The conundrum of pediatric papulosquamous disorders: a tale of two cases

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Received: 30 July 2020
Accepted: 17 October 2020

Papulosquamous disorders constitute one of the common pediatric dermatoses. They are a heterogeneous group consisting of psoriasis, eczema, pityriasis rubra pilaris, and other conditions, where the differential diagnoses are close to each other clinically. Moreover, the presentation of the same disease in children can differ from that occurring in adults. We report two infants with erythematous scaly papules and plaques diagnosed as two different disorders, albeit with a grossly similar appearance. Both these dermatoses are considered to be quite uncommon in infancy and can pose significant challenges in management**.** These disorders sometimes present with atypical variants, rendering it increasingly difficult to distinguish them. Some of these disorders tend to progress to erythroderma, and the infant may occasionally present directly in the erythrodermic stage, further compounding the conundrum. In such situations, identification of subtle clinical and histopathological clues allows accurate diagnosis, which is imperative for appropriate prognostication and treatment. This article illustrates the importance of histopathology and immunohistochemistry in aiding diagnosis and outlines the management of these disorders while emphasizing the importance of timely intervention in averting inadvertent complications in these young individuals.

Keywords: pediatric, papulosquamous, psoriasis, pityriasis rubra pilaris

Iran J Dermatol 2022; 25: 76-81

DOI: 10.22034/ijd.2020.241056.1180

INTRODUCTION

Papulosquamous disorders constitute about 9-10% of all pediatric dermatoses [1]. They are a heterogeneous group comprising a vast array of specific conditions like psoriasis [2], pityriasis rubra pilaris, keratosis follicularis, eczemas, and ichthyosis, many of which might mimic each other clinically. Moreover, the presentation of the same disease in children can differ from that occurring in adults. Individual disorders sometimes present with atypical variants, making it increasingly difficult to distinguish between them. Some of these disorders tend to progress to erythroderma, and the infant may occasionally present directly in the erythrodermic stage, further compounding the conundrum. In such situations, identification of subtle clinical and histopathological clues allows an accurate diagnosis, which is imperative for appropriate prognostication and treatment. Herein, we report two infants with erythematous scaly papules and plaques diagnosed as two different disorders, albeit with a grossly similar appearance. Both these dermatoses are considered to be quite uncommon in infancy and can pose significant challenges in management. They have a protracted relapsing course and can negatively impact the individual's quality of life.

CASE PRESENTATION

Case 1

A one-year-old female child, the second product of a non-consanguineous marriage, presented with scaly rashes that had appeared 15 days beforehand. The lesions had first appeared over the scalp and later involved the rest of the body. There was no significant personal or family history. On examination, she had multiple erythematous, scaly plaques over the trunk, back, groin, and scalp, also involving the retro auricular areas (Figure 1). All the laboratory parameters were within normal limits. She was diagnosed with sebo-psoriasis and started on beclomethasone (0.025%) and clotrimazole (1%) combined lotion for the scalp and beclomethasone (0.025%) cream with white soft paraffin for truncal lesions along with a 25 mg fluconazole tablet daily for seven days. The child is under regular follow-up with reduced erythema and scaling.

Case 2

A seven-month-old male child, the second product of a non-consanguineous marriage, presented with rashes that had appeared 15 days earlier. There was no history of lesions at birth (collodion membrane) or significant family history. Examination revealed multiple well-defined scaly erythematous follicular papules and plaques over the face, scalp, extremities, and lower back with palmoplantar hyperkeratosis extending to the lateral and dorsal aspects of the hands and feet (Figure 2). Koebnerization and islands of normal skin were noted (Figure 3). Differential diagnoses considered were psoriasis, pityriasis rubra pilaris, erythrokeratodermia variabilis, and progressive symmetric erythrokeratodermia. The baby was treated with white petroleum jelly, beclomethasone (0.025%) cream, and liquid paraffin. Despite this, the lesions progressed to erythroderma over the next 20 days. The mucosal examination was unremarkable. Laboratory investigations showed anemia (8.9 gm%) and leucocytosis (13,000 cells/mm^3). Histopathology

