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CASE REPORT

Pityriasis rubra pilaris: report of a rare case

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Abstract

Objectives: Report the case of pityriasis rubra pilaris (PRP) in a male patient at the age of 2 years and 10 months. **Methods:** A review of the medical record as well as the literature regarding the present conditions, via PubMed database. **Results:** Male patient, 2 years and 10 months, referred for the first year of appearance of pruritic erythematodescamative annular plaques in lower limbs with dissemination to the trunk, upper limbs, and face, associated with scaling in the plantar region and nail dystrophy. The biopsy presented stratum corneum parakeratotic with hypergranulosis and moderate acanthosis. The diagnosis of pityriasis rubra pilaris was considered conclusive. Isotretinoin and oral antihistamine were prescribed, having complete improvement. **Conclusions:** PRP remains a challenging disease, be it on its pathogeny, diagnosis or treatment. Randomized studies would aid on establishing a standardized treatment for PRP, improving the quality of life of patients suffering from this disease.

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INTRODUCTION

Pityriasis Rubra Pilar (PRP) is a rare disease, first described by Claudius Tarral in 1828, and named by Besnier in 1889[1]. PRP is characterized by a dermatological disorder of keratinization, with follicular and palmoplantar hyperkeratosis, in addition to red-orange scaly plaques.

Although familiar forms are described, they are mostly acquired[2]. The incidence of PRP is low[3], and the disease is equally present between the sexes, except in children where male patients are more affected in a 3:2 ratio[4].

Griffiths, in 1980[5], suggested a classification of PRP into 5 categories, considering the age of onset, clinical course and prognosis. Recently, a 6th category was proposed, associated with HIV[6].

Since the etiology and pathogenesis of this dermatosis are unknown, the treatment adopted so far is empirical[1]. Vitamin-A deficiency, association with immune and infectious diseases such as HIV, and malignant diseases are suggested pathogenic mechanisms[7].

In the present report, we present a case of PRP in a male patient aged 2 years and 10 months.

METHODS

We made a review of the patient's medical record and a literature review in the Pubmed database.

RESULTS

Male patient, 2 years and 10 months old, born by vaginal delivery, referred by another service with a previous diagnosis of atopic dermatitis, was seen at the outpatient clinic of the Dermatology service. The mother reported the appearance of pruritic erythematous-scaly annular plaques on the lower limbs (Figure 1), with subsequent progression to the trunk (Figure 2), upper limbs and face, associated with skin peeling in the plantar region and nail dystrophy (Figure 3). At the time, the patient was using a urea-based emollient; however, he was intolerant due to the burning sensation. He had been previously hospitalized in another institution for 15 days, at the age of 1, for pneumonia and bronchiolitis. There was no family history of similar skin lesions, and his father was atopic (rhinitis). The following diagnostic hypotheses were investigated: psoriasis, pityriasis rubra pilaris, erythrokeratoderma variabilis, atopic dermatitis and ichthyosis. A skin biopsy was ordered and dexchlorpheniramine was prescribed three times a day, oral and topical steroids on the sole of the foot. Laboratory tests performed - blood count, liver function test, renal function, TSH and free T4 - showed normal results. Serologies for hepatitis B, hepatitis C, HIV and syphilis were non-reactive. The biopsy showed skin with a parakeratotic stratum corneum over the epidermis with hypergranulosis and moderate acanthosis. There was no inflammatory infiltrate in the epidermis. In view of the clinical and histopathological findings, the hypothesis of pityriasis rubra pilaris was considered a conclusive diagnosis. We prescribed isotretinoin 0.5 mg/kg/day and an antihistamine orally, and the patient was instructed to maintain hydration, obtaining an improvement in the condition presented, without adverse effects and with outpatient follow-up by the pediatric dermatology team.



Figure 1. Confluent erythematous desquamative plaques with well-defined edges on the left lower limb.

DISCUSSION

Pityriasis Rubra Pilar is an inflammatory dermatosis that affects any age group, with the highest incidence peak between 40 and 60 years of age, and the lowest peak in the first decade of life[8].

The disease is classified into 6 subtypes[5,6]. Type I of the Griffiths classification is considered the classic adult type, and it



Figure 2. Confluent erythematous desquamative plaques with poorly delimited borders on the trunk.



