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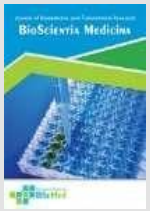

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From Patches to Plaques: A Diagnostic Challenge in a Case of Erythroderma Secondary to Pityriasis Rubra Pilaris

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A B S T R A C T

Background: Erythroderma, a dramatic and potentially life-threatening condition characterized by fiery redness engulfing over 90% of the skin's surface, presents a formidable diagnostic challenge due to its myriad underlying causes. Among these, pityriasis rubra pilaris (PRP), a rare inflammatory skin disorder, stands out with its distinctive features and often perplexing presentation. This case unveils the intricate diagnostic journey of a young man whose erythroderma masked an underlying PRP, further complicated by the subtle interplay of stress. **Case presentation:** An 18-year-old male presented with a one-month history of alarming erythroderma accompanied by distressing itching, fever, and sleep disturbances. Adding to the complexity, he exhibited characteristic 'nappes claires' – islands of normal skin amidst the erythrodermic sea – a hallmark of PRP. Palmoplantar keratoderma, alopecia areata, and ectropion further painted an intriguing clinical picture. Histopathological examination revealed the telltale 'checkboard' pattern, confirming PRP as the culprit. Notably, the patient's history revealed a compelling link between stress and disease exacerbation, adding a psychosomatic dimension to the case. Systemic corticosteroids and methotrexate, alongside topical emollients, brought about significant clinical improvement, underscoring the importance of early diagnosis and targeted treatment. **Conclusion:** This case underscores the critical need to consider PRP in the labyrinth of erythroderma diagnoses, particularly when 'nappes claires' and a history of stress are intertwined. By shining a light on the diagnostic subtleties and therapeutic nuances of PRP-associated erythroderma, this report empowers clinicians to navigate the complexities of this rare and challenging condition, ultimately improving patient outcomes and quality of life.

1. Introduction

Erythroderma, also known as exfoliative dermatitis, is a severe and potentially life-threatening dermatological condition characterized by diffuse erythema and scaling that affects a substantial portion, typically exceeding 90%, of the body surface area. This striking clinical presentation represents the culmination of a variety of underlying etiologies, posing a significant diagnostic challenge for clinicians. The diverse range of potential causes includes pre-existing inflammatory skin diseases, adverse drug reactions, underlying malignancies, and systemic disorders, each with its own unique pathogenesis and clinical course. Accurate and timely identification of the specific etiology is crucial as it guides appropriate management strategies and ultimately influences patient prognosis. Among the inflammatory dermatoses that can manifest as erythroderma, pityriasis rubra pilaris (PRP) is a relatively rare but clinically important entity. PRP is a chronic

inflammatory skin disorder characterized by the development of reddish-orange plaques. A distinctive feature of PRP is the presence of islands of uninvolved skin within the generalized erythema, referred to as "nappes claires". These areas of sparing, while characteristic, may be subtle or obscured in cases where erythroderma is the predominant presentation, further complicating the diagnostic process. The precise prevalence of PRP remains elusive, with estimates suggesting a range of approximately 1 in 5,000 to 50,000 individuals in the United Kingdom. This variability in prevalence estimates may be attributed to factors such as geographical differences, diagnostic challenges, and variations in disease classification. The etiology of PRP is complex and not yet fully understood, but it is believed to involve a combination of genetic predisposition, immune dysregulation, and environmental triggers.[1-4]

The diagnosis of PRP can be particularly challenging in cases where the patient presents with erythroderma. In such instances, the classic clinical features of PRP, such as the aforementioned "nappes claires," follicular papules, and palmoplantar keratoderma, may be less prominent or more difficult to discern amidst the widespread erythema and scaling. A thorough and detailed history, coupled with a meticulous physical examination, is therefore of paramount importance in guiding the diagnostic workup. Histopathological examination of skin biopsies plays a crucial role in confirming the diagnosis of PRP. The characteristic histopathological findings include hyperkeratosis (thickening of the stratum corneum), parakeratosis (retention of nuclei in the stratum corneum), and a distinctive "checkboard" pattern of alternating orthokeratosis (keratinization without retention of nuclei) and parakeratosis. Focal acantholytic dyskeratosis (premature separation of keratinocytes with abnormal keratinization) may also be observed. These histopathological features, in conjunction with the clinical presentation, aid in differentiating PRP from other erythrodermic conditions. The management of erythroderma secondary to PRP necessitates a multimodal therapeutic approach aimed at controlling inflammation, alleviating symptoms, and improving the patient's overall quality of life. Systemic corticosteroids are frequently employed to achieve rapid control of the acute inflammatory process, particularly in cases of severe erythroderma. These agents exert potent anti-inflammatory effects by suppressing various components of the immune system. However, due to the potential for significant side effects with long-term use, systemic corticosteroids are typically used for short-term management or as a bridge to other therapies.[5-7]