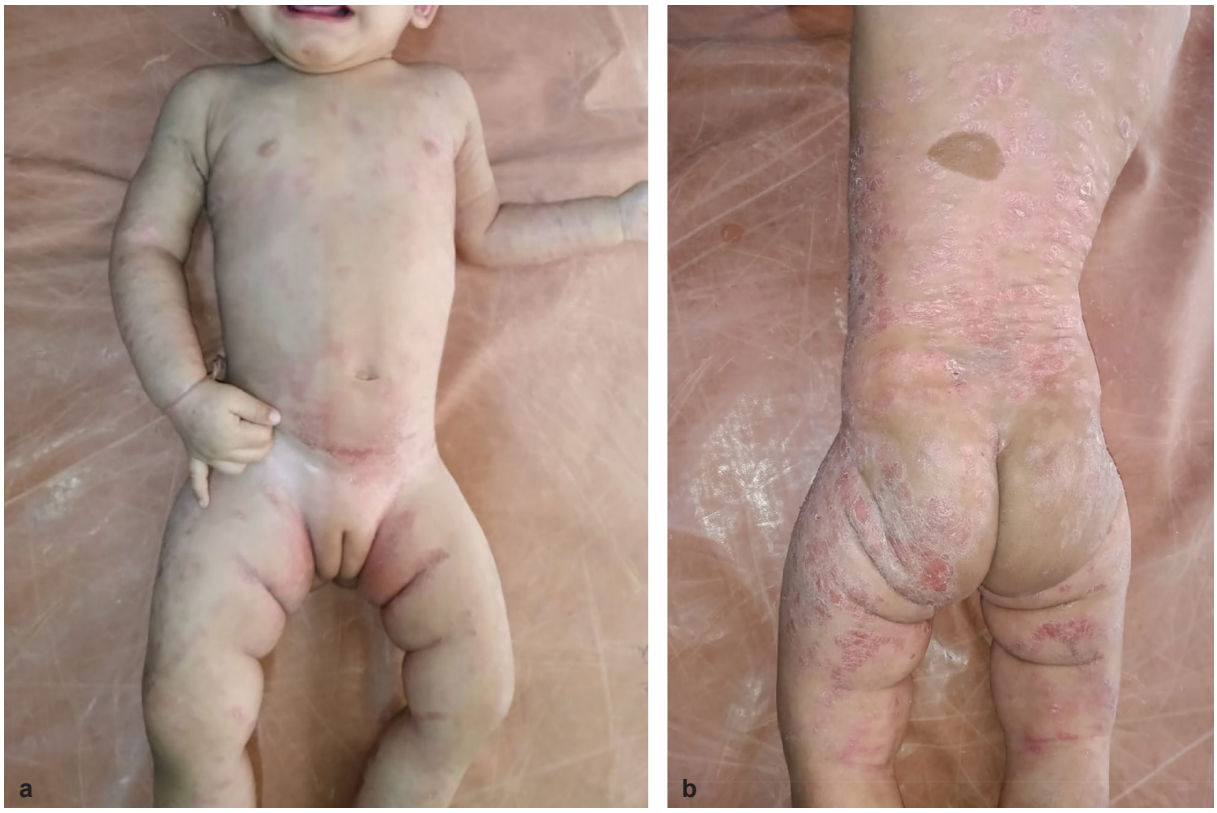

Figure 1. (a, b) First case showing multiple erythematous scaly plaques over the groins, buttocks, back, and posterior thighs.

