Generalized erythematous scaly papular lesions

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CLINICAL FINDINGS

A 44-year-old male patient presented with widespread itchy skin eruption all over his body. The condition started 3 months back, and had gradual onset and slow progressive course. Personal or family history of the same condition were negative. The patient had history of intermittent fever. He was treated with different types of topical steroids and antihistamines with unsatisfactory results. Cutaneous examination showed diffuse scaly erythematous papular lesions on face, trunk, upper and lower limbs, with areas of normal skin observed in between erythematous lesions. Joint pain and swelling were not observed, and body temperature was 38 degree. Laboratory investigation including CBC, ESR, liver profile, kidny profile, thyriod profile, were within normal limits. Also, blood, urine and throat cultures were negative. In addition, serological investigation for connective tissue and infectious diseases were within normal limits. General and systemic examination did not show any abnormal findings.

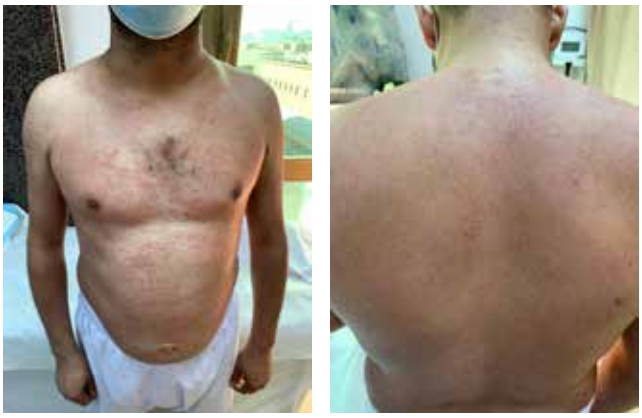

Fig. 1 Diffuse erythematous scaly papular lesions on trunk, upper limb.

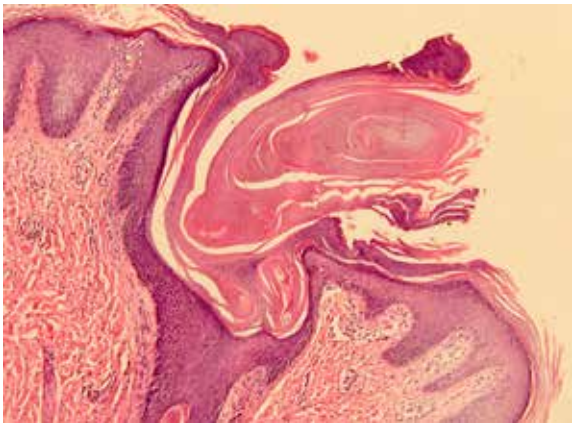

Fig. 2 Hyperkeratosis, parafollicular parakeratosis, dilated follicle filled with keratin. Acanthosis and exocytosis of lymphocytes with focal spongiosis. Mild elongation of rete ridges. Superficial perivascular inflammatory infiltrate formed of lymphohistiocytic cells.

What is your clinical diagnosis?

- Mycosis fungiodes
- Erythrodermic pityriasis rupra pilaris
- Erythrodermic Pityriasis rosea
- Drug eruption

A skin biopsy was done from lesion in trunk and histopathological examination revealed hyperkeratosis and parafollicular parakeratosis with dilated follicle filled with keratin. Epidermal acanthosis with suprapapillary thickeninig. There were no atypical haleod cells or collections within the epidermis and there was no exocytosis of neutrophils in single units or in collections.

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Superficial perivascular infilammatory infiltrate formed of lymphohistiocytic cells. (Fig. 2). The patient was treated with methotrexate (MTX) 20 mg weekly for one month and he achieved complete cure. His condition did not recur after a year of follow-up.

DIAGNOSIS

Adult erythrodermic pityriasis rupra pilaris

DISCUSSION

Pityriasis rubra pilaris (PRP) is an uncommon skin condition characterized by erythematous follicular papules and desquamation in diverse clinical presentations.[1] According to Griffith's classification, there are two forms of adult-onset PRP: classic adult type (Type 1) and atypical adult type (Type 2). PRP type 1 is the most frequent and has an abrupt onset. Type 6, that is typically found in HIV/AIDS patients, has been characterized and added to the categorization.[2]

Clinically, PRP shows follicular keratosis, perifollicular erythema, and diffuse palmoplantar hyperkeratosis. It commonly begins on the scalp and spreads in a cephalocaudal pattern. PRP can sometimes be extremely diffuse, resulting in erythroderma.[3]

Erythroderma is an uncommon inflammatory condition characterized by erythema and scales covering more than 90% of the body. Hebra first described it in 1868.[4,5] Among the etiologic causes, pre-existing skin disorders are the most common. Previous studies have found a prevalence of pre-existing skin disorders ranging from 25% to 74.4 percent.[4-7] Psoriasis is the most prevalent dermatosis that produces erythroderma. In order of frequency, other causes include medicines, cancer, and idiopathic erythroderma[4,5]

Erythroderma can be caused by a variety of factors, but one of the most uncommon is (PRP). In studies, the causes of erythroderma were reported between 0.38 to 8.20%. 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. Akhyani et al.[6] reported the highest PRP rate in erythrodermic patients, in which it was found to be the cause of erythroderma in 8 out of 97 individuals. In 260 cases of erythroderma, Li et al[7] reported that one PRP was found. Transient eruptive seborrheic keratoses has been documented in patients with PRP-related erythroderma,[8,9] which have been linked to inflammatory disorders. Seborrheic keratoses were not present in our patient.

Because PRP is a rare condition, there are not many controlled treatment studies in which a large number of patients are studied and the findings are long-term. Case reports[1,2] are the most common publications that record treatment outcomes. The efficacy of MTX in PRP is debatable, and it is not advised for long-term use due to side effects.[1] We have had good outcomes with MTX in our patient's erythrodermic PRP in the past. And, MTX therapy-related adverse effects were not observed over the 8-week treatment period.

The patient had erythroderma before being diagnosed with PRP, which is interesting. PRP should be considered in the presence of treatment-resistant erythema and scaling of lesions in adults, especially in terms of early detection and treatment to avoid erythroderma.

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