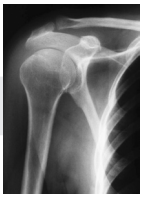

Neonatal Lupus Erythematosus: An Acquired Autoimmune Disorder and its Cutaneous Manifestations

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Key words: neonatal lupus, acquired autoimmune disease, cutaneous manifestations

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 diferent 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.

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Neonatal lupus erythematosus is an uncommon syndrome that was first described in 1954 by McCuistion and Schoch [1]. It is a model of passively acquired autoimmune disease that affects the fetus, caused by the passage of specifically maternal derived antinuclear autoantibodies and extractable nuclear antigen (anti-Ro/SSA, anti-La/SSB and anti-RNP) that result from an autoimmune process in the mother, through the placenta to the fetal circulation [2]. The mother may have an autoimmune disease or may be asymptomatic.

NLE is clinically characterized mainly by cutaneous rash, congenital atrioventricular block, thrombocytopenia, leukopenia, anemia and hepatosplenomegaly in the presence of maternal derived autoantibodies. NLE resolves in the first year of life and the permanent damage is due to cardiac involvement. The literature reveals few children with NLE who developed autoantibodies or another connective tissue disease after many years of follow-up.

Roughly 50% of infants with NLE have skin involvement. Typical skin rashes of NLE resemble subacute cutaneous lupus; they can be present at birth or appear in the first months of life. The most common types of skin rashes consist of annular, erythematous and scaly lesions. The face, scalp and periocular distributions (eye mask/raccoon-like appearance) are characteristic of NLE. It usually resolves within the first 6–8 months of life, as clearance of maternal autoantibodies from the infant's circulation occurs. Sometimes it can result in a mild hypo or hyperpigmentation on the malar area or in telangiectatic maculae, but it usually does not result in scarring or in an atrophic lesion. Histologically, epidermal basal cell damage, hyperkeratosis, atrophy of epidermis, epidermal colloid bodies, thickened basement membrane, vacuolation of the basal layer, and dermal mononuclear cell infiltrate are observed [3].

The literature reports some uncommon skin lesions in children with NLE: multiple morphea [4], discoid lesions associated with lupus erythematosus profundus [5], papulo-erythematous rash with dermal infiltrate of cells of myelomonocytic origin [6], congenital cutaneous lupus with atrophic lesions [7-9], and NLE presenting with acral papules before facial annular lesions develop [10].

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 scanty 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

NLE = neonatal lupus erythematosus

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.

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