Disease-modifying agents, such as methotrexate, are often necessary for long-term management of PRP, particularly in chronic or recalcitrant cases. Methotrexate is an antimetabolite that exhibits immunomodulatory properties, inhibiting dihydrofolate reductase and thereby interfering with DNA synthesis and cellular proliferation. It has been shown to be effective in inducing and maintaining remission in PRP, with response rates ranging from 33% to 75% in some studies. Regular monitoring for potential side effects, such as hepatotoxicity and bone marrow suppression, is essential in patients receiving methotrexate therapy. In addition to systemic therapies, topical treatments play an important role in the management of erythroderma secondary to PRP. Topical emollients are crucial for maintaining skin hydration and reducing dryness and scaling, which are prominent features of erythroderma. Topical corticosteroids of appropriate potency can also be used to alleviate localized inflammation and itching.[8-10] This case report describes a case of erythroderma secondary to PRP in a young adult male. The report highlights the diagnostic challenges encountered in differentiating PRP from other causes of erythroderma and emphasizes the importance of considering potential triggers for disease exacerbation

2. Case Presentation

The patient, an 18-year-old male student, presented with a constellation of dermatological symptoms that had evolved over a period of one

month. The primary complaint, as articulated by the patient, centered on the emergence of reddish patches and thick scales distributed across the entirety of his body. This widespread eruption was accompanied by intense pruritus, a distressing symptom that significantly impacted the patient's overall comfort and well-being. In addition to the dermatological manifestations, the patient reported experiencing fever and difficulty sleeping, further underscoring the systemic impact of his condition. A notable aspect of the patient's medical history was the report of a similar episode occurring approximately four months prior to this presentation. That earlier occurrence was specifically associated with a period of heightened anxiety related to his father's stroke. This historical detail suggests a potential correlation between psychological stress and the exacerbation or recurrence of the patient's dermatological condition, a factor that would later play a significant role in the diagnostic and therapeutic considerations. Importantly, the patient denied any family history of a similar illness, suggesting that the condition was unlikely to be attributable to a primary genetic predisposition. Furthermore, the patient reported having no known allergies and denied the use of any medications prior to this presentation. His social history was unremarkable, with the patient specifically denying any history of smoking or alcohol consumption. The physical examination revealed that the patient's general appearance was within normal limits. His vital signs, including temperature, heart rate, blood pressure, and respiratory rate, were all documented to be within the expected physiological ranges. However, the dermatological examination presented a much more striking and complex picture. The distribution of skin lesions was generalized, involving a significant proportion of the patient's total body surface area, estimated to be greater than 90%. This extensive involvement is clinically defined as erythroderma, a severe condition characterized by widespread erythema and scaling. The morphology of the skin lesions was notable for several key features. Erythema, or redness of the skin, was a dominant characteristic, indicating significant inflammation. Scaling was also prominent, reflecting an abnormality in the keratinization process. Importantly, the examination revealed the presence of "nappes claires," a term used to describe islands of relatively normal-appearing skin interspersed within the areas of erythema and scaling. This phenomenon is a distinctive feature often associated with pityriasis rubra pilaris (PRP), a rare inflammatory skin disorder. Additionally, follicular papules were observed, which are small, raised bumps associated with hair follicles, further supporting the possibility of PRP or other follicular-based dermatoses. Beyond the generalized skin involvement, specific regional findings were also documented. Palmoplantar involvement was characterized by hyperkeratosis, which is a thickening of the stratum corneum, accompanied by yellowish scales. This finding suggests an abnormality in the differentiation and maturation of keratinocytes in these regions, commonly seen in conditions affecting the palms and soles. Scalp involvement was manifested as alopecia areata, a condition characterized by patchy hair loss, suggesting an autoimmune process affecting hair follicles. Finally, eye involvement was noted in the form of ectropion, an outward turning of the eyelid margin, which can result from chronic inflammation or scarring of the eyelid skin. In order to further investigate the patient's condition and to assess for any systemic involvement or underlying abnormalities, a series of laboratory investigations were conducted. The complete blood count (CBC) revealed a hemoglobin level of 14.5 g/dL, which falls within the normal reference range of 13.5 to 17.5 g/dL for adult males. This finding suggests that the patient was not anemic. The white blood cell count was 8,000/µL, which is also within the normal range of 4,000 to 11,000/µL. The differential count showed neutrophils at 60%, lymphocytes at 30%, monocytes at 5%, and eosinophils at 5%. This distribution of white blood cell types is generally within normal limits, although a slight elevation in eosinophils can sometimes be seen in inflammatory skin conditions, although in this case it is within

