pilaris; TNF Tumor Necrosis Factor; IL interleukin; HIV Human Immunodeficiency Virus; HBV Hepatitis B Virus; HCV Hepatitis C Virus; TB tuberculosis.

3. Introduction

Erythroderma is an inflammatory skin syndrome, which involves erythema and scaling of more than 90% of the body's surface. It can represent the final stage of multiple pathologies, such as psoriasis, eczematous conditions, drug-induced reactions, neoplasms, and pityriasis rubra pilaris (PRP). The diagnostic approach should include a thorough medical history and physical examination. If the etiology is still unknown, it may be necessary to perform skin biopsies to obtain an accurate diagnosis [1, 2].

Specifically, pityriasis rubra pilaris (PRP) is a rare inflammatory dermatosis that can lead to erythroderma, which tends to present with a craniocaudal progression. It is macroscopically characterized by hyperkeratotic follicular papules surrounded by peri and interfollicular orange erythema, and erythematous scaly plaques. The presence of islands of spared skin between the affected areas is pathognomonic of PRP [3]. Histologically, it presents epidermal acanthosis, diffuse orthokeratosis, and dotted parakeratosis in both vertical and horizontal directions, as well as lymphohistiocytic inflammatory infiltrate on the perifollicular and perivascular surface of the dermis

***Corresponding author:** Joaquín-Ignacio Oporto, Fundación Oftalmológica Los Andes, Universidad de los Andes, Las Hualtatas 5951, Vitacura, Santiago, Chile, Tel: 223704600, Email: jioportoc@gmail.com

Received Date: June 30, 2021; **Accepted Date:** July 1, 2021; **Published Date:** July 6, 2021

[4].

The initial therapeutic approach of any patient with erythroderma, regardless of its cause, should include the nutritional evaluation of the patient, a correct hydro-electrolyte balance, measures to maintain the skin's barrier function, antihistamines to manage pruritus and the exclusion of concomitant bacterial infections [1].

Regarding the treatment of PRP, the low prevalence of the disease and the consequent lack of prospective clinical studies and international guidelines make therapeutic management a challenge [3]. Traditionally, retinoids such as isotretinoin, immunosuppressants such as azathioprine, methotrexate, and cyclosporine have been used for systemic treatment, alone or in combination with phototherapy, with high heterogeneity in responses [4]. New evidence suggests the use of biologics such as anti-tumor necrosis factor (TNF) -α, anti-Interleukin (IL) -12 / 23-, anti-IL 23, and anti-IL17A agents as treatment for both refractory cases as well as a first-line alternative [5].

In Chile there is no guide that provides a management algorithm for this pathology, making each case an individualized process in search of the optimal treatment for the patient. This situation affects patients since it increases both mortality and associated morbidity [6].

In this report we present the clinical case of a 69-year-old man with an erythroderma due to PRP that evolves with no response to conventional first-line treatment and the need for hospitalization, in order to discuss the scope of the expanding scenario of therapy for the PRP.

4. Case Report

A 69-year-old male patient, with no relevant past medical pathologies, presented to the emergency department with a 6-month history of generalized and progressive desquamation. The patient reported having started 7 years ago with pruritic scaly erythematous plaque-type lesions distributed on the face and scalp, which progressed caudally until involving the entire body's surface. He had recently consulted with a dermatology team, where he was diagnosed with erythroderma of unspecified etiology. A skin biopsy was performed and symptomatic management with topical corticosteroids was initiated. The histopathological analysis of the sample revealed evidence of psoriasiform and spongiotic dermatitis, with multifocal parakeratosis; PRP-compatible alterations with signs of grating. Treatment was adjusted, maintaining topical therapy and adding systemic corticosteroids (prednisone) along with systemic retinoids (isotretinoin).

