Clinical Image

A Distinct Annular Eruption: Neonatal Lupus Erythematous

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ABBREVIATIONS

ANC: Absolute Neutrophil Count; ANA: Antinuclear Antibody; dsDNA: Double-stranded Deoxyribonucleic acid; SS-A: Sjögren’s-syndrome-related antigen A; SS-B: Sjögren’s-syndrome-related antigen B; U1RNP: U1-ribonucleoprotein; NLE: Neonatal Lupus Erythematous

CLINICAL IMAGE

An 8-week old male born at 36 weeks gestation presented to clinic with a progressive rash. At birth, he was noted to have an erythematous atrophic macule on the upper lip. Over the next two months, he developed pink-orange, annular, scaly macules that slowly spread to involve his cheeks, forehead, and frontotemporal scalp (Figures 1,2). He was otherwise growing and developing well. Laboratory evaluation was remarkable for an elevated alkaline phosphatase of 439 (normal 150-420), hemoglobin of 8.5 (normal 9.5-14.1), platelets of 167 (normal 150-400), and an ANC of 788 (normal 1,000-8,500). Hepatic panel and head circumference were within normal limits. Autoimmune serologies were notable for positive ANA (1:160, speckled pattern), negative dsDNA, positive SS-A, negative SS-B, and negative Smith antibodies. An EKG suggested biventricular hypertrophy; however follow-up echocardiogram was within normal limits. Patient experienced fading of the lesions after treatment with topical tacrolimus at 4-month follow-up with resolution of the rash by 6 months of age. No further testing of the infant was undertaken, and he is clinically doing well. The infant’s mother had a history of Hashimoto’s thyroiditis and vitiligo; her laboratory evaluation was remarkable for a positive ANA (1:320, speckled pattern), positive SS-A, and negative anti-U1RNP antibodies. She does not currently meet criteria for a specific connective tissue disease but is being followed by Rheumatology.

Neonatal lupus erythematous (NLE) is acquired by transplacental transmission of maternal anti-SSA/Ro, anti-SSB/La,
or anti-U1RNP antibodies with resultant fetal tissue damage [1,2]. Mothers are usually asymptomatic and unaware of their autoantibody status [2]. NLE may manifest rash, cytopenias, hepatobiliary disease, congenital heart block, and cardiomyopathy [1]. The rash of NLE is comprised of erythematous annular or arcuate macules with central clearing and slightly raised margins, primarily located on the scalp and peri-orbital regions [3,4]. Less commonly, bullous lesions may present with predilection for the soles of the feet [3]. The cutaneous eruption has a tendency to be photosensitive, but may present immediately at birth or in sun-protected areas [3]. It may be confused with seborrheic dermatitis, nevus simplex, nevus flammeus, or fungal infection. Cutaneous lesions typically resolve with dearance of maternal antibodies around 6 months of age, whereas cardiac damage may be irreversible [1,4]. Congenital heart block is the most serious potential complication of neonatal lupus, with significant risk of morbidity and mortality (15-30%) often occurring within the first few months of life [1]. Hydrocephalus may also be a complication; infants with macrocephaly should therefore undergo further imaging [3].

Infants with a facial annular or polycyclic rash and/or any degree of heart block should undergo serum testing for NLE antibodies, electrocardiogram, and cardiology evaluation as warranted. Work-up of the mother should be undertaken, as even asymptomatic mothers have a 19% risk of developing systemic lupus erythematosus and a 28% chance of developing Sjögren syndrome within 10 years [5]. Lastly, parents should be counseled of the increased risk of NLE in future pregnancies, which approaches 16-24% [5].

REFERENCES