Neonatal lupus erythematosus (NLE) is an uncommon transplacentally acquired autoimmune disorder. The most common clinical manifestations are skin rash, congenital atrioventricular block, thrombocytopenia, leukopenia, anemia, and hepatosplenomegaly. Usually, the skin rash resembles subacute cutaneous lupus, but different forms of rash have been reported in neonatal lupus erythematosus and some are rare forms. NLE should be suspected in babies with atypical skin lesions, even if present at birth.

NLE = neonatal lupus erythematosus

Abstract

Neonatal lupus erythematosus is an uncommon transplacentally acquired autoimmune disorder. The most common clinical manifestations are skin rash, congenital atrioventricular block, thrombocytopenia, leukopenia, anemia, and hepatosplenomegaly. Usually, the skin rash resembles subacute cutaneous lupus, but different forms of rash have been reported in neonatal lupus erythematosus and some are rare forms. NLE should be suspected in babies with atypical skin lesions, even if present at birth.

NLE should be suspected in babies with atypical skin lesions

Díaz Jouanen [11] reported a boy infant, born to a mother with systemic lupus erythematosus, who showed some annular erythemas on his face, back, chest and extremities in the first month of life. Most of these lesions faded but some evolved to large atrophic lesions that gave rise to areas of sclerosis by the age of 2 months. Skin biopsies and clinical features were compatible with concurrent NLE and multiple morphea. The lesions did not resolve during a 3 year follow-up [11].

Nitta [5] described a Japanese female baby with NLE who had a concurrent lupus profundus on the face [5]. The girl developed scaly discoid lesions within 2 days of birth that evolved to concurrent lupus erythematosus profundus on the face in the fifth month. Depression of lupus profundus was still evident at 4 years of age. Lupus erythematosus profundus is rare in children. Its rate in adults with cutaneous lupus erythematosus ranges from 1% to 3% [11,12].

A third case with atypical cutaneous manifestation was a 3 week old female newborn. Immunostaining (biopsy) of the
papulo-erythematous eruptions on her trunk and limbs showed a dermal infiltrate of cells of myelomonocytic origin, as seen in hematodermic neoplasm or macrophage activation syndrome. The child had no other signs to support these diagnoses. The lesions eventually evolved to an erythematous rash on the face and eyelids, compatible with NLE. She and her mother were positive for anti-SSA and anti-SSB antibodies. Her clinical examination was normal by age 10 months [6].

**Neonatal lupus erythematosus, an uncommon syndrome considered a model of passively acquired autoimmune disease, is characterized mainly by cutaneous rash and congenital atrioventricular block in the presence of maternal-derived autoantibodies**

In the few reports of congenital lupus erythematosus with atrophy, the babies’ face and scalp had cutaneous atrophic/scarring lesions at birth. Diagnosis of the congenital presentation of NLE was made by skin biopsy [7,8] and antinuclear antibody test [8]. It is a very rare occurrence and suggests that sun exposure is not necessary for NLE skin lesions to develop.

Another case of NLE with uncommon skin manifestations was described by Adrian See et al. [10]. It involved a 4 week old male baby presenting with nodules/papules on the plantar surface of both feet. Biopsy was consistent with lupus erythematosus. Annular lesions appeared 2 weeks later on the face.

It is important to consider NLE as a possible diagnosis in babies with atypical skin lesions, even if they are born with them, since NLE may be associated with a congenital presentation or a rare cutaneous manifestation.

**References**


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**Capsule**

**What’s in a face?**

The brain processes objects through a series of regions along the ventral visual pathway, but the circuitry subserving the analysis of specific complex forms remains unknown. Most macaque monkeys have six patches of face-selective cortex distributed throughout the brain’s temporal lobe. These six face patches offer an ideal framework for dissecting the functional architecture of object recognition. What are the inputs and outputs of each of the six face patches? Do they form a functionally interconnected network? Or are they independent nodes, each processing a different aspect of faces? Moeller et al. directly imaged connections of the macaque face patches by combining in vivo microstimulation and brain imaging. Stimulation of the lateral middle face patch resulted in strong, specific activation of all five other face patches. Stimulation in the other face patches also resulted in specific activation of a subset of face patches. The face patches are thus part of an interconnected network, which may generate a hierarchy of face processing stages.

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