journalofclinicalcases.com

Journal of Clinical Cases

ISSN 2769-9366

Case Report

Mimicking The "Great Imitator": Pityriasis Rubra Pilaris Misdiagnosed As Systemic Lupus Erythematosus: A Case Report.

Ioana Burciu[1], Mihai Augustin Oprea[1], Viviana-Cristina Pavel[1], Valer Mihai Pompilian[1,3], Daniela Nestor[1,3], Razvan Theodor Andrei[1].

Affiliations:

1. *3rd Internal Medicine Department, Colentina Clinical Hospital, Bucharest, Romania.*
2. *Pathological Anatomy Department, Colentina Clinical Hospital, Bucharest, Romania.*
3. *"Carol Davila" University of Medicine and Pharmacy, Bucharest.*

Abstract

Pityriasis rubra pilaris (PRP) is a rare skin condition, currently classified as a keratinization disorder. It is associated with various malignancies and autoimmune diseases. There are six clinical subtypes and can affect both adults and children. Due to its rarity, is often initially misdiagnosed and mistreated. We present a case of a 53-year-old woman with PRP who was initially misdiagnosed as systemic lupus erythematosus (SLE) and treated accordingly. In our case, both the clinical signs and the laboratory testing (minor lymphopenia, positive results for both antinuclear antibodies (ANA) and anti-dsDNA antibodies) suggested a diagnosis of SLE. Treatment consisting in Prednisone 10 mg/d and Plaquenil 400 mg/d was initiated. One month after Plaquenil was introduced, patient returned to the clinic with generalized erythrodermic eruption and a severe cutaneous adverse drug reaction (post-Plaquenil) was suspected. Skin biopsy revealed hyperkeratosis with areas of parakeratosis, mild acanthosis and small foci of perivascular lymphocytic inflammation within the papillary dermis leading to the diagnosis of PRP. Following the dermatologist`s recommendation, Methotrexate 15 mg/week via subcutaneous injection was started. Prednisone was gradually tapered and discontinued with complete resolution of skin lesions at 2 months of treatment. The preliminary hypothesis concerning a post-Plaquenil eruption was ruled out based on the duration of eruption following discontinuation of the drug. In this patient's case, no underlying malignancy or autoimmune disease could be identified. This case report highlights the importance of skin biopsy whenever there is suspicion of PRP and emphasises the possibility of systemic presentation of this rare skin condition.

INTRODUCTION

Pityriasis rubra pilaris is a rare inflammatory skin condition currently classified among keratinization disorders. It can affect children and adults, with a similar prevalence in males and females. The etiology is unknown, although cases of PRP have been reported in the literature in association with viral infections (e.g., HIV), autoimmune conditions (such as dermatomyositis and SLE)(1), and neoplasms (as a paraneoplastic manifestation)(2). It is characterized by a follicular, papulosquamous, erythematous eruption, predominantly localized on the face, trunk, palms, and soles. It may also be associated with arthralgias and myalgias, and there are case reports describing PRP with an abnormal autoimmune profile(3).

Due to its heterogeneous clinical presentation and rarity, PRP is a challenging condition to diagnose. The aim of this article is to present a case of PRP initially misdiagnosed and treated as systemic lupus erythematosus, and to highlight the importance of skin biopsy in complex dermatologic conditions with atypical presentations.

CASE REPORT

We present a case of a 53 years old woman with PRP that was initially misdiagnosed as systemic lupus erythematosus. She had no prior health conditions, no toxic environmental exposure or any drug use. She presented to her local hospital for erythematous eruption on the face, associated with bilateral gonalgia and fatigue. She admitted she had a mild common cold 2 weeks before the symptoms appeared. The laboratory testing showed minor lymphopenia, antinuclear