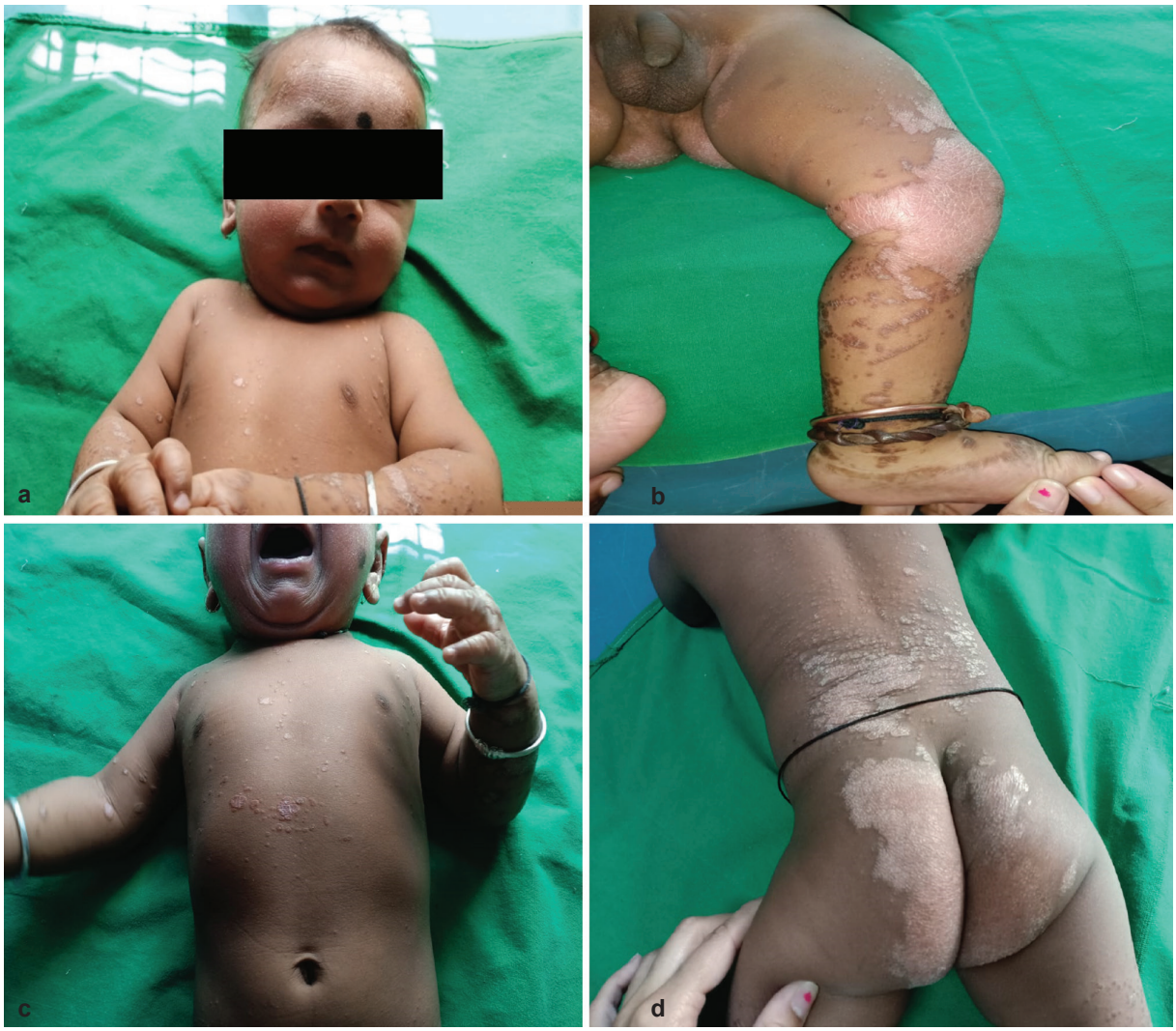

Figure 2. (a-d) Case 2 at first visit: Multiple erythematous scaly papules and well-defined plaques over the face, knees (with Koebnerization), trunk, and buttocks.

was consistent with non-psoriasis psoriasiform (Figure 4a,b). Immunohistochemistry (Ki67) showed increased expression in the suprabasal and basal layers (Figure 4c). The final diagnosis of juvenile pityriasis rubra pilaris (Type III) was made based on the clinicopathologic correlation. Prednisolone (0.5 mg/kg syrup) was started with tapering every fifth day along with intravenous amoxicillin-clavulanic acid and maintenance of adequate hydration and nutritional supplementation. He was administered oral vitamin A (25000 IU twice a week) and emollients for five months until all his skin lesions subsided (Figure 5a-c). The child also received a dose of oral vitamin A vaccine (100,000 IU) as part of the WHO immunization program. After five months of the above regimen, oral vitamin A was also discontinued. Subsequently, he is in complete remission, maintained on bland emollients.

