# Pityriasis Rubra Pilar and hypothyroidism*

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**Abstract:** Pityriasis Rubra Pilaris (PRP) is a chronic and rare papulosquamous disorder. Treatment of Pityriasis Rubra Pilaris is based on empiric evidence because of several doubts regarding its etiology and also because of its relative rarity, making randomized studies difficult to perform. Some factors suggest that the metabolism of vitamin A is involved in pathogenesis. We report a case of Pityriasis Rubra Pilaris associated with autoimmune hypothyroidism which presented rapid and complete response after thyroid hormone replacement, without any association with other systemic treatment. In literature there are only three other reports of significant improvement of the lesions after hormonal correction. Deficiency of thyroid hormone inhibits the conversion of carotene into vitamin A, which would be responsible for the occurrence of Pityriasis Rubra Pilaris in this patient.
**Keywords:** Hypothyroidism; Pityriasis rubra pilaris; Therapeutics

## INTRODUCTION

Pityriasis Rubra Pilaris is a chronic dermatosis of unknown etiology. Although there are familial cases, the majority is acquired. It affects men and women in the same proportion and presents bimodal incidence, with peaks on the 1st - 2nd and 5th- 6th decades.

Griffiths proposed its classification in five groups based on clinical appearance, behavior and prognosis.[1] Recently, a sixth type was proposed, associated with HIV, typically refractory to the treatment.[2]

The treatments adopted up to now are empiric, due to its unknown etiology. Currently, the first-line therapy is the use of systemic retinoidsm which allow the clinical improvement of the the patient due to its beneficial effect on disorders of keratinization.[3]

The methotrexate is the second-line therapy. Other therapies include immunosuppressors, such as azathioprine or cyclosporine, stanozolol, phototherapy, extracorporeal photopheresis, fumaric acid, calcipotriol and the biologicals.[3,4,5] All presented variable responses.

Initially it was believed that vitamin A deficiency caused PRP. The possibilities of infectious and autoimmune pathogenesis were also taken into account.[3]

We describe here a case of PRP associated with subclinical hypothyroidism with presented significant improvement of lesions after a short period of Levothyroxine use. Therefore, the importance of researching the alteration of thyroid function in these patients is here emphasized.

## CASE REPORT

The patient was a white man, 30 years old, with erythemato-squamous plaques in a follicular pattern affecting elbows, knees and back of hands, associated with palmoplantar hyperkeratosis (Figure 1). Initially topical corticoids were used together with salicylic acid, with no improvement. Referred to our service, a cutaneous biopsy was requested for anatomopathological (AP) exam, the main diagnostic hypotheses being Psoriasis and Pityriasis rubra pilaris. The first laboratory exams showed subclinical hypothyroidism (TSH: 11.25mU/mL; T4L:1.08ng/dL) associated with hypertriglyceridemia (235mg/dl). HIV serology was negative. It was decided to introduce levothyroxine 50mcg/day and refer the patient to the endocrinologist for follow-up. Only skin hydration was maintained. AP showed ectasia and follicular hyperkeratosis, besides lymphocytary superficial perivascular inflammatory infiltrate, favoring the clinical proposition of Pityriasis Rubra Pilaris. A considerable improvement of the lesions was noticed after only 30 days of hormone replacement (Figure 2). It was opted to follow-up with only cutaneous hydration and adjustment of thyroid function. The subsequent follow-up showed progressive improvement of lesions (Figure 3).

