Neonatal Erythroderma as an Early Sign of Primary Immunodeficiency

A 20-day-old female neonate, born to first-cousin healthy parents from Pakistan, presented to our pediatric emergency department with erythroderma and desquamation (Figure 1). She had proper weight gain and was afebrile. Findings of the physical examination showed erythroderma with scaling skin and patchy alopecia, without lymphadenopathy or hepatosplenomegaly. Blood tests showed lymphopenia (650/mmc), eosinophilia (2800/mmc), high immunoglobulin E (167 UI/mL), and lymphocyte subsets revealed a T–B–NK+ phenotype leading to a diagnosis of severe combined immunodeficiency disease. The skin biopsy highlighted parakeratosis, focal reduction of stratum granulosum, and inflammatory perivascular infiltrate in the dermis. Genetic testing confirmed the diagnosis of Omenn syndrome by showing homozygous mutation in the recombination-activating gene, which was present in heterozygosis in both parents. At the age of 3 months, the patient successfully underwent allogeneic hematopoietic stem cell transplantation from HLA-identical donor (her mother); she is currently 1-year post transplantation and fully engrafted without signs of graft-versus-host disease.

This case illustrates a diagnosis of primary immunodeficiency (PID) in an erythrodermic newborn before the development of failure to thrive, infections, diarrhea, hepatosplenomegaly, or lymphadenopathy. Erythroderma is an inflammatory skin disorder affecting more than 90% of the body surface, with erythema and scaling (Figure 2).[1] Neonatal erythroderma is a rare condition and can be the primary manifestation of a broad spectrum of conditions, from benign skin diseases to potentially fatal systemic diseases.[2] Erythroderma itself, depending on the degree of epidermal disruption, can cause serious complications such as electrolyte imbalance, hypoalbuminemia, dehydration, temperature instability, and infections, even leading to sepsis. These neonates need a warm and humid environment and require an adequate liquid intake and topical therapy with emollients to guarantee proper skin hydration.[3]

As reported,[4-6] differential diagnoses of neonatal erythroderma include monogenic skin diseases (eg, epidermolytic

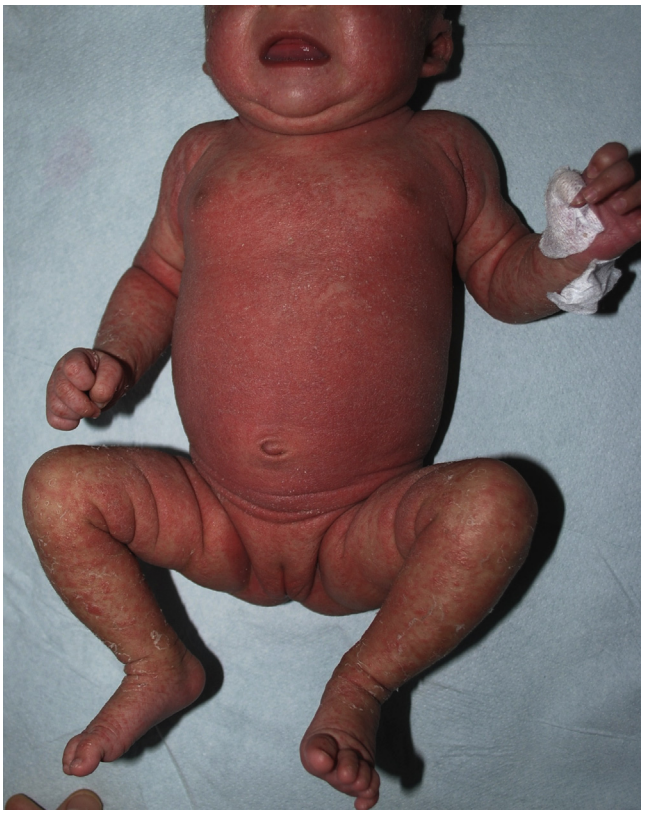

Figure 1. Erythrodermal and desquamative lesions extended to the whole body but more prominent at the limbs in a 20-day-old neonate with no other symptoms.