The patient evolved with no response to treatment. Upon physical examination, generalized erythema, erythematous-scaly plaques, palmoplantar hyperkeratosis and onychogryphosis were noticed

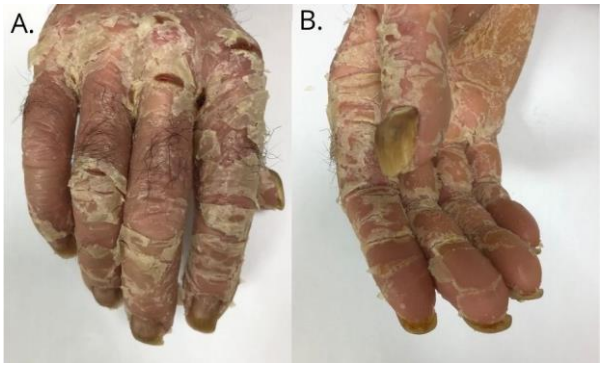

Figure 1: (A) Severe erythematous-scaly plaques, palmar hyperkeratosis, fissures in the skin. (B) Onychodystrophy and onychogryphosis in his fingernails.

He was hospitalized to optimize his treatment given cutaneous stiffness that limited movement, painful fissures in the skin and poor thermoregulation, associated with ocular compromise characterized by blepharitis and ectropion

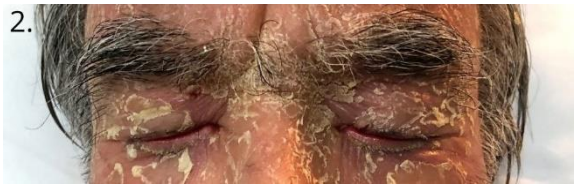

Figure 2: Eye compromise characterized by blepharitis and ectropion.

During his stay, general measures were intensified, with special care in the hydration and thermoregulation of the patient. His usual moisturizing lotion was replaced by solid petroleum

jelly, which achieved greater flexibility of the skin and comfort.

Therapeutic alternatives were discussed with the internal medicine and dermatology teams. The patient was receiving isotretinoin along with prednisone, both in doses of 20 mg per day. Given that until now he had presented null response to conventional systemic treatment, it was decided to escalate to immunosuppressive therapy with cyclosporine. Before starting the new therapy, an exhaustive study of possible coinfections was carried out, specifically for HIV, HBV, HCV and TB, obtaining negative results for all of them.

It was decided that the patient was a candidate to start treatment with cyclosporine at a dose of 150 mg every 12 hours and the dose of prednisone was doubled. After adjusting the therapy, he remained hospitalized for 5 days with serial monitoring of kidney and liver functions, and electrolyte levels, presenting a good response to the new drug. Therefore, he was discharged with outpatient follow-up.

Two weeks after discharge, he went to his dermatology check-up, where a decrease in desquamation, less limitation of movement and a clear improvement in thermoregulation was observed.

5. Discussion

Erythroderma is the clinical finding of erythema and scaling that involves more than 90% of the skin. It has a variable epidemiology, approximately 9.4 cases per year were reported in a retrospective study from Portugal [7]. Locally, it has been quantified at 21.7 cases per 100,000 dermatological consultations [8]. Excluding the pediatric population, the age of presentation usually varies between 41 to 61 years, being usually over 45 years of age. A higher prevalence in men has also been described, with a ratio of 2-4 to 1 [1].

Low associated mortality has been reported in studies in recent decades, probably secondary to advances in hospital management and new therapeutic alternatives. Despite this, a retrospective study from 2014 in Denmark reported that 39.6% of patients with erythroderma died within the first 3 years after being hospitalized [2].

The therapeutic challenge posed by erythroderma as a syndrome is that it can be caused by multiple diseases and external factors. Among the etiologies, one of the rare ones is PRP, which is a papulosquamous and inflammatory skin condition, with highly varied clinical characteristics. The definitive diagnosis is obtained with a skin biopsy that presents compatible histopathological findings [3].

The treatment of PRP is initially similar to any erythroderma, with emphasis on the patient's hydro-electrolyte support [1]. Currently there is no established algorithm for its treatment, and its development will depend on small series of cases, reports and opinions based on experience, since no randomized clinical studies on this pathology have been published. [3]

Firstly, for symptomatic relief, emollients, keratolytics and systemic antihistamines have been used with good reported results [9]. However, systemic therapies constitute the main pillar of management, with highly heterogeneous results. Among the most used alternatives are retinoids, corticosteroids, immunosuppressants and biologics [3].