normal limits. The platelet count was 250,000/μL, which is within the normal range of 150,000 to 450,000/μL, indicating normal platelet production and function. The erythrocyte sedimentation rate (ESR) was measured at 20 mm/hr, which is slightly elevated above the normal range of 0 to 15 mm/hr. The ESR is a non-specific marker of inflammation, and its elevation suggests the presence of an inflammatory process within the body. The C-reactive protein (CRP) level was 5 mg/L, which is at the upper limit of the normal range of 0 to 5 mg/L. CRP is another acute-phase reactant protein that is produced by the liver in response to inflammation. While within the normal range, the value supports the presence of an inflammatory process, as indicated by the elevated ESR. Liver function tests were performed to assess the integrity and function of the liver. The aspartate aminotransferase (AST) level was 25 U/L, and the alanine aminotransferase (ALT) level was 20 U/L, both of which are within the normal reference range of up to 40 U/L. These findings indicate that there was no evidence of significant hepatocellular injury or liver inflammation. The alkaline phosphatase (ALP) level was 80 U/L, which is within the normal range of 40 to 129 U/L, suggesting normal biliary function. The bilirubin level was 1 mg/dL, which is within the normal range of 0.3 to 1.2 mg/dL, indicating normal bilirubin metabolism and excretion. Renal function tests were also conducted to evaluate the patient's kidney function. The blood urea nitrogen (BUN) level was 15 mg/dL, which is within the normal range of 7 to 20 mg/dL. The creatinine level was 1 mg/dL, which is within the normal range of 0.7 to 1.3 mg/dL. These results indicate that the patient's renal function was within normal limits. In addition to the laboratory investigations, imaging studies were performed. A chest X-ray was conducted, and the results were reported as normal, indicating the absence of any significant pulmonary abnormalities. To further elucidate the nature of the patient's dermatological condition, a histopathological examination of a skin biopsy was performed. The epidermal layer showed hyperkeratosis, parakeratosis, alternating orthokeratosis, and focal acantholytic dyskeratosis, arranged in a "checkboard" pattern. Hyperkeratosis, as previously mentioned, is a thickening of the stratum corneum, the outermost layer of the epidermis. Parakeratosis refers to the retention of nuclei in the keratinocytes of the stratum corneum, which is an abnormal finding. Orthokeratosis is the normal process of keratinization without the retention of nuclei. The alternating pattern of orthokeratosis and parakeratosis, described as a "checkboard" pattern, is a characteristic histopathological feature often associated with PRP. Focal acantholytic dyskeratosis indicates premature separation of keratinocytes with abnormal keratinization. The dermis showed a mild perivascular lymphocytic infiltrate, suggesting a mild inflammatory response in the dermal layer of the skin. Based on the synthesis of the clinical presentation, laboratory investigations, and histopathological findings, a primary diagnosis of erythroderma secondary to pityriasis rubra pilaris was established. It is crucial to emphasize that erythroderma, as observed in this case, is not a diagnosis in itself but rather a clinical manifestation of an underlying condition. In this instance, PRP was identified as the underlying etiology. In the process of arriving at the primary diagnosis, several differential diagnoses were considered. These included psoriasis, a chronic autoimmune skin condition characterized by erythematous plaques with silvery scales; atopic dermatitis, a chronic inflammatory skin condition often associated with allergies; seborrheic dermatitis, a common skin condition affecting areas rich in sebaceous glands; and drug reaction, which can mimic a variety of dermatological conditions. However, the constellation of clinical features, particularly the presence of "nappes claires," the characteristic palmoplantar keratoderma, the scalp involvement with alopecia areata, and the distinctive histopathological findings, ultimately supported the diagnosis of erythroderma secondary to PRP (Table 1).

The management of this patient with erythroderma secondary to pityriasis rubra pilaris (PRP) was initiated with a comprehensive treatment regimen designed to