***Corresponding Author:** **Ioana Burciu**, 3rd Internal Medicine Department, Colentina Clinical Hospital, Bucharest, Romania, **Email:** ioana.burciu@yahoo.com.
Received: 21-May-2025, Manuscript No. JOCC - 4885 **; Editor Assigned:** 22-May-2025 **; Reviewed:** 06-June-2025, QC No. JOCC - 4885 **; Published:** 09-June-2025, **DOI:** 10.52338/jocc.2025.4885.
Citation: **Ioana Burciu**. Mimicking the "Great Imitator": Pityriasis rubra pilaris misdiagnosed as systemic lupus erythematosus: a case report. Journal of Clinical Cases. 2025 June; 5(1). doi: 10.52338/jocc.2025.4885.

antibodies (ANA) and anti-dsDNA antibodies both positive in low titer with no other abnormalities. The diagnosis of systemic lupus erythematosus was made and treatment consisting of corticotherapy (Prednisone 10 mg/d) and Plaquenil 400 mg/d was started. One month after the initiation of Plaquenil she returned to the clinic with generalized erythrodermic eruption and soon she was transferred to our clinic with the suspicion of severe cutaneous adverse drug reaction (post-Plaquenil). At admission, the patient was stable, with an erythrodermic eruption consisting of confluent plaques disseminated all over the body, most predominantly to her trunk and face. The eruption was slightly pruritic, and she had no mucosal involvement. She complained about gonalgia, describing it as mechanical joint pain.

Plaquenil treatment was discontinued, and the patient received pulse therapy with intravenous methylprednisolone 500 mg/day for 3 days, followed by oral corticosteroids (Prednisone 30 mg/day) with gradual tapering. Local treatment consisted of topical corticosteroids.

Although a drug eruption was still strongly suspected, the initial diagnosis of systemic lupus erythematosus remained uncertain, as the clinical presentation did not fully support this etiology.

A skin biopsy was performed, which revealed prominent follicular plugging, compact hyperkeratosis, alternating parakeratosis, and acanthosis, with minimal perivascular lymphocytic infiltrate. Based on these histopathological findings, a diagnosis of pityriasis rubra pilaris was made. It is important to highlight that no basal stratum atrophy was found. (4)

Laboratory testing showed a normal hemogram and peripheral blood smear. ANA, anti-dsDNA, anti-Ro, anti-La, and anti-Sm antibodies were within normal limits, as were rheumatoid factor, anti-CCP antibodies, and complement levels (C3, C4).

To exclude a possible paraneoplastic etiology, a computed tomography scan of the lungs, abdomen, and pelvis was performed, which revealed no evidence of solid malignancies. Serum protein electrophoresis was normal. A viral panel including hepatitis B and C, HIV, and EBV was also negative. Musculoskeletal ultrasound of both knees showed no signs of active inflammatory joint pathology.

As the extensive investigations revealed no identifiable underlying cause, the diagnosis of idiopathic pityriasis rubra pilaris was established. The initial diagnoses of drug-induced eruption and systemic lupus erythematosus were ruled out.

The patient was referred to a dermatologist and initiated systemic treatment with methotrexate 15 mg/week via subcutaneous injection. Prednisone was gradually tapered and discontinued after three weeks.

After two months of methotrexate treatment, the patient presented with complete resolution of skin lesions.

Figure 1. Confluent erythrodermic ring-like lesions disseminated on the trunk and on the limbs.