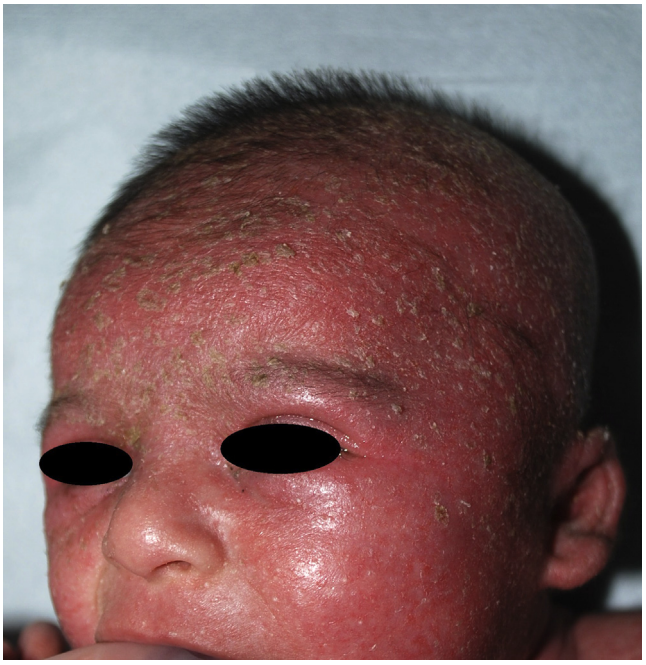

Figure 2. Erythema and scaling affecting more than 90% of the body surface area in a neonate.

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ichthyosis, Netherton syndrome), infections, benign skin diseases (ie, infantile psoriasis, atopic dermatitis, or seborrheic dermatitis), immune disorders, metabolic diseases, and drug-related erythroderma. Failure to thrive, severe infections, neurologic symptoms, or signs of metabolic imbalance are red flags for urgent diagnostic and therapeutic interventions.[4]

Erythroderma with alopecia, failure to thrive, diarrhea, hepatosplenomegaly, and lymphadenopathy are suggestive of PID, a complex of signs and symptoms called Omenn syndrome. In this case, the severe and extensive skin phenotype was due to autoreactive T cells responsible for a graft-versus-host disease-like phenotype.

The correct management of erythrodermic neonates is a multistep procedure and always requires the knowledge of family and medical history and a physical examination; a dermatologic visit is recommended. Because benign erythematous disorders are rare in neonates, further analyses, such as complete blood count, serum electrolytes, blood gas analyses, and serum IgE levels, are often necessary.[4] Immunologic phenotype and genetic tests are crucial in case a PID is suspected. In desquamative erythroderma, a skin biopsy is also important.[7] Performing an early diagnosis is a key element, as its delay, in most cases of neonatal erythroderma, may lead to fatal consequences. ■

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Giant Urticaria and Acral Peeling in a Child with Coronavirus Disease 2019

A healthy 6-year-old girl presented with pruritic skin eruptions. The child was on the sixth day of isolation, with her mother suffering from a mild form of coronavirus disease 2019 (COVID-19) with ageusia and a single febrile episode.

The next day, the child developed fever and pharyngodynia. In the emergency department, a nasal swab for severe acute respiratory syndrome coronavirus 2 (both molecular and antigen tests) was positive, and she was admitted to the COVID-19 unit of our institute.

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Skin examination revealed fleeting urticarial lesions lasting <24 hours and migrant appearance with polycyclic contours consistent with the diagnosis of acute viral giant urticaria (**Figure**, A-C). Two days after the onset of the skin lesions, a desquamation of the distal phalanges of the hands and feet appeared with cyanosis of the apical portion of the nail bed (**Figure**, D).

The remaining physical examination and blood tests were unremarkable. No cardiac or respiratory abnormalities or signs suggestive of Kawasaki disease were evident. An oropharyngeal swab permitted us to rule out a streptococcal infection.

The patient's fever disappeared quickly, lasting only 24 hours. Antihistamine therapy was given for symptomatic