Clues in the Diagnosis of Pityriasis Rubra Pilaris: A Case Report and Literature Review Comparing it to Psoriasis

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ABSTRACT:
Pityriasis rubra pilaris (PRP) is a rare papulosquamous skin disease characterized by palmoplantar keratoderma, erythroderma, orange plaques amidst normal skin and nail changes. A severe case of PRP can mimic erythrodermic psoriasis; therefore, accurate diagnosis is challenging. Clinical presentation, dermoscopic findings, and skin biopsy play pivotal roles in differentiating PRP from psoriasis. We present a case report and review the literature about the imperative diagnostic clues to distinguish PRP from psoriasis.

Key words: Pityriasis rubra pilaris, Psoriasis, Papulosquamous disease

Introduction

Pityriasis rubra pilaris (PRP) is a rare papulosquamous disease. Based on the age of onset, clinical features, and prognosis, PRP can be classified into six subtypes. The most common type is type I (classic adult) which presents with erythematous plaque, palmoplantar keratoderma, nail plate thickening with subungual hyperkeratosis, and nail discoloration. In a severe case, the lesion may coalesce and turn into erythrodermic state, in which scattering areas of normal skin (so-called "islands of normal skin/sparing or skip areas") surrounded by orange plaque[1]. Psoriasis can present as erythematous plaque with silvery scale together with nail abnormality; hence, their similarity may lead to differential-diagnostic confusion[2]. Herein, we demonstrate a diagnostic pitfall of a case representing papulosquamous diseases and also the important clinical clues in distinguishing between PRP and psoriasis.

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Case report

39-year-old Thai woman presented with a persistent rash that had troubled her for over a decade. She had previously tried herbal remedies for a year, along with acitretin for an unknown duration, all without experiencing any clinical improvement. Approximately five years later, she sought a second opinion at a hospital, leading to a thorough physical examination. Clinical observations revealed the presence of large, erythematous, non-scaly plaques on her trunk and limbs. Some of her nails also displayed distal subungual hyperkeratosis, with no signs of hair and oral mucosal involvement. She was diagnosed with psoriasis vulgaris at this point of time and had been treated with a weekly dose of methotrexate (MTX) at 10 mg and topical Liquor Carbonis Detergens in 0.02% triamcinolone acetonide twice a day. Besides these treatments, mild topical steroids were recommended for the flexural regions of her body. However, her condition did not improve. Consequently, MTX dosage was increased to 15 mg weekly, resulting in a noticeable improvement in her lesions so she had continued taking the weekly dose of MTX.

Approximately one year later, she was hospitalized because of cystitis and she did not receive any treatment for her skin condition then the lesion got worse. She experienced a severe, progressively itchy rash. Upon another physical examination, she exhibited multiple scaly erythematous patches on the face, scalp, trunk, and extremities. By PASI-score assessment, the lesion yielded a value of 6.60 (Figure 1). Additionally, numerous follicular papules were observed on the back, and palmoplantar keratoderma on both hands. At this point, her skin condition was suspicious of pityriasis rubra pilaris (PRP). Consequently, a punch biopsy was conducted. The pathological finding revealed an acanthotic epidermis with marked hyperkeratosis, alternating ortho-parakeratosis, and superficial perivascular lymphocytic infiltration in the dermis (Figure 2). The diagnosis of PRP was confirmed. She was then placed under a vitamin A derivative (acitretin), a moderate-potency steroid, along with symptomatic treatment.