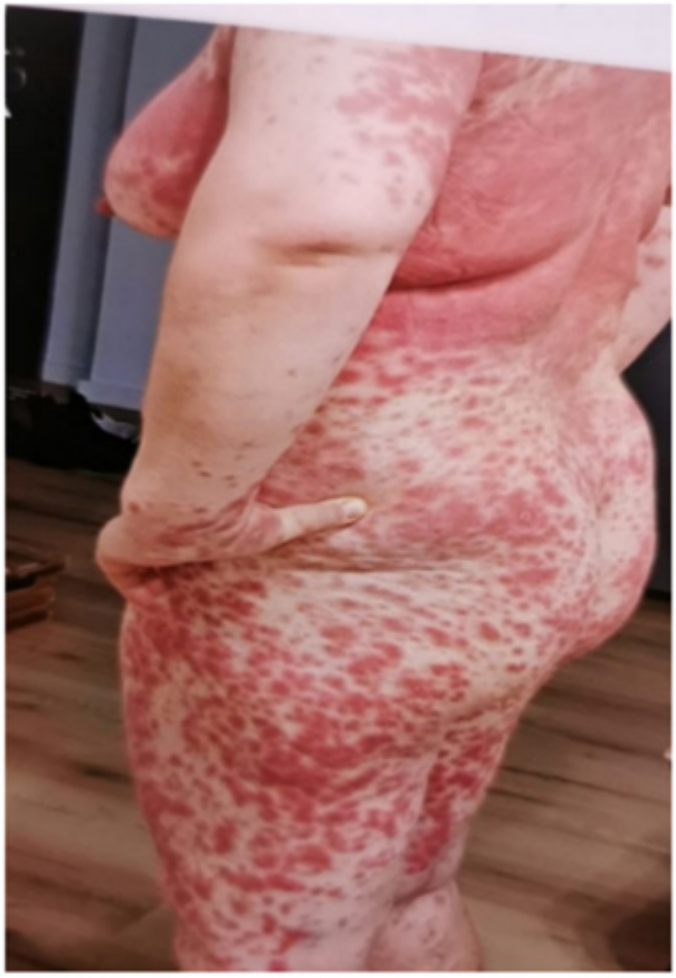


Figure 2. Clinical presentation with erythematous confluent scaly plaques after 1 month of Plaquenil treatment