## DISCUSSION

The not very clear pathogenesis of PRP is the main culprit for the multiple and ineffective treatments

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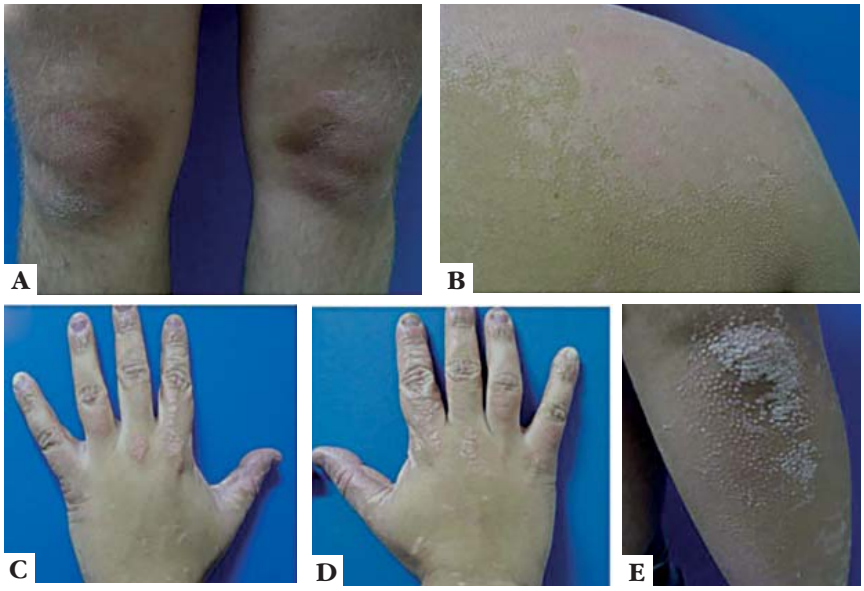


**Figure 1:** Erythemato-squamous plaques with follicular pattern involving elbows, knees, back of hands and trunk

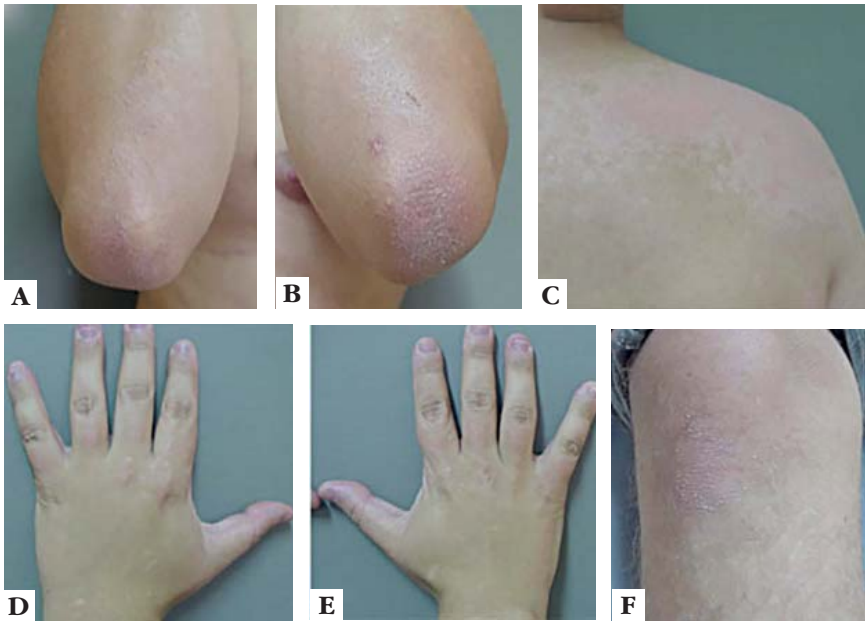


**Figure 2:** Significant improvement of lesions after 30 days of hormone replacement

employed. For many years PRP was associated with vitamin A deficiency, hypothesis corroborated by its histological similarity to hypovitaminosis A and the clinical improvement of patients after high doses of this vitamin. However, vitamin A replacement fell into disuse due to its inconsistent results. The frequent finding of normal serum levels of vitamin A suggests that there would be a failure in its metabolism and not a deficiency.

