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Severe Combined Immunodeficiency Revealed by Neonatal Erythroderma

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Severe combined immunodeficiency (SCID) is a rare syndrome caused by a variety of genetic abnormalities and characterized by a lack of T-lymphocyte and B-lymphocyte function. It is a frequent cause of erythroderma in the neonatal period and should be considered in cases of severe erythroderma or erythroderma resistant to standard treatments. [1] We report the case of a one-week-old neonate, the youngest of 2 siblings, from a 1st-degree consanguineous marriage and with a history of death in a brother at the age of 1 month. The pregnancy was poorly monitored, carried to term and delivered vaginally. Birth weight was 2900g. The newborn was admitted for erythroderma evolving since D4 of life, initially managed as toxic erythema of the newborn, hospitalized in view of the aggravation of the clinical symptomatology with the appearance of fever and hypotonia. Clinical examination revealed a diffuse erythematosquamous rash, with a diffuse infiltrate and no organomegaly. Blood counts were consistent with hyperleukocytosis and lymphopenia. CRP and procalcitonin were elevated. Renal and hepatic function were normal. Protein levels were low. HIV serology was negative. Chest X-ray was unremarkable. Bacteriological samples were negative. The diagnosis of severe post-natal infection was evoked. The patient was started on broad-spectrum bi-antibiotic therapy. The patient died within 48 hours. Given this clinical picture, an immune deficiency was strongly suspected, given the anamnestic and clinico-biological data.

In a retrospective study by Pruszkowski et al, immune deficiency was diagnosed in 30% of patients with neonatal erythroderma [2]. An immune work-up is then required, based on a blood count, protein electrophoresis, weighted immunoglobulin assay and lymphocyte phenotyping [1]. A skin biopsy will help orient the diagnosis, demonstrating a significant inflammatory infiltrate and keratinocyte necrosis [3]. Erythroderma warrants appropriate in-hospital management by specialized teams, aimed at optimizing the skin barrier and preventing metabolic, hydroelectrolytic and infectious complications (Figure 1).

Keywords: Erythroderma; Immune deficiency; Newborn; Sepsis

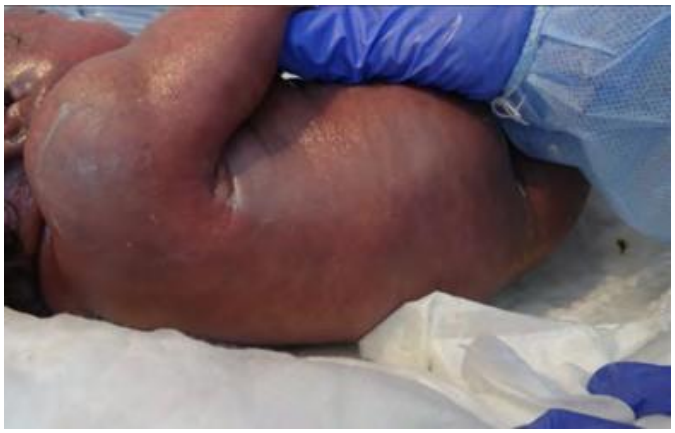

Figure 1: Diffuse erythematous-scaly eruption in a one-week-old newborn.

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