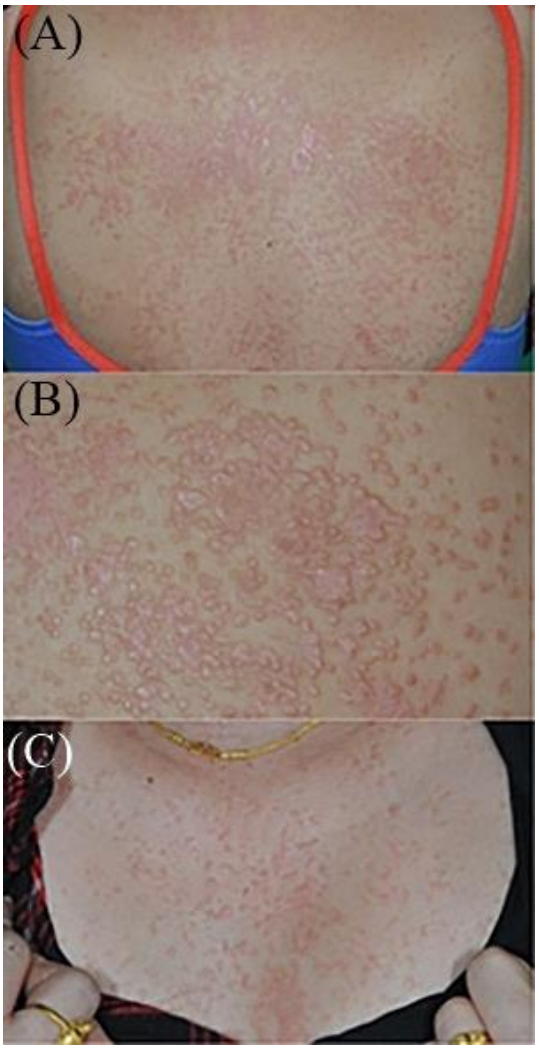

Figure 1 The clinical manifestation in the patient was scaly erythematous plaques on the upper back (A,B), multiple follicular papules on the upper back (B) and upper chest (C)

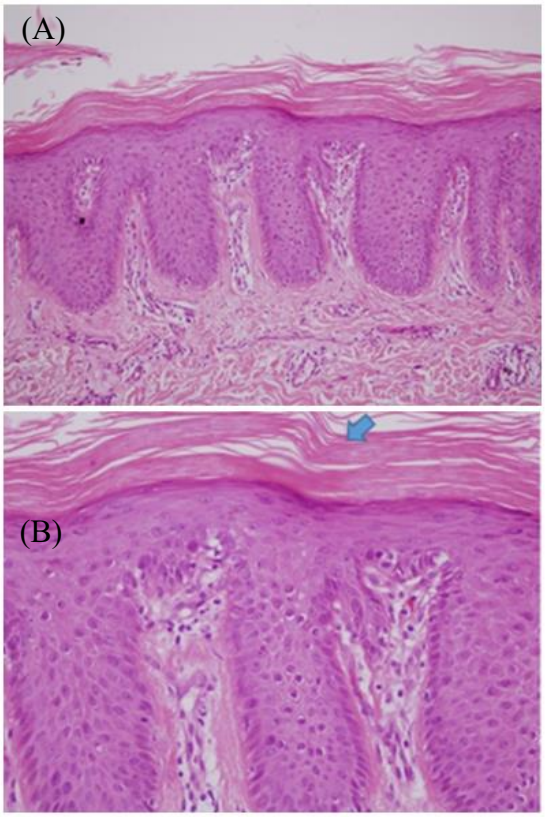

Figure 2 The histopathological findings (H&E (A): 20x, (B): 60x) revealed acanthotic epidermis with marked hyperkeratosis, alternating ortho-parakeratosis, superficial perivascular lymphocytic infiltration in the dermis (arrow depicts alternating ortho-parakeratosis)

Discussion

The differential diagnosis of PRP from other papulosquamous diseases is crucial; particularly from psoriasis because they need different treatments. Severe forms of erythroderma can manifest in the early stage of papulosquamous diseases, including psoriasis and PRP leading to mistaken of one disease for another, as demonstrated in this case. The diagnosis of PRP is primarily based on the clinical manifestation and pathological findings. Therefore, a skin biopsy is essential for a case suspected of PRP. The dermoscopic study has been recently used to obtain information useful for differentiation between PRP and psoriasis. The comparison of clinical clues, and dermoscopic and histopathologic findings are shown in Table 1[2].

