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Late onset pityriasis rubra pilaris type IV treated with low-dose acitretin

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Abstract

Pityriasis rubra pilaris is a chronic inflammatory dermatosis of unknown etiology and great clinical variability. It has been divided into six categories. Types III, IV, and V occur in childhood and are distinguished by their clinical presentation, age of onset, and course. We report a 19-year-old male patient with a 2-week history of pruritic, scaling dermatosis of the hands, feet, elbows, and knees. He had no family history of skin disease. On physical examination, we observed circumscribed, reddish-orange, scaling plaques affecting the elbows and knees and a waxy palmoplantar keratoderma. The skin biopsy showed acanthosis, alternating orthokeratosis, parakeratosis, and follicular plugging suggestive of pityriasis rubra pilaris. The patient started treatment with oral acitretin, 25 mg every other day. The treatment was tolerated well, and after 6 months the lesions had resolved completely. Pityriasis rubra pilaris is a chronic papulosquamous disorder of unknown pathogenesis, characterized by reddish-orange scaly plaques, palmoplantar keratoderma, and keratotic follicular papules. There is still no consensus regarding the treatment, but therapeutic options include systemic retinoids, particularly acitretin in the recommended dose of 0.5 to 0.75 mg/kg/day. In our case, the patient was treated with a low-dose regimen of acitretin, which was effective and well tolerated.

Keywords: Pityriasis rubra pilaris, treatment, acitretin

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Introduction

Pityriasis rubra pilaris (PRP) is a rare inflammatory dermatosis of unknown origin and considerable clinical heterogeneity (1). It has been divided into six types, each with a particular clinical presentation, age of onset, and course (2). Type IV, or the circumscribed juvenile form, accounts for approximately 25% of cases of PRP (2). It usually occurs in prepubertal children and its main features are hyperkeratotic perifollicular papules forming sharply demarcated erythematous plaques, localized to the elbows, knees, and dorsal aspects of the hands and feet. An orange palmoplantar keratoderma is frequently present (3). Due to the clinical variability observed in this disease, the treatment is challenging and the response is variable, making it difficult to predict the outcome of the treatment. A standard therapeutic approach does not exist because the cases are few and treatment is protracted. Retinoids are among the most frequently used medications and have variable effectiveness (1).

Case report

A 19-year-old male patient was observed in our department due to a 2-week, mildly pruriginous eruption affecting his elbows, knees, and palmoplantar regions. He mentioned taking ibuprofen 400 mg for sore throat 1 week before the onset of the dermatosis. His past medical record was otherwise irrelevant and he had no family history of other skin diseases, including psoriasis or ichthyosis.

Upon physical examination, the patient had circumscribed, well-defined, scaling salmon plaques, distributed symmetrically on the elbows, knees, and dorsal aspects of the hands and feet, along with an orange keratoderma (Fig. 1a–c). No mucosal lesions (specifically, in the buccal mucosa) or nail changes were found.

A skin biopsy was performed, revealing alternating orthokeratosis and parakeratosis, confluent hypergranulosis, and follicular plugging, compatible with the clinical diagnosis of pityriasis rubra pilaris. The patient's weight was 70 kg. Due to the extent of the lesions and the impact they were having on the patient, he was medicated with acitretin, 25 mg every other day. No topical treatment was prescribed. One month after beginning the treatment, there was already a marked improvement of the dermatosis (Fig. 1d–f). After 6 months, the lesions had completely resolved and acitretin was stopped (Fig. 1g–i). The treatment was very well tolerated by the patient, with the only side effect being mild dryness of the lips. The lesions have not recurred after 1 year of follow-up.

Discussion

PRP is a chronic papulosquamous disorder characterized by reddish-orange scaly plaques, palmoplantar keratoderma, and keratotic follicular papules (1). The disease may progress to erythroderma with distinct areas of uninvolved skin, known as "islands of sparing" (1).

It has an estimated incidence of 1:3,500–5,000 patients and affects individuals of all racial backgrounds equally, with no gender predilection (1).

Although its pathogenesis is still unknown, some conditions such as abnormalities in vitamin A metabolism leading to vitamin-A deficiency, infections, trauma, impaired immunologic response, rheumatism, and malignancy have been implicated (4).

Histology helps in making the diagnosis by excluding other dermatoses such as psoriasis; however, it is not pathognomonic. Hyperkeratosis with alternating orthokeratosis and parakeratosis forming a checkerboard pattern in the stratum corneum, focal or confluent hypergranulosis, follicular plugging with perifollicular parakeratosis forming a shoulder effect, thick suprapapillary plates, broad rete ridges, narrow dermal papillae, and sparse superficial dermal lymphocytic perivascular infiltration are frequent findings (5).