Vitamin A circulates in the human plasma as retinol, linked to a retinol binding specific carrier protein (RBP).[6]

In the Finzi et al study a low level of this binding protein was found, which suggests the decrease of retinol arrival to the epidermis, which would explain the best clinical response with the use of topical vitamin A or retinoic acid (active form of

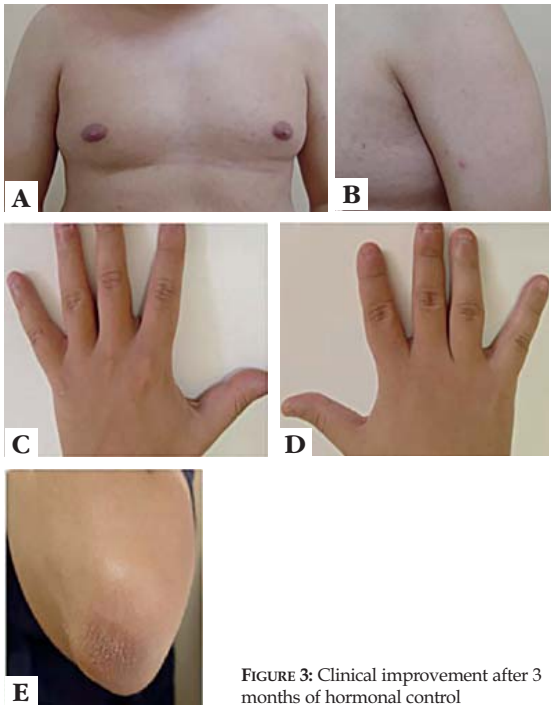


**Figure 3:** Clinical improvement after 3 months of hormonal control

retinol) in comparison with parenteral and oral use of the vitamin.[6]

Other authors believe in the alteration of the immune system due to its association with other autoimmune diseases (myasthenia gravis, hypothyroidism and celiac disease), malignancies (bronchogenic, renal cell and hepatocellular carcinomas) and HIV infection.

Hypothyroidism is directly linked to vitamin A metabolism alteration. The thyroid hormone is fundamental to the conversion of ingested carotenes to vitamin A. The hypothyroidism patients present almost total blockage of this conversion and only a small amount of carotenoids, not converted to vitamin A, are absorbed by humans.[7]

In this case, we show the dramatic improvement of lesions after thyroid hormone correction, without the use of any other therapy. In the literature there are only 3 similar cases reported. The first was in 1964, in a review article.[8] Tunessen et al described the second case of erythrodermic PRP in a 4-year-old child, associated with severe hypothyroidism.[7] The last described case we found was from Orlandini et al, about a 24-year-old man with erythrodermic PRP who presented complete improvement of the clinical picture after 3 months of hormone replacement.[9]

We cannot discard clinical improvement as natural evolution of the disease, for spontaneous remission can occur in up to 80% of the cases, specially in the erythrodermic form; however, it only occurs after 1 to 3 years of evolution and systemic treatments are normally necessary to hasten recovery. In all described cases, clinical improvement was obtained, in average, 30 to 90 days after the start of hormone replacement, without any association with other therapy, strongly suggesting there is direct effect of this treatment on PRP.

Hypothyroidism is also associated with cholesterol level changes. In our case, besides hormone level changes, the increase of triglycerides (TG) was observed. The deficiency of thyroid hormones diminishes the rate of TG removal.

In literature some cases were described about patients whose TG serum level increased after treatment with high doses of vitamin A. Nevertheless, the true causal relation of this increase regarding treatment with vitamin A has been questioned. On these studies cholesterol levels were not dosed before the start of treatment. Perhaps these patients had already had their thyroid function altered and the simple hormonal adjustment led to clinical improvement.[10]

Pityriasis rubra pilaris associated with hypothyroidism could be a particular subtype of acquired PRP, such as the previously described association with HIV. This way, we could assume that PRP would be the cutaneous clinical manifestation of metabolic alteration of vitamin A, with acquired (infectious, autoimmune, paraneoplastic) or familial causes (deficiency of RBP).

To confirm such suppositions more studies with larger numbers of cases are necessary; however, the relative rarity of this dermatosis makes it difficult.

Notwithstanding, this association with hypothyroidism, although uncommon, must be always investigated in the cases of PRP due to the spectacular effect of thyroid hormone replacement and absence of collateral effects. ❑

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