A 24-day-old African female presented with cutaneous targetoid lesions that had begun ten days earlier. The patient had been born at term, after an uncomplicated pregnancy. She had been diagnosed with citrullinemia type 1 in the first week of life (high serum citrulline level of 858 mol/L and hyperammonemia of 55 mol/L), and arginine supplementation was initiated. No other drugs were administered, and maternal medications were excluded. The mother did not have any known systemic diseases. Physical examination revealed multiple annular erythematous plaques of variable size, sharply margined, on the forehead, neck and upper trunk (Figure 1A, 1B). The palms, soles and mucous membranes were spared. The rest of her exam was normal with no evidence of cardiac abnormalities or hepatosplenomegaly.

Initial laboratory evaluation showed only neutropenia (830/mm³). Red blood cell and platelet count, hepatic function, and serum creatinine were normal. Syphilis and viral serologies (cytomegalovirus, Epstein-Barr virus and herpes simplex) were negative, as was mycological examination of skin scrapings. A skin biopsy revealed degeneration of the basal cell layer, necrotic keratinocytes within an atrophic epidermis and dermal perivascular lymphocytic infiltration (Figure 2A, 2B).

Further laboratory tests showed positive anti-SSA/Ro52 (474.6 U/mL), anti-SSa/Ro60 (744 U/mL), anti-SSb/La (1550 U/mL) and high titre antinuclear antibody (1:320) at the patient. Mother’s serological examination revealed an antinuclear antibody titre of 1:1280; anti-SSa/Ro52 (3443.1 U/mL), anti-SSa/Ro60 (1352.2 U/mL) and anti-SSb/La (15500 U/mL) antibodies were positive. The patient was diagnosed with Neonatal lupus erythematosus on the basis of the clinical, histopathological and laboratory findings.

Cardiologic investigation revealed no abnormalities and the patient was treated with a low-potency topical steroid. The neutropenia disappeared after six weeks.

**FIGURE 1.** A) Erythematous, annular plaques located on the forehead, neck and trunk. B) Periorbital erythematous plaques with violaceous center.
and the lesions healed with residual hyperpigmentation within three months. The diagnosis of citrullinemia type 1 was later confirmed by molecular tests (a compound heterozygous mutation in ASS1 gene).

Neonatal lupus is a rare immune-mediated disease that results from transplacental transfer of maternal antibodies against Ro, La and, less commonly, U1-ribonucleoprotein. While some mothers may have an autoimmune disease, half of them are asymptomatic at the time of childbirth. To our knowledge, neonatal lupus had never been reported in association with citrullinemia, but according to some studies, arginine and its precursor citrulline may play an important role in T cell function.

Neonatal lupus can affect different organs, mainly the skin and heart. Cutaneous findings consist of erythematous annular plaques, seen most commonly on scalp and periorbital regions. They are observed in approximately 40% of the cases, and generally develop during the first two months of life. Although the skin lesions are usually characteristic, they pose a diagnostic challenge in the absence of maternal history, and they are sometimes misdiagnosed as seborrheic dermatitis, tinea faciae, or erythema multiforme. Cardiac disease may present with total atrioventricular block, which is the most severe complication for the baby, with significant morbidity and mortality. Other findings may include hepatobiliary dysfunction, cytopenias and neurologic involvement.

Although cutaneous manifestations are benign and self-limited, early recognition may help identify life-threatening complications of neonatal lupus.

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