DISCUSSION

Although psoriasis affects 0.5 – 2% of children and is the most common pediatric papulosquamous disorder, infantile psoriasis (Case 1) is relatively rare [2-4]. The initial lesions, manifesting as recalcitrant diaper dermatitis, can be atypical and misleading. In contrast with adults, there is greater

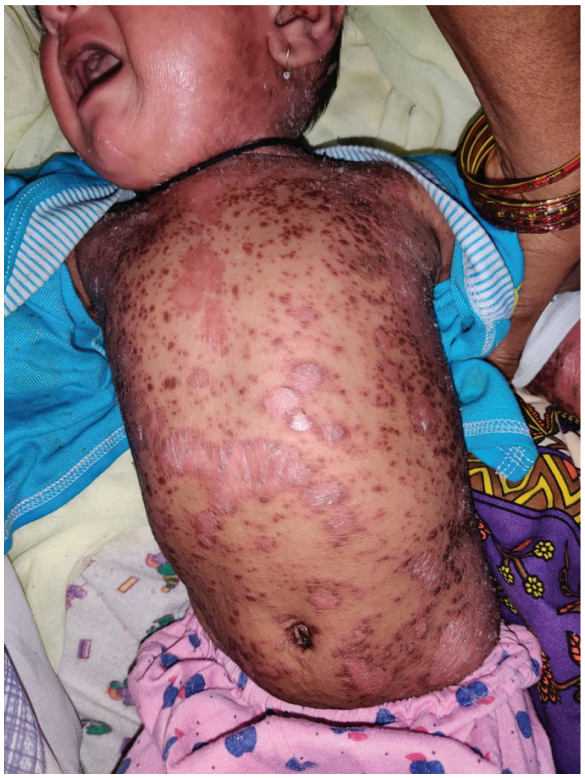

Figure 3. Second case showing multiple erythematous scaly follicular papules and plaques with islands of normal skin in between.

involvement of the scalp (58%), face, ears, and flexors, with the lesions being relatively thinner, softer, and less scaly. Psoriasis is often associated with seborrheic dermatitis (as in our case). Localized disease can be managed with topical medications like emollients and corticosteroids. Rapidly evolving, severe disease warrants the use of systemic modalities like methotrexate, cyclosporine, and acitretin.

Erythroderma has been extensively studied in adults, but the condition remains elusive in the pediatric population. Few studies have found infections like staphylococcal scalded skin syndrome (SSSS) and candidiasis, ichthyosiform erythroderma, and infantile seborrheic dermatitis as the leading causes of erythroderma in children [5].

Managing pediatric erythroderma is challenging. These children are at risk for potentially life-threatening complications like hypernatremic dehydration and hyperpyrexia. Careful monitoring of vital signs and maintenance of euthermia and fluid and electrolyte balance are essential. Other modalities include the application of emollients, wet dressings, topical steroids (limited areas only), and systemic antibiotics.

Pityriasis rubra pilaris (PRP, Devergie's disease, lichen ruber acuminatus) is an uncommon papulosquamous disorder with unknown etiology. It is classified into six types based on clinical appearance, age of onset, and prognosis. Clinically, it is characterized by pityriasiform scaling, palmoplantar keratoderma, and erythematous hyperkeratotic perifollicular papules, which may progress to plaques or erythroderma as seen in our second case. Juvenile PRP is categorized into three types (classical, circumscribed, and atypical). Although the relationship between PRP and psoriasis is not clear, they share clinical and histopathological similarities, so lesions of PRP are easily misdiagnosed as psoriasis. The absence of Auspitz and candle grease signs in PRP are instant diagnostic clues. The salient histopathological features (which differ according to stage of evolution) of PRP are alternating ortho and parakeratosis, hypergranulosis, irregular acanthosis, thick suprapapillary plates, and sparse lymphocytic perivascular infiltrate and follicular plugging. Munro's microabscesses and suprapapillary thinning (classic features of psoriasis) are conspicuously absent. Despite this, the findings are often reported as psoriasiform dermatosis, while the differential diagnosis is even more difficult in erythrodermic patients.