address both the acute inflammatory process and the chronic nature of the underlying skin disorder. This therapeutic strategy encompassed both systemic and topical medications, with careful attention to the patient's clinical response and the need for ongoing maintenance therapy. Furthermore, patient education and counseling played an integral role in the overall management plan, focusing on stress management and optimal skin care practices. The initial treatment phase focused on rapidly controlling the widespread inflammation and alleviating the patient's distressing symptoms. This was achieved through a combination of systemic and topical therapies, strategically selected to target different aspects of the disease process. Systemic medications were crucial in addressing the extensive erythroderma and the associated systemic symptoms, such as fever. The cornerstone of the systemic treatment was the administration of methylprednisolone, a potent corticosteroid. Methylprednisolone was initiated at a dosage of 48 mg, administered orally once daily. Corticosteroids exert their therapeutic effects through various mechanisms, including suppression of inflammatory cytokine production, inhibition of leukocyte migration and activation, and reduction of vascular permeability. In the context of erythroderma, the primary goal of corticosteroid therapy is to rapidly reduce inflammation, thereby alleviating erythema, scaling, and pruritus. The high initial dosage of methylprednisolone reflects the severity of the patient's condition and the need for prompt control of the inflammatory process. The duration of this initial high-dose corticosteroid therapy was planned for two weeks. In addition to corticosteroids, methotrexate was introduced as a disease-modifying agent. Methotrexate was administered at a dosage of 10 mg, given orally once weekly. Methotrexate is a folate antimetabolite that exerts immunomodulatory and anti-inflammatory effects. It inhibits dihydrofolate reductase, an enzyme involved in DNA synthesis and cellular proliferation. In dermatology, methotrexate is commonly used in the management of chronic inflammatory skin diseases, including psoriasis and PRP. Its use in PRP aims to provide long-term control of the disease, reduce the need for chronic corticosteroid therapy, and prevent disease flares. The initiation of methotrexate early in the treatment course reflects an intention to establish a foundation for long-term management, even as corticosteroids provide rapid initial relief. The duration of methotrexate administration was also planned for two weeks initially, with the understanding that it would likely be continued for a more extended period based on the patient's response. To mitigate potential side effects associated with methotrexate therapy, folic acid supplementation was included in the regimen. Folic acid was administered at a dosage of 5 mg, given orally once daily, except on the days when methotrexate was administered. Methotrexate can interfere with folate metabolism, potentially leading to adverse effects such as mucositis, gastrointestinal disturbances, and bone marrow suppression. Folic acid supplementation helps to counteract these effects by providing an alternative source of folate. The duration of folic acid supplementation was aligned with the initial two-week period of methotrexate therapy. To address the patient's pruritus, cetirizine, an antihistamine, was prescribed. Cetirizine was administered at a dosage of 10 mg, given orally once daily. Antihistamines block the action of histamine, a mediator of allergic and inflammatory responses, thereby reducing itching. While not directly targeting the underlying inflammatory process of PRP, cetirizine provides symptomatic relief, improving the patient's comfort and quality of life. The duration of cetirizine therapy was planned for two weeks, consistent with the initial phase of intensive treatment. Topical medications were used as adjunctive therapies to address the local manifestations of the disease, such as scaling and inflammation, and to support skin barrier function. Desoximetasone cream, a high-potency topical corticosteroid, was prescribed for application to affected skin areas. The cream was applied twice daily. Topical corticosteroids exert anti-inflammatory effects by inhibiting the release of inflammatory mediators and suppressing local immune responses. Their use

helps to reduce erythema, scaling, and pruritus in localized areas. Desoximetasone, being a high-potency steroid, was likely used for areas of more severe inflammation or thicker plaques, with careful consideration to avoid prolonged use on sensitive areas to minimize the risk of local side effects such as skin atrophy or telangiectasia. The duration of desoximetasone cream application was planned for two weeks. In addition to the active medications, Vaseline album, a petrolatum-based emollient, was prescribed for application to the entire skin surface. Vaseline album was applied twice daily. Emollients play a crucial role in the management of erythroderma by providing hydration to the skin, reducing dryness and scaling, and restoring the skin barrier function. The widespread erythema and scaling in erythroderma disrupt the normal skin barrier, leading to increased transepidermal water loss and impaired barrier function. Emollients help to restore the barrier, prevent further moisture loss, and improve skin comfort. The use of Vaseline album was planned for two weeks initially, but with the understanding that emollient therapy would be a crucial ongoing component of the patient's skin care regimen. The patient's clinical course was closely monitored through regular follow-up visits, during which the effectiveness of the treatment regimen was assessed, and modifications were made as necessary. At the week 3 follow-up visit, a clinical evaluation of the skin lesions and an assessment of the severity of itching were conducted. The evaluation revealed a significant improvement in both erythema and scaling. The widespread redness and the thick scales that had characterized the patient's initial presentation had noticeably diminished. Furthermore, the patient reported a reduction in the intensity of pruritus, indicating a positive response to the treatment regimen. This improvement suggested that the initial treatment phase had been effective in controlling the acute inflammatory process and alleviating the patient's symptoms. Based on the observed clinical improvement, a modification was made to the systemic corticosteroid therapy. The methylprednisolone dosage was tapered from 48 mg daily to 24 mg daily. This tapering strategy reflects the principle of gradually reducing corticosteroid dosage as the disease comes under control, in order to minimize the risk of corticosteroid-related side effects. The reduced dosage of methylprednisolone was continued once daily for one week. At the week 4 follow-up visit, further clinical evaluation of the skin lesions and assessment of pruritus were performed. The patient demonstrated continued improvement in his dermatological condition. The erythema and scaling had further subsided, and only minimal residual erythema and scaling were noted. The patient also reported minimal itching, indicating a sustained reduction in pruritus. These findings confirmed the ongoing effectiveness of the treatment regimen and the successful tapering of the corticosteroid dosage. In light of the continued improvement, a further modification was made to the methylprednisolone therapy. The dosage was tapered again, from 24 mg daily to 12 mg daily. This step-wise reduction in corticosteroid dosage aimed to gradually wean the patient off corticosteroid therapy while maintaining disease control. The reduced dosage of methylprednisolone was continued once daily for one week. At the week 5 follow-up visit, a final assessment of the skin lesions and pruritus was conducted. The patient exhibited further improvement, with near-complete resolution of erythema and scaling. Only trace residual erythema and minimal scaling were observed. The patient reported no itching, indicating a complete resolution of pruritus. These findings demonstrated a successful response to the treatment regimen, with the patient achieving significant clinical improvement. Based on the near-complete resolution of the patient's dermatological condition, the methylprednisolone therapy was discontinued entirely. This decision reflects the successful tapering of the corticosteroid dosage and the achievement of disease control without the need for ongoing corticosteroid therapy. The long-term management phase focused on maintaining disease remission, preventing disease flares, and addressing the chronic