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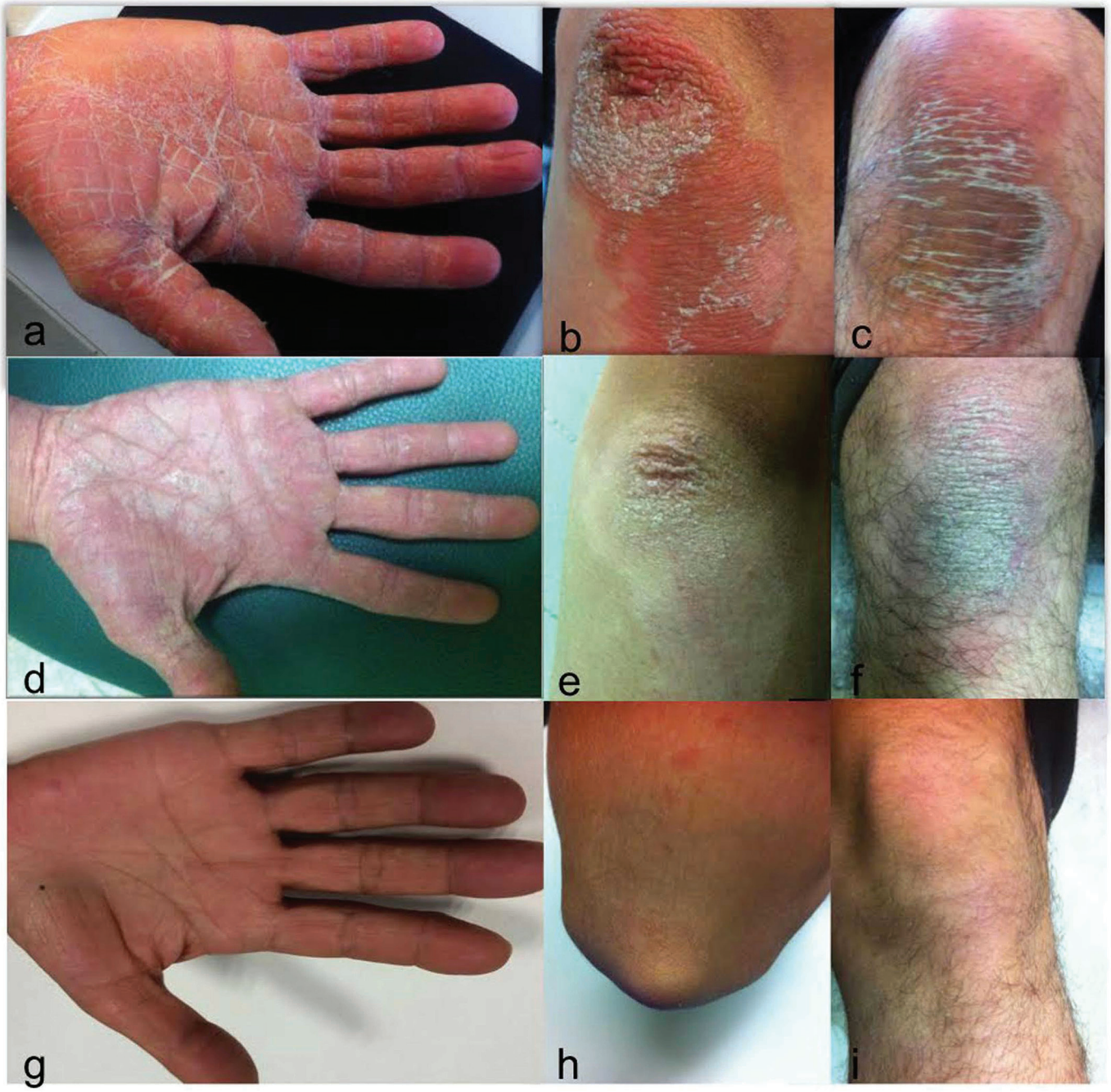

Figure 1 | Clinical aspect of the lesions (a–c: right palm, right elbow, and right knee before treatment, respectively; d–f: right palm, right elbow, and right knee 1 month after beginning treatment, respectively; g–i: right palm, right elbow, and right knee 6 months after beginning treatment, respectively).

There are multiple treatment options for this entity. Because there are no controlled trials to evaluate the efficacy and safety of treatments in patients with PRP, the use of these agents is based on clinical reports. There have been reports on the use of topical agents such as corticosteroids and vitamin D derivates, vitamin A, photo(chemo)therapy, and systemic therapies such as systemic retinoids, azathioprine, methotrexate, cyclosporine, and biological agents (6, 7).

Systemic retinoids are one of the first-line treatments for this condition (8). Dicken reported that, out of 15 patients with PRP treated with oral retinoids, 10 had complete clearing of the lesions and two had partial clearing after 4 to 6 months of therapy (6). The recommended dose of oral acitretin is 0.5 to 0.75 mg/kg/day (8). Response to retinoid therapy is usually evident within 3 to 6 months, although longer courses of treatment are necessary in some patients (6). In our case, we decided to use acitretin because its use in this disease is better documented than isotretinoin and allitretinoin.

As is known, retinoids produce a number of side effects that are dose-related. These include alopecia, cheilitis or chapped lips, dermatitis, epistaxis, nail fragility, peeling rash, xerosis and pruritus, dryness of the eyes, which may result in discomfort, photophobia, blepharoconjunctivitis, and keratitis (9). In our case, the patient was treated with a low-dose regimen of acitretin, which was effective and well tolerated. This highlights the option of using lower doses of retinoids in type IV PRP, reducing the risk of drug-related side effects and non-adherence by the patient.

Finally, this is a case of a late onset type IV PRP, which usually occurs in prepubertal children, showing that it can also affect young adults.

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