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Early Presentation of Pityriasis Rubra Pilaris Mimicking Tinea Corporis: Diagnostic Challenges of a Rare Skin Condition

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 Dayang Haniffa Abang Hashim
ACDE 1 Ilham Ameera Ismail
CD 2 Zahrah Tawil
ADE 1 Haizlene Abd Halim

1 Department of Primary Care Medicine, Universiti Teknologi MARA, Sungai Buloh, Selangor, Malaysia
2 Department of Pathology, Hospital Selayang, Ministry of Health, Selangor, Malaysia

Corresponding Author: Ilham Ameera Ismail, e-mail: ilham874@uitm.edu.my
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Patient: Female, 61-year-old
Final Diagnosis: Pityriasis rubra pilaris
Symptoms: Skin lesions • pruritis
Medication: —
Clinical Procedure: Skin biopsy
Specialty: Dermatology • Family Medicine • Pathology

Objective: Rare disease

Background: Pityriasis rubra pilaris (PRP) is a rare chronic inflammatory skin condition characterized by follicular, papulosquamous, reddish-orange scaling, palmoplantar keratoderma, and erythema with islands of sparing. Its heterogeneous clinical presentation makes the diagnosis of PRP quite challenging, especially at the initial presentation, as it can mimic common skin conditions.

Case Report: We present a case with an early presentation of PRP in a 61-year-old Malay woman with underlying uncontrolled diabetes, and discuss evolving clinical course of her disease. She presented to a primary care clinic with a 3-week history of itchy, ring-like skin lesions that started on her neck and chest but subsequently spread widely on her chest, back, and upper extremities. She was first treated as having extensive tinea corporis but responded poorly to multiple courses of antifungal treatment. An initial skin biopsy that was taken at the dermatology clinic revealed features suggestive of erythema annulare centrifugum. However, despite topical steroid treatment, her skin condition evolved further and she developed generalized erythroderma along with follicular hyperkeratosis and palmoplantar keratoderma. A repeat biopsy finally confirmed the diagnosis of PRP.

Conclusions: Making the diagnosis of PRP is challenging for clinicians. However, clinicians should approach any common skin problem that does not respond to treatment appropriately, with consideration of other uncommon skin disorders. A repeat skin biopsy may be considered if there are any doubts about the diagnosis. A clinical and histopathological correlation is important to aid in the diagnosis of PRP.

Keywords: Annular Erythema • Dermatology • Pityriasis Rubra Pilaris • Tinea

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936906

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Background

Pityriasis rubra pilaris (PRP) is a rare skin condition that was first described in the 1800s [1-3]. Due to the rarity of the disease, the worldwide incidence rate of PRP is not known. Approximately 1 in 5000 cases in an outpatient dermatology in the United Kingdom in 1980 was diagnosed with PRP [3], with no recently published data. In 2020, a Facebook PRP group has approximately 1500 members worldwide [4]. There are 6 clinical subtypes of PRP [3,5]; with the classical adult-type (Type I) being the commonest [6]. Other types are Type II, the atypical adult-type; Type III, the classical juvenile type; Type IV, the circumscribed juvenile type; Type V, the atypical juvenile type; and Type VI, which is PRP associated with HIV/AIDS. PRP can affect any race and can occur at any age and in either sex. Although there are cases of the familial autosomal dominant form of PRP [7] and reports have shown an association with drugs such as tyrosine kinase inhibitors and autoimmune diseases such as myasthenia gravis, autoimmune thyroiditis, and celiac disease [8,9], the exact etiology and pathogenesis of PRP remain unclear. PRP has been reported in patients with concurrent medical conditions such as diabetic mellitus, allergic rhinitis, dyslipidemia, depression, arthritis, cardiovascular diseases, hypothyroidism, malignancies, HIV, and other skin conditions [5,10,11].