nature of PRP. Methotrexate, which was initiated during the initial treatment phase, was continued as a long-term maintenance therapy. The dosage remained at 10 mg, administered orally once weekly. Methotrexate plays a crucial role in maintaining disease remission and preventing flares of PRP. Regular monitoring for potential side effects associated with long-term methotrexate therapy is essential. This includes periodic blood tests to assess liver function, complete blood count to monitor for bone marrow suppression, and other relevant investigations. Follow-up visits were scheduled monthly to assess the patient's clinical status and monitor for any adverse effects of methotrexate. Emollients, such as Vaseline album, were continued as a long-term topical therapy. The emollients were applied twice daily. The consistent use of emollients is crucial for maintaining skin hydration, supporting skin barrier function, and preventing dryness and scaling, which are common features of PRP. Patient education on proper emollient application and the importance of regular use is essential. Patient education and counseling were integral components of the long-term management plan, aimed at empowering the patient to actively participate in their care and minimize the risk of disease exacerbations. Given the patient's history of a previous episode associated with stress, stress management techniques were emphasized. The patient received education and counseling on various stress reduction strategies, such as relaxation techniques, mindfulness, and cognitive behavioral therapy. The importance of managing stress as a potential trigger for PRP exacerbation was highlighted. These discussions occurred at every visit to reinforce the importance of stress management. The patient received comprehensive education on proper skin care practices to maintain skin health and prevent flares of PRP. This included guidance on gentle cleansing, avoiding harsh soaps or detergents, using lukewarm water for bathing, and patting the skin dry rather than rubbing. The importance of consistent emollient use and recognizing early signs of a potential flare were emphasized. These recommendations were reinforced at every visit to ensure adherence and address any emerging concerns (Table 2).

3. Discussion

Erythroderma, characterized by widespread erythema and scaling affecting a large proportion of the body surface area, poses a significant diagnostic challenge due to its diverse range of underlying etiologies. As observed in this case, the initial presentation of erythroderma can obscure the characteristic features of specific dermatoses, making it difficult to arrive at a definitive diagnosis. The differential diagnosis of erythroderma is broad, encompassing a variety of conditions, including inflammatory skin diseases such as psoriasis, atopic dermatitis, and seborrheic dermatitis, as well as drug reactions, malignancies, and systemic disorders. In this particular case, the patient presented with a one-month history of erythroderma accompanied by pruritus, fever, and sleep disturbances. The extensive involvement of the skin, estimated to be greater than 90% of the body surface area, underscored the severity of the condition. The diagnostic process was further complicated by the fact that erythroderma can be the final common pathway for a variety of dermatological conditions, making it essential to carefully evaluate the patient's history, physical examination findings, and laboratory and histopathological investigations to determine the underlying cause. The importance of a thorough and detailed history in the evaluation of erythroderma cannot be overstated. In this case, the patient's history of a similar episode four months prior, associated with anxiety related to his father's stroke, provided a crucial clue to the potential role of stress in triggering or exacerbating his condition. This historical information, combined with the findings of the physical examination and subsequent investigations, ultimately led to the diagnosis of PRP.[11,12]

Table 1. Summary of patient findings.

