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Neonatal Lupus Erythematosus with Atypical Hypopigmented Atrophic Facial Lesions and a Concomitant Nuchal Capillary Malformation in an Infant with Skin of Color: A Case Report

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Abstract | **Case Report**

Neonatal lupus erythematosus (NLE) is a rare acquired autoimmune condition resulting from transplacental transfer of maternal autoantibodies, most commonly anti-Ro/SSA and anti-La/SSB. Cutaneous disease is its most frequent manifestation and classically presents as annular or polycyclic erythematous plaques in a periorbital "raccoon-eye" distribution. We describe a 5-week-old male infant with skin of color who presented with progressive hypopigmented, atrophic facial macules that had been preceded by crusted, erosive lesions present since birth. An incidental non-inflammatory erythematous patch over the occipital and nuchal region, consistent with a nevus simplex, was also noted and had been stable since birth. Laboratory work-up confirmed positive anti-Ro/SSA antibodies with negative anti-La/SSB antibodies, mild normochromic normocytic anemia and a markedly elevated erythrocyte sedimentation rate; cardiac and hepatic evaluations were normal. The infant improved substantially with strict photoprotection and a low-potency topical corticosteroid. This case illustrates an atypical, predominantly atrophic-hypopigmented cutaneous phenotype of NLE that may be under-recognized in infants with darker skin, and an exceptionally rare co-occurrence with a capillary vascular malformation, the pathogenic significance of which coincidental versus a shared underlying mechanism remains to be clarified.

Keywords: Lupus erythematosus, Cutaneous – Nevus – Child – Case report - Sub-Saharan Africa.

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INTRODUCTION

Neonatal lupus erythematosus is a rare, passively acquired autoimmune syndrome mediated by transplacental transfer of maternal anti-Ro/SSA and anti-La/SSB antibodies. [1] The condition affects an estimated 1 in 12,500–20,000 live births in the United States. [2,3] Cases have also been reported across sub-Saharan Africa, although the population prevalence in this region remains undefined. [3,4] Cutaneous involvement is the most common presenting feature, typically evolving over the first weeks of life and often triggered by ultraviolet light exposure; the classic phenotype is an annular or polycyclic erythematous eruption with a centrofacial and periorbital "raccoon-eye" distribution. [1,5,6] Atypical morphologies including purpuric, telangiectatic, and hypopigmented variants are increasingly reported and may be more likely to be missed or misdiagnosed, particularly in infants with darker skin phototypes, in whom erythema is less conspicuous. [2,3] Congenital vascular malformations such as nevus simplex are common, sporadic, and not otherwise linked to autoimmune disease. [1,2] Their co-occurrence with neonatal lupus erythematosus is, to our knowledge, exceptionally rarely documented. We report this association together with an atypical hypopigmented, atrophic cutaneous phenotype of NLE, and discuss the diagnostic and pathogenic implications.

CASE REPORT

A 5-week-old male infant was brought for dermatological consultation for progressive hypopigmented facial macules. He was born at term by uncomplicated vaginal delivery to a 27-year-old primigravida mother with no known autoimmune or

other chronic disease and no history of miscarriage. The father, aged 50 years, was in good general health with no relevant medical history.

Crusted, erosive facial lesions had been present since birth (day 0) and healed spontaneously over two weeks, leaving progressively enlarging hypopigmented macules. The mother had applied topical trolamine (Biafine®) for three weeks without improvement, prompting referral to dermatology.

General examination showed a well-appearing infant with normal growth parameters and mildly pale conjunctivae. Dermatological examination revealed atrophic, hypopigmented macules with fine overlying scale, disseminated across the face and coalescing into a continuous band across the periorbital and upper mid-facial region (Figure 1). A separate, non-inflammatory erythematous patch with an irregular, geographic border and a roughly ovoid shape (long axis 8 cm) was noted over the occipital and nuchal region, consistent with a nevus simplex; this lesion had been stable since birth (Figure 2). Cardiovascular examination and the remainder of the systemic examination were unremarkable.

Post-inflammatory hypomelanosis, neonatal vitiligo, and seborrheic dermatitis were initially considered. The atrophic quality of the lesions argued against these diagnoses and raised suspicion for neonatal lupus erythematosus, prompting serological investigation.

Infant serology was positive for anti-Ro/SSA antibodies (213.2 U/mL) and negative for anti-La/SSB antibodies. Full blood count showed normochromic, normocytic anemia (hemoglobin 8.4 g/dL) with normal white cell count (8,050/µL) and platelet count (205,000/mm³). Erythrocyte sedimentation rate was markedly elevated (50 mm at 1 hour; 103 mm at 2 hours), while C-reactive protein was negative. Liver enzymes were normal (AST 14 IU/L, ALT 7 IU/L), and cardiac evaluation (ECG and echocardiography) was normal.

The mother was clinically asymptomatic for connective tissue disease. Maternal serology showed a positive speckled antinuclear antibody pattern (indirect immunofluorescence, titer 1:1280) with negative anti–double-stranded DNA (FEIA). Maternal anti-Ro/SSA antibodies were positive; anti-La/SSB antibodies were negative.

The infant was managed with strict photoprotection, a low-potency topical corticosteroid applied to the facial lesions, and a bland emollient for the remainder of the body.