PRP usually presents with 2 primary features – follicular hyperkeratotic papules and palmoplantar keratoderma – but it can have a range of varying clinical presentations. The typical clinical presentation is the appearance of small follicular hyperkeratotic papules, with a yellowish ring surrounding a central, smooth keratotic plug. This appearance is termed "nutmeg-grater sign". It can also present as inflamed plaques with islands of sparing, pityriasiform scales, and erythroderma. Other initial clinical presentations include erythema of the extensor surfaces of knees and elbows, skin lichenification, ichthyosis, eczema-like skin changes, hair loss, and nail thickening or discoloration [10]. The diagnosis of PRP is made by correlating the clinical and histopathologic features. The rarity of PRP and its varied range of clinical presentations make the diagnosis of the condition quite challenging, especially in the primary care setting, where the diagnosis is often delayed. Only approximately half of classic PRP patients were correctly diagnosed when they presented early [11]. The most common misdiagnoses include psoriasis, contact dermatitis, and eczema or spongiotic dermatitis [10]. About 2% of PRP patients were initially diagnosed as having tinea [10].

Case Report

A 61-year-old woman presented to a primary care outpatient clinic with a 3-week history of multiple small, red, itchy, ring-like spots that initially appeared on her face and neck. The rash then increased and spread widely throughout her whole body. She visited 2 primary care clinics and was twice treated unsuccessfully with short-course topical miconazole 1% cream. She had underlying allergic rhinitis, dyslipidemia, and poorly-controlled diabetes, with HbA1c 10.5%. Her medications were oral Janumet (metformin/sitagliptin), gliclazide MR, atorvastatin, loratadine, and budesonide nasal spray. There was no history of taking any new medications and she did not have any drug allergies. She was a non-smoker.

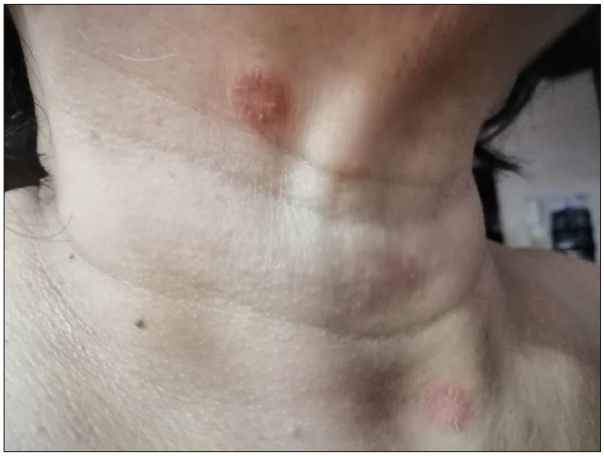

Figure 1. Initial presentation: Well-demarcated, erythematous, rounded, scaly, annular-like lesions mimicking tinea corporis.

On examination, there were raised, well-demarcated, erythematous, rounded, annular-like lesions on her face, neck, chest, abdomen, back, and upper extremities. Some of the lesions had reddish-orange periphery scales (**Figure 1**). Results of other system examinations were unremarkable. The diagnosis of extensive tinea corporis was made based on physical examination findings and her underlying poorly-controlled diabetes. She was given oral fluconazole 200 mg weekly and was referred to an outpatient dermatology clinic.

She was seen at the dermatology clinic 1 month later. Her skin condition had not improved, despite the course of oral fluconazole. A skin biopsy was taken, and investigations including a complete blood count, renal profile, liver function test, ANA, C3, C4, and viral screening. A skin scraping for fungal culture and sensitivity was also taken and it was negative. Autoimmune and viral screening results were negative, and all other blood investigation results were within normal ranges.

The first skin biopsy result suggested a possible diagnosis of erythema annulare centrifugum (EAC) (**Figure 2**). She was subsequently prescribed betamethasone valerate 0.1% topical cream twice daily over her body and clobetasone 0.05% topical

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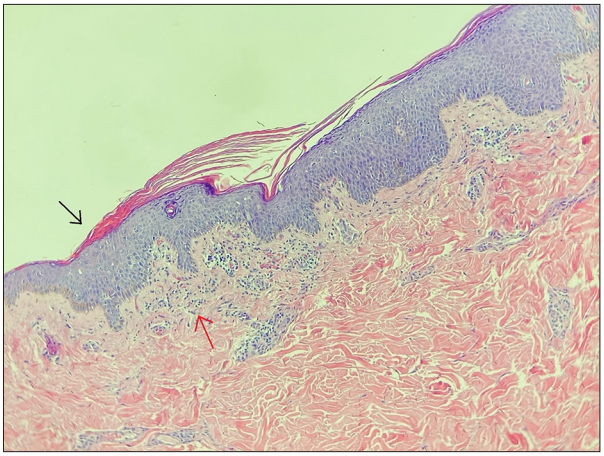

Figure 2. First biopsy. Section of the skin shows an epidermal layer with focal parakeratosis (black arrow), mild spongiosis, and perivascular lymphoid aggregate (red arrow), which are features suggestive of EAC.

cream twice daily over her face for a month. She was also given aqueous cream emollient and oral cetirizine 10 mg daily.