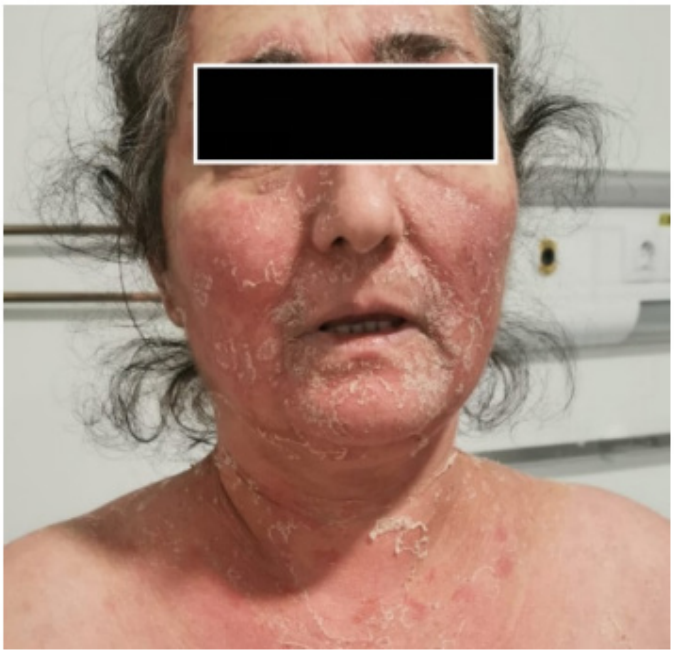


Figure 3. A dilated follicular ostium with focal parakeratosis. HE x100

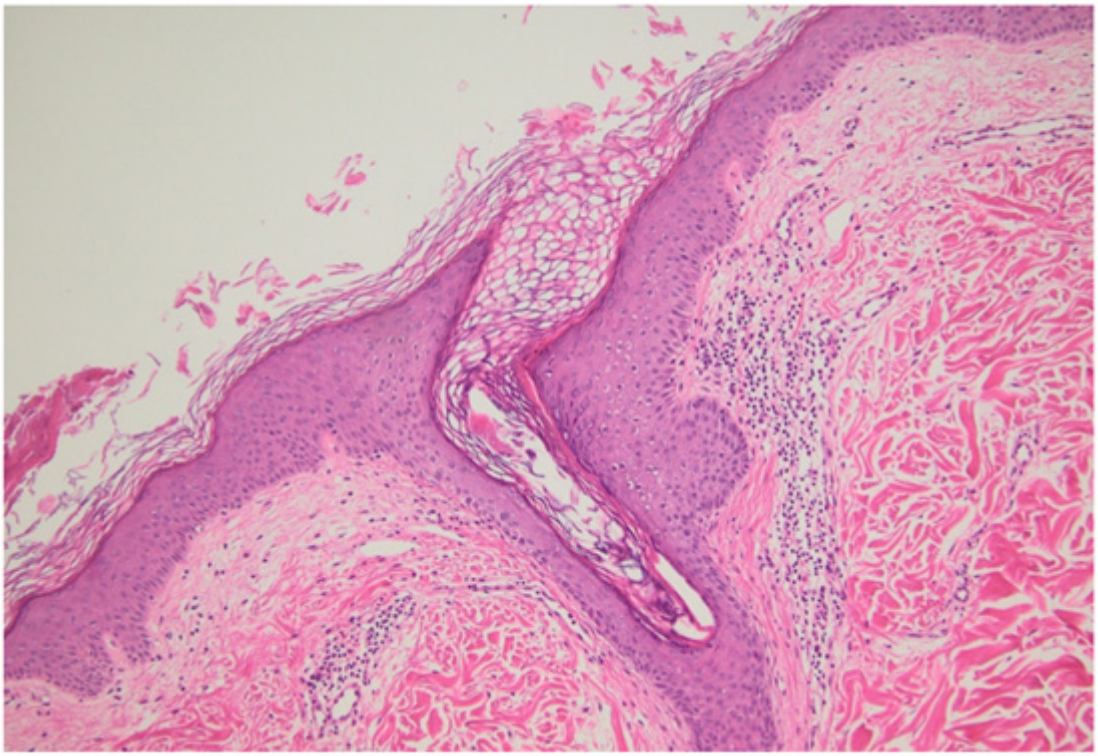


Figure 4. Hyperkeratosis with areas of parakeratosis; mild acanthosis; small foci of perivascular lymphocytic inflammation within the papillary dermis. HE x100.