The causes of non-psoriasis psoriasiform dermatoses in children include seborrheic dermatitis, PRP, psoriasiform syphilids, pityriasis rosea, and pityriasis lichenoides. Lesional upregulation of Ki67 (a marker to detect S, G2, and M phases of cell cycle), as demonstrated in our second case, supports the view that PRP is a hyperproliferative disorder of epidermal keratinocytes [6]. The significantly higher expression of these indices in psoriasis can be utilized to confirm doubtful cases. Juvenile PRP can be treated with topical therapy alone, as this variant (Type III) is expected to undergo natural remission within one to two years. Hence the consequences of overzealous treatment should be considered. However, our patient did not respond to topical corticosteroids and emollients and progressed to erythroderma. As prednisolone led to only transient improvement, we contemplated starting either methotrexate or cyclosporine in an attempt to

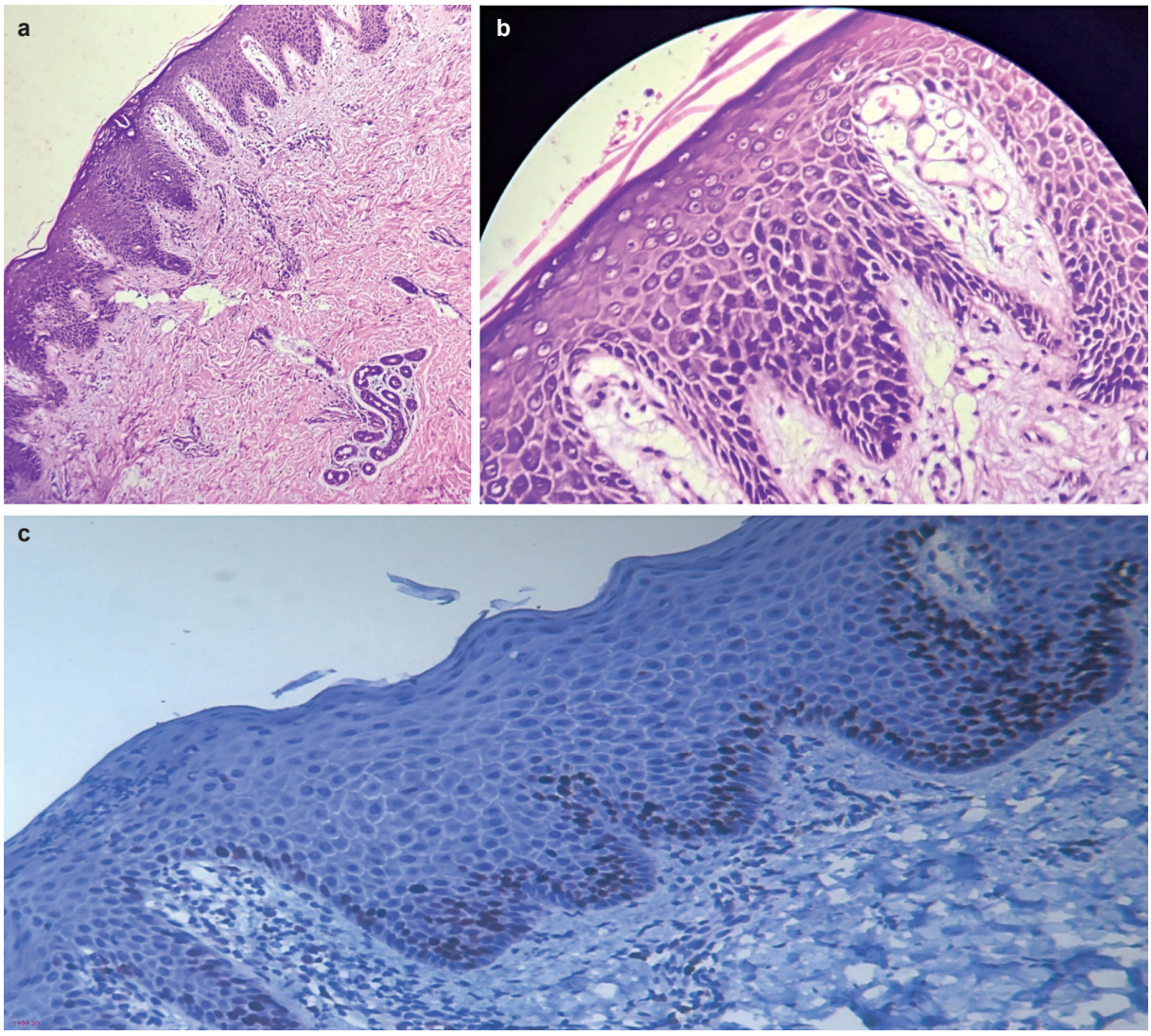

Figure 4. (a) Histopathology of an erythematous plaque from the second case showing alternating orthokeratosis and parakeratosis and regular elongation of rete ridges with follicular plugging (H&E stain, ×10). (b) Histopathology from an erythematous plaque of the second case demonstrated thick suprapapillary