Outcome and follow-up

At four-week follow-up, marked clinical improvement was observed, with regression of the hypopigmented facial macules and residual post-inflammatory hyperpigmentation (Figure 3). The nuchal capillary malformation persisted unchanged. No systemic involvement was identified during follow-up. The mother was referred to internal medicine for further autoimmune evaluation and longitudinal follow-up.



Figure 1: Hypopigmented, atrophic facial macules at 5 weeks of age



Figure 2: Capillary malformation of the nuchal and occipital region



Figure 3: Evolution of the lesions after four weeks of treatment

DISCUSSION

Neonatal lupus erythematosus is caused by passive transplacental transfer of maternal anti-Ro/SSA and, less consistently, anti-La/SSB antibodies. [1] Its incidence is approximately 2% among infants born to anti-Ro/anti-La positive mothers, rising to 18–20% in subsequent pregnancies. [6] Roughly 25% of mothers of affected infants are asymptomatic for autoimmune disease at delivery, and about 50% develop symptoms within three years. [1,7,8] NLE therefore frequently serves as a sentinel diagnosis for latent maternal connective tissue disease, as illustrated in the present case.

Cutaneous disease is the most common manifestation of NLE, followed by hematological, hepatobiliary and cardiac involvement. [10,11] Skin lesions typically appear within the first weeks of life, often after ultraviolet exposure, and classically present as annular or polycyclic erythematous plaques with a centrofacial, periorbital "raccoon-eye" distribution. [1,5,6] Atypical variants purpuric, telangiectatic or hypopigmented are increasingly recognized. [2,3]

In our patient, the lesions were initially crusted and erosive, evolving into atrophic, hypochromic macules an uncommon phenotype that may correspond to an intense initial inflammatory phase progressing to post-inflammatory atrophy. This phenotypic variability underscores the need for a high index of suspicion for any atypical neonatal facial dermatosis. This point is clinically important in infants with darker skin phototypes: the erythema that defines the classic "raccoon-eye" pattern is far less conspicuous on pigmented skin, and hypopigmented, atrophic lesions may instead be mistaken for vitiligo, post-inflammatory hypomelanosis or tinea versicolor, potentially delaying diagnosis and systemic screening.

Positive anti-Ro/SSA antibodies in the infant confirmed the diagnosis, even in the absence of anti-La/SSB antibodies. [2,3,4] The mild anemia observed is consistent with the generally transient, favorable-prognosis hematological involvement described in NLE. The markedly elevated erythrocyte sedimentation rate, although non-specific, is unusual in purely cutaneous forms and may reflect a modest degree of systemic inflammatory activity. The principal clinical concern in NLE remains cardiac involvement, particularly congenital complete heart block, which is the main determinant of mortality. Although absent in our patient, systematic cardiological screening is mandatory regardless of the severity or extent of cutaneous involvement.

A distinctive feature of this observation is the co-occurrence with a nevus simplex. Congenital capillary malformations arise from sporadic anomalies of cutaneous vascular development and are not otherwise linked to autoimmune disease. [12] To our knowledge, this association is exceptional and has been very sparsely documented in the literature. Several explanations can be considered. A chance co-occurrence cannot be excluded, given the relatively high background prevalence of nevus simplex in the general population. [12] Alternatively, a shared underlying immunological or vascular developmental susceptibility could theoretically predispose to both findings; this hypothesis remains speculative and unproven. On balance, the first explanation coincidence appears more plausible, and larger case series would be required to establish any genuine pathogenic link.

This case has several limitations. The absence of long-term follow-up precludes assessment of the eventual course of the cutaneous lesions and of secondary autoimmune disease in the mother. In addition, the absence of skin histology limits fine pathophysiological characterization of the lesions.

Management of cutaneous NLE relies principally on strict photoprotection, which is essential to prevent worsening of lesions. Topical corticosteroids or calcineurin inhibitors may be used for inflammatory forms. [3,13,14] The disease typically resolves over several months in parallel with clearance of circulating maternal antibodies, although residual pigmentary or atrophic changes may persist, as observed in our patient.

Learning points

Neonatal lupus erythematosus can present with atypical, purely hypopigmented and atrophic facial lesions rather than the classic annular "raccoon-eye" erythema and should remain a differential diagnosis for any atypical neonatal facial dermatosis.

Diagnosis may be more challenging and delayed in infants with darker skin phototypes, in whom erythema is less visible and hypopigmented lesions can mimic vitiligo or post-inflammatory hypomelanosis.

Diagnosis is confirmed by maternal and infant anti-Ro/SSA and anti-La/SSB serology; a single positive antibody (typically anti-Ro/SSA) is sufficient.

Systematic cardiac (ECG and echocardiography) and hepatic evaluation is mandatory in all infants with NLE, irrespective of the severity of cutaneous involvement.

A diagnosis of NLE in an infant should prompt referral of the mother for autoimmune work-up, as up to half of initially asymptomatic mothers develop connective tissue disease within a few years.

Patient's perspective

The infant's mother reported that she had first applied a topical trolamine emollient for three weeks without improvement before seeking dermatological review, reflecting the initial impression that the facial

lesions represented a minor, self-limiting skin irritation rather than a systemic disease requiring investigation.

Patient consent for publication

Written informed consent was obtained from the infant's parents for publication of this case report and the accompanying clinical images.

Contributors

All listed authors contributed to the clinical care of the patient, acquisition and interpretation of data, and drafting or critical revision of the manuscript, and approved the final version submitted.

Competing interests

None declared.

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