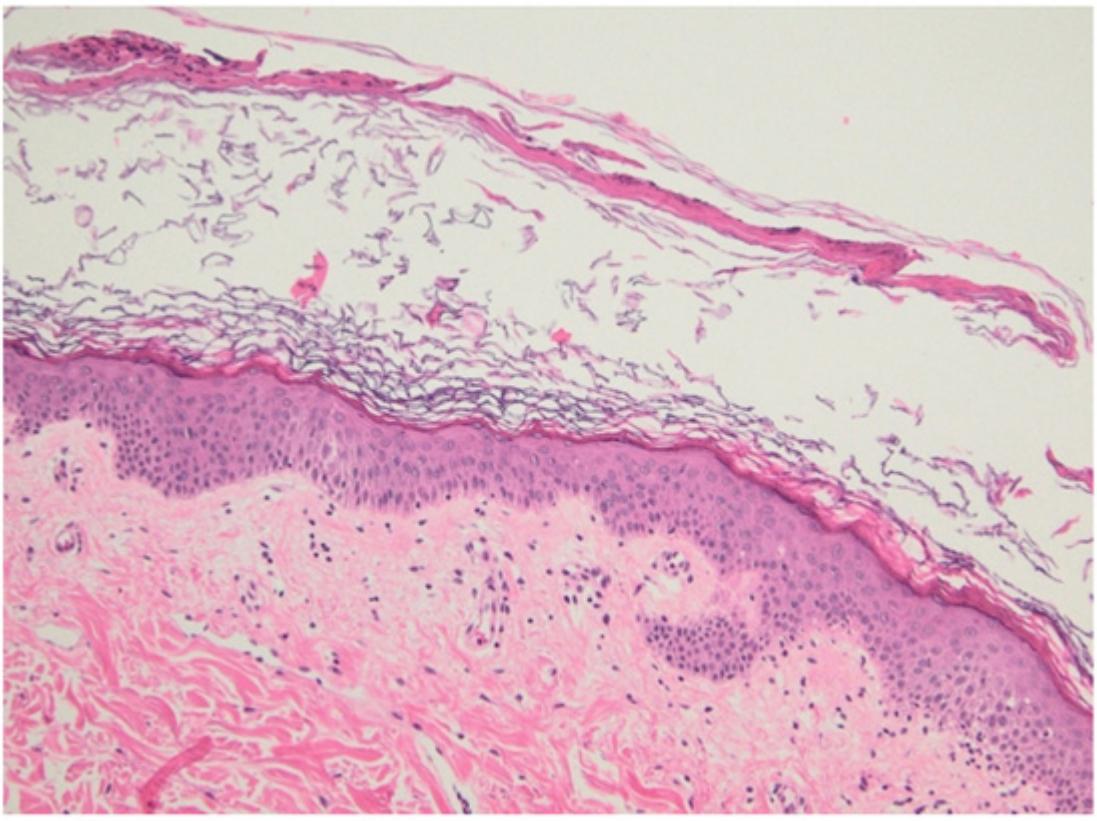
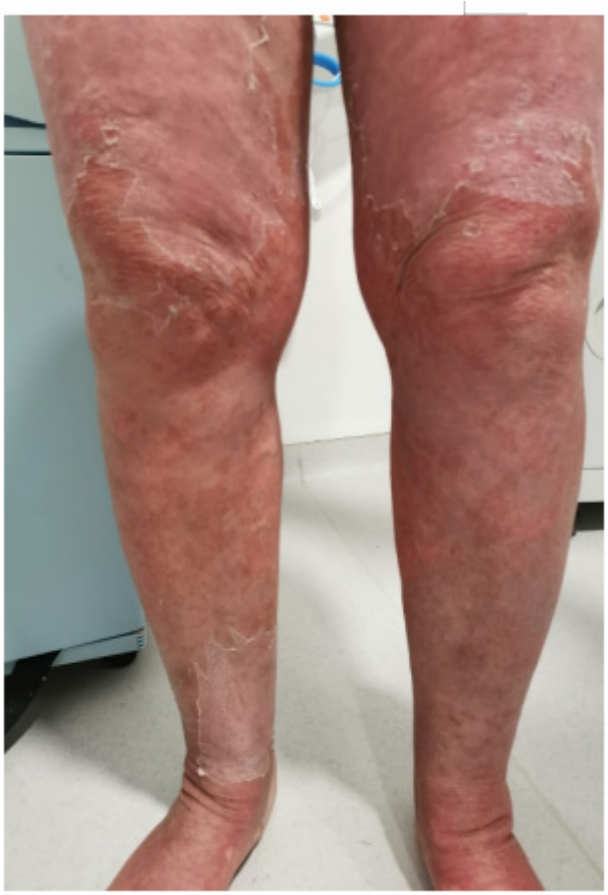


Figure 5. Large patches of skin desquamation and reduced erythema five days after the starting of topical corticosteroids.



DISCUSSION

Most cases of PRP are sporadic and of unknown etiology(5). In the present case, the abnormal laboratory findings—specifically the presence of autoantibodies typically associated with lupus—combined with arthralgia, led to an initial misdiagnosis.

There are a few reports in the literature describing PRP cases associated with both abnormal immunological findings and systemic symptoms such as arthralgia and myalgia(3). At the time of admission to our clinic, lupus-specific autoantibodies – anti-dsDNA, anti-Ro, anti-La, and anti-Sm antibodies (tested using the ELISA method) were negative, despite the patient experiencing a disease flare. It is worth noting that, at the time of the first presentation, a skin biopsy was not performed. There are also reported cases in which the diagnosis of PRP was only established after two or more skin biopsies(6).

In consideration of the preliminary hypothesis concerning the occurrence of a post-drug eruption, the duration of the eruption exceeded the period of administration. Moreover, the cessation of drug administration did not exert an influence on the progression of the eruption.

Pityriasis rubra pilaris is a rare and diagnostically challenging condition. Once PRP is diagnosed, clinicians should investigate potential associated abnormalities, guided by the patient's initial clinical presentation.

There are six recognized types of PRP, each with distinct clinical features and prognoses.(7) The most common one is the classical adult-onset PRP, which represents 50% of all cases. It can associate with malignancies, so a diagnosis of PRP requires further investigations.

In our patient's case, no underlying malignancy or autoimmune disease could be identified.