Figure 3. Onychodystrophy and orange-tinted keratoderma in the plantar region, associated with fissures and diffuse skin peeling.

corresponds to 55% of the PRP cases. The atypical form of the adult (type II) is the uncommon variant, being present in 5% of all the cases of the disease. Considered the classic juvenile type, type III corresponds to 10% of cases and affects the age group from 5 to 10 years of age. The clinical features of this subtype are like PRP type I and include orange-red plaques with islands of healthy skin, palmoplantar orange keratoderma, perifollicular keratotic papules, and are usually sporadic. Type IV, juvenile circumscribed, represents 25% of all cases of this dermatosis, being characterized by clearly outlined areas of follicular hyperkeratosis and erythema of the knees and elbows in a prepubertal child. According to Griffiths, type V, atypical juvenile, is responsible for 5% of the cases. Its clinical presentation evolves with follicular hyperkeratosis, palmar-plantar lamellar keratoderma, sclerodermiform alterations of the hands and feet, and is generally associated with a family history. Type IV of the disease is associated with HIV. Skin lesions in PRP result from hyperproliferation of keratinocytes in the epidermis and inflammation in the dermis[7].

The diagnosis of PRP is commonly confused with other papulosquamous and erythrodermic disorders such as psoriasis. The recognition of the 6 forms of this dermatosis is made especially by clinical findings, associated with anatomopathological analysis[9]. There are no serological markers of the disease[9]. In addition to the characteristics of the 6 types described, PRP in adults usually starts on the face and scalp, while in young people it starts in the lower half of the body[10].

While clinically, it may be difficult to differentiate PRP from psoriasis, and histological analysis is the essential criterion for diagnosis. Histologically, PRP presents acanthotic epidermis, mild spongiosis, focal or confluent hypergranulosis and alternation between orthokeratosis and parakeratosis displayed in the horizontal and vertical directions[9,11-14]. Polymorphonuclear cells and Munro's microabscess are always absent in PRP and may be present in psoriasis[9].

The patient reported here was classified as PRP type III, for presenting red-orange plaques with islands of healthy skin, orange palmoplantar keratoderma, and perifollicular keratotic papules. Histology showed a parakeratotic stratum corneum over the epidermis with hypergranulosis and moderate acanthosis, corroborating the literature.

Due to the lack of knowledge about the etiopathogenesis and the rarity of PRP, to date there are no randomized, controlled and double-blind studies with therapeutic options[9]. Some authors believe in the abnormal manifestation of vitamin-A metabolism, as patients with PRP commonly respond to retinoid therapy. Others link the disease to immune dysregulation. A third theory argues that juvenile PRP is a superantigen-mediated disease, as is guttate psoriasis[15]. Therefore, there is no evidence for standard care. In practice, options for topical, systemic or other treatments are available. However, there are forms of the disease that can have spontaneous remission.

The first-line treatment adopted is the systemic use of oral retinoids, which allow for clinical improvement of the patient due to their beneficial effect on keratinization disorders[6,11]. Isotretinoin 1 mg/kg/day is preferable to etretinate 0.5 mg/kg/day, due to its slower release, increasing the risk of teratogenicity[9]. Methotrexate is considered second-line therapy, used when there is no response to retinoids[2,8,9].

Topical and keratolytic emollients are mainly used in children[5,9]. Other therapeutic options include phototherapy, psoralen plus Ultraviolet-A light therapy (PUVA), cyclosporine or biological drugs[9]. For systemic treatment in children, there is the off-label use of acitretin (0.5 mg/kg/day for 3-5 months) associated with narrow band UVB or isotretinoin (0.5-1 mg/kg/day). If the case is severe and refractory to treatment, methotrexate, cyclosporine, azathioprine (in those older than 10 years) or anti-TNF-α biological agents can be used[9]. The treatment of the reported case was supported by the current literature. In the absence of treatment or in cases of therapeutic failure, the complications of this disease range from painful and disabling palmoplantar keratoderma, nail dystrophy and onychoptosis to erythroderma[9].

CONCLUSION

PRP is still a challenging disease, whether in its pathology, diagnosis, or treatment. Searches for biomarkers, such as genetic mutations linked to disease development and progression could contribute to diagnosis and prognosis. Randomized studies could contribute to the establishment of a standardized treatment for PRP, contributing to improve the quality of life of affected patients.

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