Feature	Details
Anamnesis	
Age	18 years
Gender	Male
Occupation	Student
Chief complaint	Reddish patches and thick scales all over the body, accompanied by itching
Duration of illness	1 month
Associated symptoms	Fever, difficulty sleeping
History of similar illness	Yes, 4 months prior, associated with anxiety related to father's stroke
Family history of similar illness	No
Medications	None
Allergies	None
Social history	
Smoking	No
Alcohol consumption	No
Physical Examination (Figure 1)	
General appearance	
Vital signs	Within normal limits
Dermatologic examination	
Distribution of skin lesions	Generalized, involving >90% of body surface area
Morphology of skin lesions	Erythema, scaling, 'nappes claires', follicular papules
Palmoplantar involvement	Hyperkeratosis with yellowish scales
Scalp involvement	Alopecia areata
Eye involvement	Ectropion
Laboratory Investigations	
Complete blood count	
Hemoglobin	14.5 g/dL (normal range: 13.5-17.5 g/dL)
White blood cell count	8,000/μL (normal range: 4,000-11,000/μL)
Platelet count	250,000/μL (normal range: 150,000-450,000/μL)
Differential count	Neutrophils 60%, Lymphocytes 30%, Monocytes 5%, Eosinophils 5%
Erythrocyte sedimentation rate (ESR)	20 mm/hr (normal range: 0-15 mm/hr)
C-reactive protein (CRP)	5 mg/L (normal range: 0-5 mg/L)
Liver function tests	
Aspartate aminotransferase (AST)	25 U/L (normal range: 0-40 U/L)
Alanine aminotransferase (ALT)	30 U/L (normal range: 0-40 U/L)
Alkaline phosphatase (ALP)	80 U/L (normal range: 40-129 U/L)
Bilirubin	1 mg/dL (normal range: 0.3-1.2 mg/dL)
Renal function tests	
Blood urea nitrogen (BUN)	15 mg/dL (normal range: 7-20 mg/dL)
Creatinine	1 mg/dL (normal range: 0.7-1.3 mg/dL)
Urinalysis	Normal
Imaging Studies	
Chest X-ray	Normal
Histopathological Examination (Figure 2)	
Epidermis	Hyperkeratosis, parakeratosis, alternating orthokeratosis, and parakeratosis ('checkboard' pattern)
Dermis	Mild perivascular lymphocytic infiltrate
Clinical Diagnosis	
Primary diagnosis	Erythroderma secondary to pityriasis rubra pilaris
Differential diagnoses	Psoriasis
	Atopic dermatitis
	Seborrheic dermatitis
	Drug reaction

Table 2. Treatment and follow-up.

Phase	Treatment	Frequency	Duration	Follow-Up	Outcome
Initial Treatment					
Systemic	Methylprednisolone 48 mg	Once daily	2 weeks	-	-
	Methotrexate 10 mg	Once weekly	2 weeks	-	-
	Folic acid 5 mg	Once daily (except on days of MTX administration)	2 weeks	-	-
	Cetirizine 10 mg	Once daily	2 weeks	-	-
Topical	Desoximetasone cream 0.25%	Twice daily	2 weeks	-	-
	Vaseline album	Twice daily	2 weeks	-	-
Follow-Up (Week 3)					
Assessment	Clinical evaluation of skin lesions, severity of itching	-	-	-	Significant improvement in erythema and scaling; reduced itching
Treatment Modification	Methylprednisolone tapered to 24 mg daily	Once daily	1 week	-	-
Follow-Up (Week 4)					
Assessment	Clinical evaluation of skin lesions, severity of itching	-	-	-	Continued improvement; minimal erythema and scaling; minimal itching
Treatment Modification	Methylprednisolone tapered to 12 mg daily	Once daily	1 week	-	-
Follow-Up (Week 5)					
Assessment	Clinical evaluation of skin lesions, severity of itching	-	-	-	Further improvement; near-complete resolution of erythema and scaling; no itching
Treatment Modification	Methylprednisolone discontinued	-	-	-	-
Long-Term Management					
Systemic	Methotrexate 10 mg	Once weekly	Ongoing	Monthly	Maintenance of remission; monitoring for potential side effects of methotrexate
Topical	Emollients (e.g., Vaseline album)	Twice daily	Ongoing	-	Maintenance of skin hydration
Patient Education and Counseling					
Stress management techniques	-	-	-	Every visit	To minimize potential triggers for PRP exacerbation
Skin care recommendations	-	-	-	Every visit	To maintain skin health and prevent flares