plates and absence of Munro's microabscesses (features compatible with the clinical diagnosis of pityriasis rubra pilaris while ruling out psoriasis) (H&E stain, ×40). (c) Immunohistochemistry Ki-67 marker in second case showing increased proliferation in the suprabasal and basal layers.

achieve rapid and sustained resolution. However, both these drugs were precluded due to anemia and the concurrent outbreak of the coronavirus disease 2019 pandemic. Fortunately, the child responded remarkably well to therapeutic doses of vitamin A.

CONCLUSION

Papulosquamous lesions in infancy can be a diagnostic dilemma and therapeutic challenge, particularly in the erythrodermic stage. Meticulous clinical and histopathological examination with immunohistochemistry (in selected cases) is the key to appropriate management. Despite the availability of multiple therapeutic options for infantile psoriasis and juvenile PRP, the choice of therapy should be dictated by the risk-benefit ratio.

Conflict of interest: None declared.

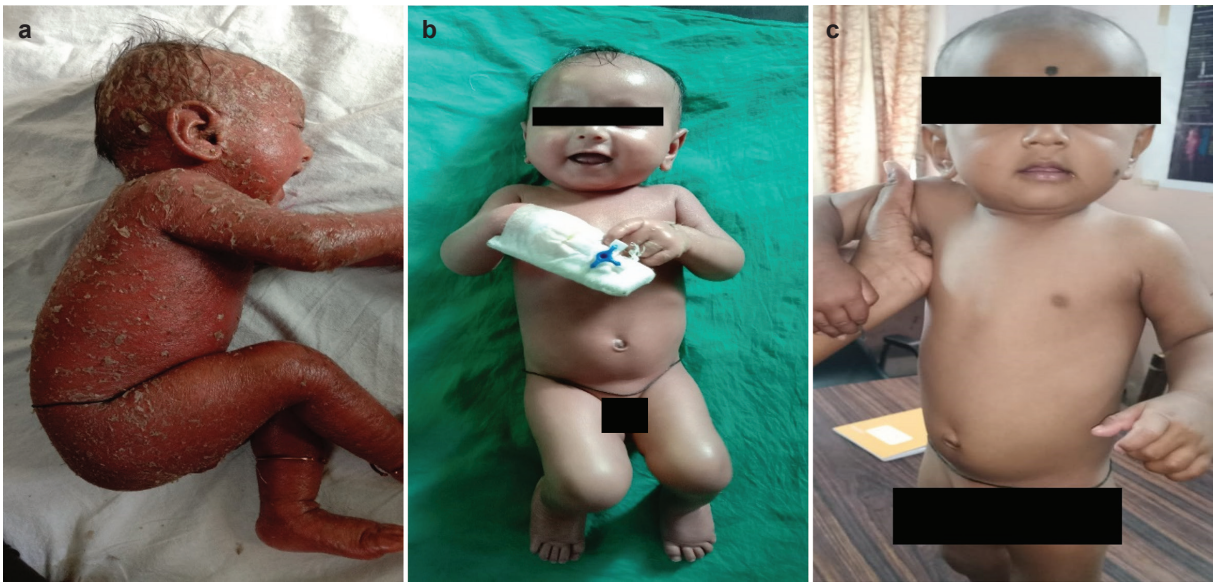

Figure 5. (a) Second case presenting with erythroderma. (b) Clinical improvement after starting prednisolone and vitamin A. (c) Complete resolution of lesions after five months.

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