Oral retinoids are often used as first-line agents, such as acitretin and isotretinoin. Based on multiple small case series, a significant clinical response could be obtained in 4 to 25 weeks regarding degrees of erythema, induration, and scaling [9, 10].

Refractory cases that have received retinoids or with contraindications for their use can benefit from the use of immunosuppressive drugs such as methotrexate, azathioprine and cyclosporine. The most widely used is methotrexate, with a response rate of approximately 39% [3]. On the other hand, phototherapy, either alone or in combination with another treatment, represents an alternative for disseminated disease. [3]

In case of poor response to the treatments already described, there is also the option of using biological therapies such as inhibitors of TNF-a, IL12, IL-23p40, IL-17A, given the relationship existing both at the molecular and clinical level between PRP and psoriasis. Despite the risks and high cost that these entail, their use has become more common [5].

Regarding the patient presented, this man was affected by a severe form of PRP without compromise of other organs or relevant comorbidities, so in congruence with the therapeutic measures already mentioned, he was prescribed oral retinoids and corticosteroids. Due to his refractoriness to treatment, during the hospitalization it was decided to initiate immunosuppressive therapy with cyclosporine, observing a favorable response to the pharmacological adjustment.

It is important to emphasize that the therapeutic options depend on the local reality. For example, in this case despite the severity of the case, immunosuppressants or biological were not used as first line agents due to the cost associated with them for this type of rare pathologies.

In conclusion, a case of PRP erythroderma successfully managed with immunosuppressants is presented with the aim of showing the importance of choosing the appropriate treatment according to the severity of the disease and the patient's comorbidities. Although retinoids are still considered one of the mainstays of treatment, immunosuppressants and biologics have progressively gained attention for their mechanism of action and effectiveness. In any case, the development of therapeutic guidelines and a structured algorithm based on large randomized clinical studies is still essential for a standardized approach.

Conflicts of interest

The authors declare no conflict of interest.

6. References:

1. Shim TN, Berth-jones J. Erythroderma. In: Lebwohl M, Hey-mann W, Berth-Jones J, Coulson I. Treatment of skin disease: Comprehensive therapeutic strategies. Saun-ders; 2014; 234-238.
2. Egeberg A, Thyssen J, Gislason G, Skov L. Prognosis after hospitalization for erythroderma. Acta Derm Venereol. 2014; 7: 959-962.
3. Roenneberg S, Biedermann T. Pityriasis rubra pilaris: algorithms for diagnosis and treatment. J Eur Acad Dermatol Venereol. 2018; 32: 889-898.
4. Wang D, Chong VC, Chong WS, Oon HH. A review on pityriasis rubra pilaris. Am J Clin Dermatol. 2018; 19: 377-390.
5. Napolitano M, Abeni D, Didona B. Biologics for pityriasis rubra pilaris treatment: a review of the literature. J Am Acad Dermatol. 2018; 79: 353-359.
6. Kamarachev J, Grozdev I, Darlenski R, Tsankov N. Pityriasis rubra pilaris as a systemic disease. Clin Dermatol. 2019; 37: 657-662.
7. César A, Cruz M, Mota A, Azevedo F. Erythroderma. A clinical and etiological study of 103 patients. J Dermatol Case Rep. 2016; 10: 1-9.
8. Vargas-Mora P, Valenzuela-ahumada F, Muñoz-alvear P, Morales Huber C. Estudio clínico-histopatológico de las eritrodermias en un hospital universitario: 14 años de experiencia. Rev chil dermatol. 2019; 35: 90-94.
9. Klein A, Landthaler M, Karrer S. Pityriasis rubra pilaris: a review of diagnosis and treatment. Am J Clin Dermatol. 2010; 11: 157-170.
10. Dicken CH. Treatment of classic pityriasis rubra pilaris. J Am Acad Dermatol. 1994; 31: 997-999.

Citation: Mori A, Sacco DM, Sandoval JD. and Oporto JI. Treatment Approach to Erythrodermic Pityriasis Rubra Pilaris: Case Report. SunKrist Journal of Dermatology and Skin Diseases. 2021; 2: 1002.