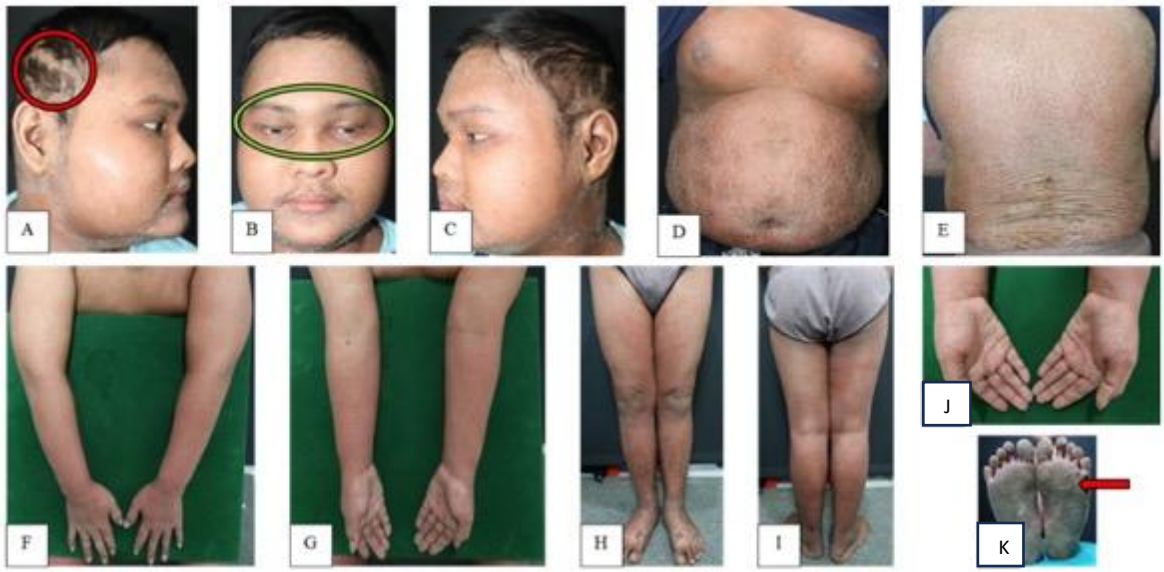

Figure 1. Clinical manifestations of the patient (A). The scalp region showed alopecia areata (red circle) (B). In the orbital region, there was an ectropion (green circle) (C-K). The generalized region shows discrete reddish patches and multiple papules, partially confluently formed nappes claires. (C-K) Bilateral palmoplantar region showing hyperkeratosis with yellowish-colored scales (red arrow).

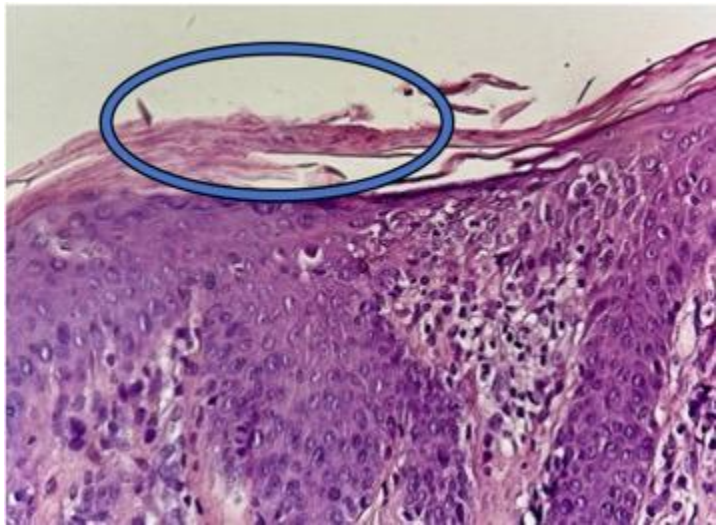

Figure 2. Histopathological examination. The epidermal layer shows hyperkeratosis, parakeratosis, and orthokeratosis in a checkboard pattern accompanied by focal acantholic dyskeratosis (blue circle) (100x magnification; H&E staining).

Pityriasis rubra pilaris is a relatively rare inflammatory skin disorder that can manifest as erythroderma. It is characterized by the development of reddish-orange plaques, follicular papules, and a distinctive pattern of sparing within the areas of erythema, known as "nappes claires." In cases where erythroderma is the predominant presentation, these characteristic features may be less apparent, making the diagnosis more challenging. In this case, the patient exhibited several clinical features suggestive of PRP. The presence of "nappes claires," although potentially subtle in the context of erythroderma, was a significant finding. These islands of relatively normal-appearing skin within the generalized

erythema are a hallmark of PRP and aid in differentiating it from other erythrodermic conditions. The patient also presented with follicular papules, which are small, raised bumps associated with hair follicles, further supporting the possibility of PRP. Palmoplantar involvement, characterized by hyperkeratosis with yellowish scales, was another notable clinical feature in this case. Palmoplantar keratoderma is a common manifestation of PRP, reflecting an abnormality in the keratinization process in these regions. The scalp involvement, manifested as alopecia areata, was an interesting finding. Alopecia areata is an autoimmune condition characterized by patchy hair loss, and its presence in this patient suggests the potential for associated autoimmune phenomena in PRP. Additionally, the patient exhibited ectropion, an outward turning of the eyelid margin. Ectropion in PRP may result from chronic inflammation and keratinization affecting the eyelid skin. Histopathological examination of skin biopsies plays a crucial role in confirming the diagnosis of PRP. The characteristic histopathological findings include hyperkeratosis, parakeratosis, and a "checkboard" pattern of alternating orthokeratosis and parakeratosis. Focal acantholytic dyskeratosis, which is premature separation of keratinocytes with abnormal keratinization may also be observed. In this case, the histopathological examination revealed hyperkeratosis, parakeratosis, alternating orthokeratosis, and parakeratosis in a "checkboard" pattern, accompanied by focal acantholytic dyskeratosis. These findings were consistent with the histopathological features of PRP and supported the clinical diagnosis.[13,14]