However, the rash did not respond to the treatment. She visited a private primary care clinic due to worsening skin lesions and severe pruritus. She was given a course of oral clarithromycin 500 mg twice daily. She then developed widespread erythroderma 3 days after taking the antibiotics and was promptly admitted to the hospital due to a suspicion of severe antibiotic allergy. In the ward, her skin condition evolved to generalized erythroderma with islands of sparing (**Figure 3**). There

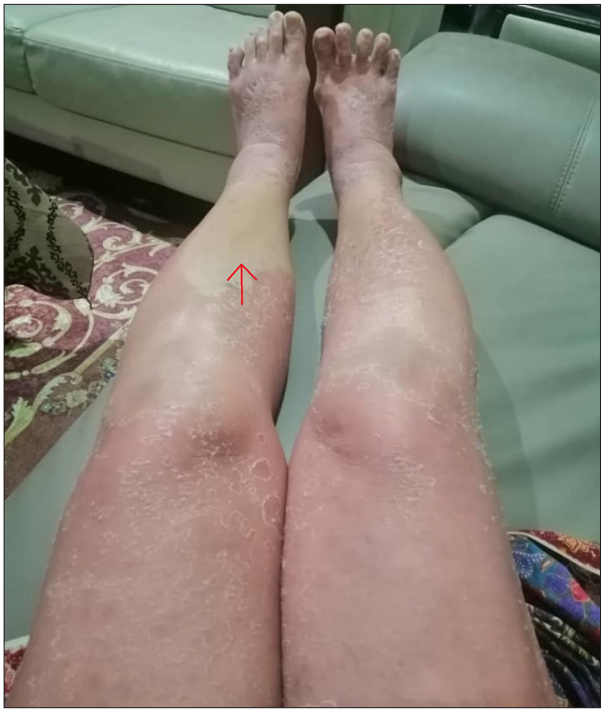

Figure 3. Erythroderma with islands of sparing (red arrow) on the lower limbs.

was waxy keratoderma on the palms and soles, which started to desquamate after a few days of treatment (**Figure 4**). There were also orange-red plaques with follicular prominences, but they were poorly appreciated after initiating treatment. The new evolving presentation of skin features suggested the

