

An Acute Onset Erythrodermic Adult Pityriasis Rubra Pilaris Case and Response to Treatment with Methotrexate

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Abstract

Observations: Pityriasis rubra pilaris (PRP) is a rare skin disease characterized by erythematous follicular papules and desquamation. Clinically follicular keratosis, perifollicular erythema, and palmoplantar hyperkeratosis are observed but erythroderma may occur rarely. PRP is one of the rare causes of erythroderma. We identified a PRP case that rapidly progressed to erythroderma in a 49-year-old male patient with no PRP diagnosis before. It is presented due to being a rare case of erythroderma.

Introduction

Pityriasis rubra pilaris (PRP), a rare skin disease which can occur in various clinical manifestations, is characterized by erythematous follicular papules and desquamation [1]. According to the classification made by Griffith, there are two adult-onset PRP types which are classic adult type (Type 1) and atypical adult type (Type 2). Type 1 is the most common PRP type and is usually acute onset. Recently a special PRP type commonly seen in HIV/AIDS patients has been described and is added to the classification as Type 6 [2].

Follicular keratosis, perifollicular erythema, and palmoplantar hyperkeratosis are observed clinically in PRP. It usually starts from the scalp and spreads by a cephalocaudal distribution. Sometimes PRP can be very diffuse causing erythroderma [3]. An acute onset erythrodermic adult PRP case and response to treatment with methotrexate is presented because of being a rare erythroderma reason.

Case Report

Forty-nine-year-old male patient referred to our clinic from out of town with the complaint of widespread redness, scaling and itching throughout the body. In his history we learned that his complaints of seborrhea and dandruff on the scalp started 2 months ago. The patient had no reduction in complaints after the use of topical ketoconazole, clobetasol propionate and betamethasone, and a week later had redness, itching and cracking on all of his body. The dermatological examination shown white color fine scales on the diffuse erythematous background on the scalp, face, whole trunk (Figures 1 and 2) and extremities (Figure 3) were found. Both palmar and plantar had slightly yellowish hyperkeratosis. The patient complained from severe itching and 38 degree fever was observed from time to time. Laboratory results were not abnormal of the patient that also had negative blood, urine and throat cultures. In the histopathological examination of the skin biopsy, alternating parakeratosis in both vertical and horizontal, irregular acanthosis, expansion of the rete ridges and wide suprapapiller areas were observed. There was a slight perivascular mononuc-

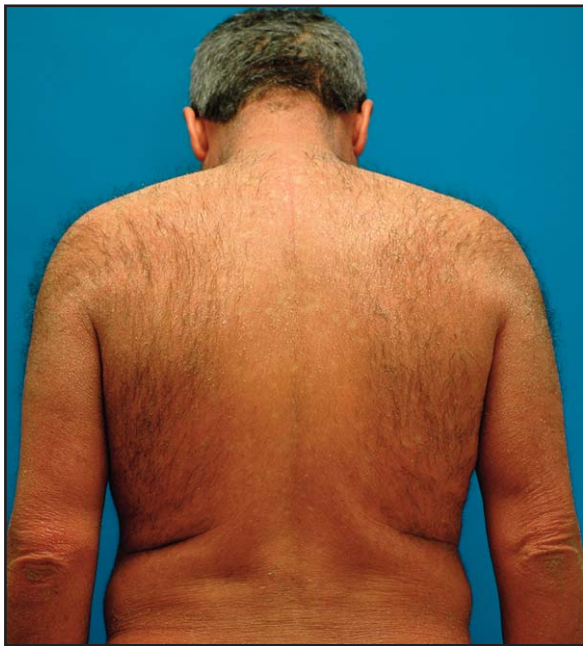


Figure 1. Widespread erythematous lesions on the back



Figure 2. White color fine scales on the diffuse erythematous background on the back

lear leukocyte infiltration in the dermis (**Figure 4**). In the treatment, initially a single dose of 40mg triamcinolone acetate IM, followed by prednisolone at the dose of 20mg/day was continued for three days. Upon the conformation of PRP with the biopsy, prednisolone was stopped reducingly and 20mg/week dose of methotrexate (MTX) was started. The treatment with MTX was discontinued at the end of 10 weeks when the patients complaints regressed after treatment and began to be followed up with acitretin therapy. At the end of one years follow-up his complaints did not repeat.

Discussion

Erythroderma is a rare inflammatory disease characterized as more than 90% of the body being covered by erythema and scales. It was

described by *Hebra* in 1868 [4, 5]. Pre-existing skin disorders are the most common cause among etiologic factors. In previous studies the frequency of pre-existing skin disorders have been reported between 25% and 74.4% [4, 5, 6, 7]. The most common disease that causes erythroderma of dermatoses is psoriasis. Other causes include medications, malign-

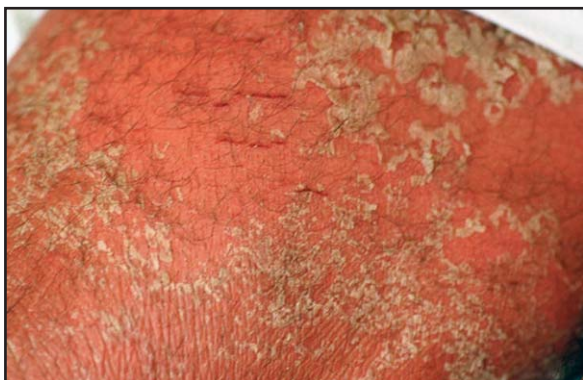


Figure 3. Lesions on the legs



Figure 4. Alternating parakeratosis, irregular acanthosis, and expansion of the rete ridges were seen (H+E x 200)

nancies and idiopathic erythroderma in order of frequency [4, 5].

PRP is one of the rare causes of erythroderma. In studies, the causes of erythroderma were reported between 0.38 to 8.2 0%. Adışen et al [4] reported one PRP in 50 patients with erythroderma. Rym et al [5] reported only one PRP related erythroderma in a 80 diseases series. The maximum PRP rate in erythrodermic patients was reported by Akhyani et al [6] which was responsible for the etiology in 8 of 97 patients with erythroderma. Li et al [7] reported one PRP was identified in 260 cases of erythroderma. In patients with PRP related erythroderma, transient eruptive seborrheic keratoses known to have association with inflammatory diseases have been reported [8, 9]. Our patient did not have the formation of seborrheic keratoses.

Due to PRP being a rarely seen disease, there are controlled studies of treatment where many patients are evaluated and the results are long-term. Publications reporting the results of treatment is usually in the form of case reports [1, 2]. MTX efficacy in PRP treatment is controversial and not recommended for long-term treatment due to side effects [1]. We got good results with MTX in the treatment of the erythrodermic stage of our patient. We did not observe MTX therapy related side effects during the 10 weeks of treatment.

It is interesting that the patient developed erythroderma before the diagnosis of PRP. Considering PRP in the presence of treatment-

resistant erythema and scaling of the scalp lesions in adulthood is important especially in terms of early detection and treatment to prevent possible erythroderma.

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