The etiology of PRP is complex and not fully understood, but it is believed to involve a combination of genetic predisposition, immune dysregulation, and environmental triggers. Stress has been implicated as a potential trigger for PRP exacerbation in some cases. In this case, the patient's history revealed a compelling link between stress and disease exacerbation. He reported a similar episode of erythroderma four months prior, which was associated with anxiety related to his father's stroke. The current presentation was also preceded by a period of stress related to school exams. These historical details suggest a strong correlation between psychological stress and the exacerbation or recurrence of PRP in this patient. The mechanisms by which stress may trigger or exacerbate PRP are not fully elucidated. However, it is postulated that stress-induced immune dysregulation may play a role. Stress can activate the hypothalamic-pituitary-adrenal (HPA) axis and the sympathetic nervous system, leading to the release of various hormones and neurotransmitters, such as cortisol and catecholamines. These stress mediators can influence immune function by affecting the production and activity of cytokines, chemokines, and other immune mediators. In susceptible individuals, stress-induced immune dysregulation may contribute to the development or exacerbation of inflammatory skin conditions like PRP. It is important to note that while stress may play a role in some cases of PRP, it is not the sole causative factor. Genetic predisposition and other environmental triggers are also likely to be involved. However, the recognition of stress as a potential trigger is crucial for the holistic management of PRP patients. Addressing stress through various stress management techniques may help to prevent or minimize disease exacerbations.[15,16]

The management of erythroderma secondary to PRP requires a multimodal therapeutic approach aimed at controlling inflammation, alleviating symptoms, and improving the patient's overall quality of life. This often involves a combination of systemic and topical therapies, tailored to the individual patient's needs and disease severity. In this case, the patient was initially treated with systemic corticosteroids, methotrexate, folic acid, and cetirizine, along with topical desoximetasone cream and Vaseline album. Systemic corticosteroids, such as methylprednisolone, are frequently used to achieve rapid control of the acute inflammatory process, particularly in cases of severe erythroderma. Corticosteroids exert potent anti-inflammatory effects by suppressing various components of the immune

system. In this case, methylprednisolone was initiated at a dosage of 48 mg daily, and the patient showed significant improvement in erythema and scaling. However, due to the potential for significant side effects with long-term use, systemic corticosteroids are typically used for short-term management or as a bridge to other therapies. In this case, the methylprednisolone dosage was gradually tapered as the patient's condition improved, and ultimately, the corticosteroid therapy was discontinued. Disease-modifying agents, such as methotrexate, are often necessary for long-term management of PRP, particularly in chronic or recalcitrant cases. Methotrexate is an antimetabolite that exhibits immunomodulatory properties. It has been shown to be effective in inducing and maintaining remission in PRP. In this case, methotrexate was initiated at a dosage of 10 mg weekly and continued as a long-term maintenance therapy. Regular monitoring for potential side effects, such as hepatotoxicity and bone marrow suppression, is essential in patients receiving methotrexate therapy. In addition to systemic therapies, topical treatments play an important role in the management of erythroderma secondary to PRP. Topical emollients are crucial for maintaining skin hydration and reducing dryness and scaling, which are prominent features of erythroderma. In this case, Vaseline album was used as an emollient and applied twice daily. Topical corticosteroids of appropriate potency can also be used to alleviate localized inflammation and itching. In this case, desoximetasone cream, a high-potency topical corticosteroid, was applied twice daily to affected skin areas.[17,18]

This case also highlights the potential for extracutaneous manifestations and associated conditions in PRP. The patient developed alopecia areata and ectropion during the course of his illness. Alopecia areata is an autoimmune condition characterized by patchy hair loss. Its association with PRP, as seen in this case, suggests a potential link between these two conditions, possibly mediated by shared immunopathogenic mechanisms. Further research is needed to fully elucidate the relationship between PRP and alopecia areata. Ectropion, the outward turning of the eyelid margin, is another noteworthy extracutaneous manifestation observed in this patient. In the context of PRP, ectropion may result from chronic inflammation and keratinization affecting the eyelid skin. The chronic inflammatory process can lead to scarring and contracture of the eyelid tissues, ultimately resulting in ectropion. The development of these extracutaneous manifestations underscores the systemic nature of PRP and the potential for involvement beyond the skin. Clinicians should be aware of these potential associations and carefully evaluate PRP patients for extracutaneous signs and symptoms.[19,20]

4. Conclusion

This case report elucidates the complexities inherent in diagnosing erythroderma, particularly when it masks the underlying pityriasis rubra pilaris (PRP). The case underscores the critical importance of a meticulous clinical examination, with a keen focus on subtle yet pathognomonic signs such as 'nappes claires', which are areas of sparing within the erythrodermic skin. A thorough history, including potential triggers like stress, is also crucial. The characteristic 'checkboard' pattern observed in histopathological examination definitively confirmed the diagnosis of PRP. This case further highlights the potential association of PRP with extracutaneous manifestations such as alopecia areata and ectropion, expanding the clinical spectrum of this rare condition. The successful management of this patient involved a combination of systemic corticosteroids for rapid control of inflammation and methotrexate for long-term disease management, alongside diligent supportive care with topical emollients. This case emphasizes the need for clinicians to be vigilant in recognizing PRP amidst the myriad causes of erythroderma and to consider the potential role of stress in disease exacerbations. By enhancing awareness of the diagnostic subtleties and therapeutic strategies for PRP-associated erythroderma, this

report contributes to improved clinical acumen in managing this challenging condition.

5. References

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