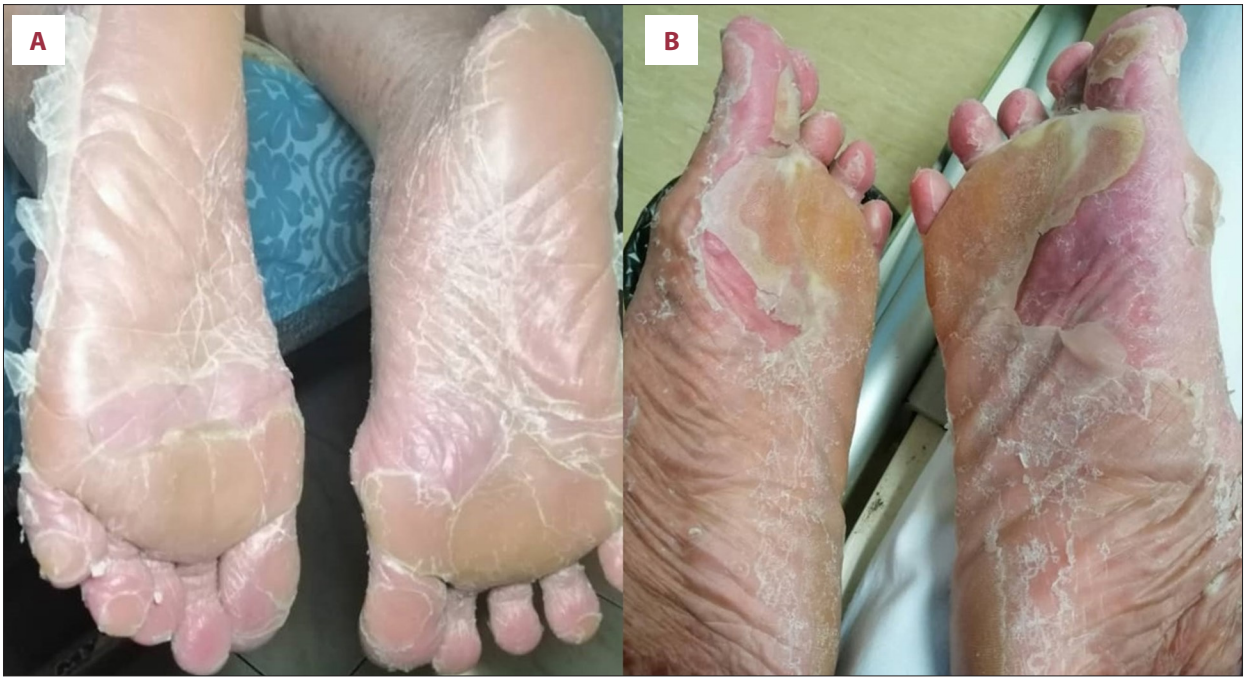

Figure 4. Palmoplantar waxy keratoderma (**A**) and desquamation (**B**) on the soles.

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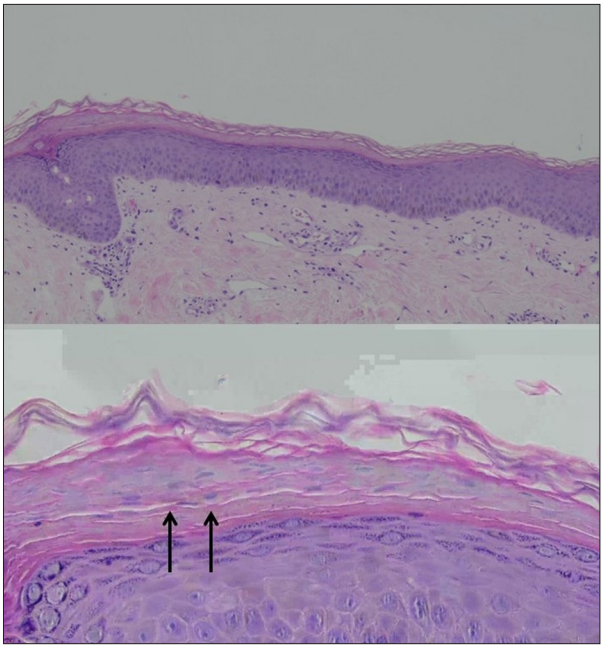

Figure 5. Second biopsy: Acanthosis, spongiosis, focal prominent granular layer, thick suprapapillary plate. The dermis shows perivascular lymphoplasmacytic and occasional eosinophilic infiltration. The stratum corneum shows a vague alternating area of orthokeratoses and parakeratoses (double arrows), which are features consistent with pityriasis rubra pilaris.

diagnosis of classical adult-type pityriasis rubra pilaris. A second skin biopsy was performed. The correlation of clinical and histological findings (**Figure 5**) confirmed the diagnosis of pityriasis rubra pilaris. She was initially treated with methotrexate but there was minimal response. She was then switched to oral acitretin 25 mg daily and the rash began to resolve.

The diagnosis of PRP was made 4 months after the initial presentation. Three months after the diagnosis of PRP and appropriate treatment, her skin condition improved. After 6 months of acitretin therapy, her PRP was in remission (**Figure 6**).

Discussion

The patient presented to a primary care clinic at an early stage of PRP with lesions that were similar to tinea corporis. Tinea corporis is a skin fungal infection that is common in tropical regions where the weather is warm and humid [12], such as Malaysia. In addition, the patient also has poorly-controlled diabetes, a known predisposing factor for tinea infection. The diagnosis of tinea corporis can be made with microscopic examination of skin scrapings stained with potassium hydrochloride (KOH). However, tinea corporis is usually diagnosed clinically, with the findings of typical pruritic, well-demarcated, erythematous, annular, raised scaling patches with central clearing [13].

There are no known serologic or immunohistochemical markers to aid or confirm the diagnosis of PRP [6]. Although additional tests are often done to rule out other possible differential diagnoses, the results are often normal. The diagnosis of PRP is made by correlating the clinical findings and the histopathological features. However, in the present patient, the diagnosis was challenging in the beginning as the clinical findings mimicked tinea corporis, and the initial histologic findings did not demonstrate a clear diagnostic picture of PRP.