The treatment of PRP involves both systemic and topical approaches. Topical therapy typically includes emollients and calcipotriol. Systemic treatments include methotrexate, acitretin, and isotretinoin.

Biologic therapies have shown efficacy in the treatment of PRP, including etanercept (a TNF-alpha inhibitor), ustekinumab (IL-12/IL-23 blockade), guselkumab and risankizumab (both IL-23 inhibitors), and secukinumab (IL-17 inhibitor)(8). Narrow-band ultraviolet B (NB-UVB) phototherapy and extracorporeal photopheresis seem beneficial in some cases(9).

PROGNOSIS

The prognosis for our patient is favorable. She had a rapid response to methotrexate, with complete resolution of her skin lesions. In cases of spontaneous adult-onset PRP, approximately 50% of patients will have gradual disease resolution over months to years, with a mean time of around 3 years. Once resolved, recurrence is rare. However, the remaining 50% of patients may experience a lifelong course of the disease(7).

CONCLUSIONS

Pityriasis rubra pilaris is a rare skin condition that can have systemic manifestations. It may be associated with various malignancies and autoimmune diseases. The diagnosis can be challenging for clinicians and histopathologists, particularly if they have not previously encountered the condition in their practice. As the diagnosis is often delayed, this can significantly impact the patient's quality of life. A skin biopsy should be performed whenever there is suspicion of PRP(10).

REFERENCES

1. Polat M, Lenk N, Ustün H, Oztaş P, Artüz F, Alli N. Dermatomyositis with a pityriasis rubra pilaris-like eruption: an uncommon cutaneous manifestation in dermatomyositis. Pediatr Dermatol. 2007 Mar-Apr;24(2):151-4. doi: 10.1111/j.1525-1470.2007.00364.x. PMID: 17461814.

2. Mehta N, Coates MM, Miles JA, Miedema J, Blasiak RC A

case of paraneoplastic pityriasis rubra pilaris JAAD Case Rep 2023 Jul 13;39:125-129 doi: 101016/j.jdcr202306045 PMID: 37680569; PMCID: PMC10480444.

3. Gregoriou S, Chiolou Z, Stefanaki C, Zakopoulou N, Rigopoulos D, Kontochristopoulos G Pityriasis rubra pilaris presenting with an abnormal autoimmune profile: two case reports J Med Case Rep 2009 Nov 13;3:123 doi: 101186/1752-1947-3-123 PMID: 19946540; PMCID: PMC2783063.

4. Fetter T, Braegelmann C, De Vos L, Wenzel J. Current Concepts on Pathogenic Mechanisms and Histopathology in Cutaneous Lupus Erythematosus. Front Med. 2022 May 30;9:915828.

5. A case of recurrent and paraneoplastic pityriasis rubra pilaris. https://www.jaadcasereports.org/article/S2352-5126(21)00302-7/fulltext.

6. Abang Hashim DH, Ismail IA, Tawil Z, Abd Halim H. Early Presentation of Pityriasis Rubra Pilaris Mimicking Tinea Corporis: Diagnostic Challenges of a Rare Skin Condition. Am J Case Rep. 2022 Aug 21;23:e936906. doi: 10.12659/AJCR.936906. PMID: 35988013; PMCID: PMC9404675.

7. Greiling TM, Brown F, Syed HA. Pityriasis Rubra Pilaris. [Updated 2024 Apr 30]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482436/.

8. Pityriasis rubra pilaris Sagut, Pelin et al. Journal of the American Academy of Dermatology, Volume 92, Issue 2, 376 - 378.

9. Roenneberg S, Biedermann T. Pityriasis rubra pilaris: algorithms for diagnosis and treatment. J Eur Acad Dermatol Venereol. 2018 Jun;32(6):889-898. doi: 10.1111/jdv.14761. Epub 2018 Jan 17. PMID: 29247481.

10. Niemi KM, Kousa M, Storgards K, Karvonen J. Pityriasis rubra pilaris. A clinico-pathological study with a special reference to autoradiography and histocompatibility antigens. (Dermatologica. 1976;152(2):109.).