The diagnosis for PRP varies greatly depending on its subtypes which are classified into six subtypes according to the modified Griffiths classification. The most common type is type I in which well-defined erythematous plaques, islands of normal skin- the pathognomonic sign, papules at the follicle, and palmoplantar keratoderma[3,4]. Characteristic hyperkeratotic follicular papules are prominent within the erythematous lesion and uninvolved skin. In contrast, the erythematous patches or plaque with a silvery scale, and bleeding points found in the lesion (so-called “Auspitz sign”) favor the diagnosis of psoriasis. Although the similarity of the erythrodermic state of these two diseases is problematic in diagnosis, the search for typical signs of PRP, as mentioned above, will help to differentiate between them.

Nail involvement in psoriasis can occur at the nail plate resulting nail pittings, thickening/fragility of the nail plate, and at the nail bed resulting in oil spots, distal subungual hyperkeratosis, and splinter hemorrhage. Although both diseases can have similar nail changes; however, pitting nails is not typical for PRP[5,6].

Furthermore, we can use dermoscopy to visualize tiny components of skin lesions. For psoriatic lesions, vascular patterns are red globules and ring-like vessels on a light red background, diffuse white scale patterns primarily found in the scalp, palmar, and plantar area[7]. For PRP lesions, the vascular pattern is dot or linear on a yellow-to-red background with yellowish and white scales[2].

Table 1 The comparison between clinical manifestations, histopathologic and dermoscopic findings of PRP and psoriasis

Descriptions	Disease	
	Psoriasis	**PRP**
Clinical manifestations	- Erythroderma in erythrodermic psoriasis - Well-defined erythematous plaque with white scale - Auspitz sign (bleeding spots) - *Nail involvement:* pitting nails, oil spots, hyperkeratosis at the subungual area	- Islands of normal skin - Palmoplantar keratoderma - Hyperkeratotic follicular papule (plaque) - Salmon-to-red plaques - Cephalocaudal progression - *Nail involvement*: nail plate thickening, and yellowish-to-brownish color
Pathologic findings	-Prolongation of epidermal rete ridges -Epidermal thickening -PMNs accumulation in the epidermis -Suprapapillary thinning of epidermis -Parakeratosis -Absence of the granular layer	-Acanthosis of the epidermis -Orthokeratosis with alternating spotty parakeratosis -Dermal infiltration of the lymphohistiocyte -Normal granular layer
Dermoscopic findings	-Vascular patterns: red globules, and ring-like vessels on a light red background -Diffuse white scale patterns	-Vascular patterns: dot or linear on a yellow-to-red background -Yellowish and white scales

List abbreviations: PRP, pityriasis rubra pilaris; PMNs, polymorphonuclear neutrophils

As mentioned earlier, histopathologic study give more information that will enable a physician to be more confident in distinguishing PRP from psoriasis. Nowadays it still remains the gold standard for diagnosis of PRP. The pathological changes in psoriasis are observed in both epidermis and dermis reflecting involvement of epidermal kinesis and inflammatory process. The epidermis becomes thickening with rete ridges prolongation (especially in plaque type). Parakeratosis is seen throughout the stratum corneum. Suprapapillary thinning as well as the absence of the granular layer are typical for psoriasis. The involvement of the inflammatory process is evident by the migration neutrophils (PMNs) from the dermis into the epidermis. The foci of PMNs (so-called "Munro's microabscess") are seen in the parakeratotic stratum corenum and the spinous layer (so-called "spongiform micropustule of Kogoj")[8]. On the contrary, PRP shows acanthosis of the epidermis, orthokeratosis with alternating spotty parakeratosis, dermal infiltration of the lymphohistiocyte, and completion of the granular layer[9].

Conclusion

Accurate diagnosis of PRP relies on a combination of clinical, dermoscopic, and histopathological features. Careful consideration of the patient's medical history, lesion distribution, and meticulous dermatological examination is crucial to avoid misdiagnosing PRP as psoriasis.

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