Almost 54% of PRP patients had 2 or more biopsies before the diagnosis was established [10]. Our patient underwent 2 separate skin biopsies before the diagnosis of PRP was confirmed. The first biopsy had similar morphology as the superficial variant of EAC (**Figure 2**). EAC is typically characterized histologically by perivascular lymphocytic infiltrate of lymphocytes with a "coat sleeve" appearance. The superficial type is characterized by epidermal changes like focal spongiosis,

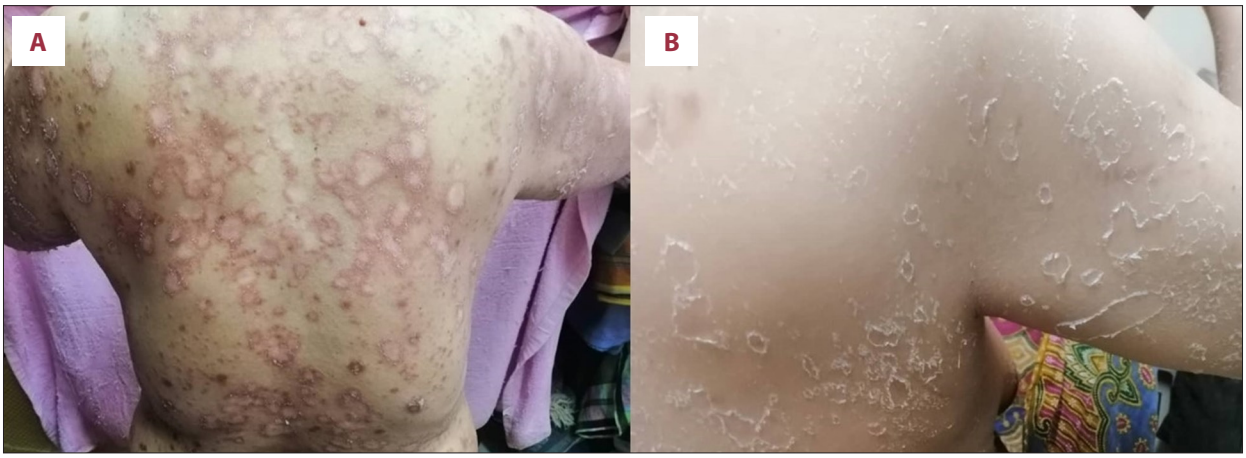

Figure 6. Treatment response. Skin condition after 2 weeks of acitretin therapy (**A**), and when the patient was entering remission (**B**).

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focal parakeratosis, epidermal hyperplasia, and papillary dermal edema [14]. The second biopsy was taken when the patient had started to develop erythroderma and palmoplantar keratoderma, manifesting the features of classical adult PRP. The morphology was well-developed and more specific. Considering the patient's evolving clinical features and correlating with these biopsy findings, the diagnosis of PRP could then be confirmed.

The treatment of PRP consists of topical and systemic therapies. In mild-to-moderate disease, especially for localized types of PRP, topical treatment with corticosteroids, calcipotriol, tazarotene, and emollients may be adequate. Moderate-to-severe diseases are treated with systemic therapy in addition to topical treatment [11]. The first-line therapy is retinoids such as isotretinoin and acitretin. Other systemic therapies include methotrexate, cyclosporine, and combinations of acitretin with narrowband-UVB, UVA1, or PUVA [11]. Our patient was initially treated with methotrexate due to the availability and cost. However, she did not respond well to the treatment. Her treatment was successful after switching to acitretin, and the skin lesions showed marked improvement.

The diagnosis of PRP is often delayed and can seriously affect a patient's life [15]. The skin lesions may worsen and evolve despite being treated by multiple doctors. The itchiness and burning sensation on the skin can affect activities of daily living. Therefore, a prompt diagnosis with appropriate management is important to improve the patients' overall quality of life [15].

Conclusions

PRP is a rare chronic inflammatory skin disorder with varying clinical presentations, often similar to some common skin problems. Making the diagnosis of PRP has proven to be challenging for clinicians. Clinicians should have a high index of suspicion toward any common skin problems that do not respond to treatment appropriately, with consideration for other rarer skin disorders such as PRP. Close follow-up of unresolved skin lesions is recommended to monitor for evolving skin changes that could aid in the diagnosis.

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Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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