Diffuse Annular Plaques in an Infant

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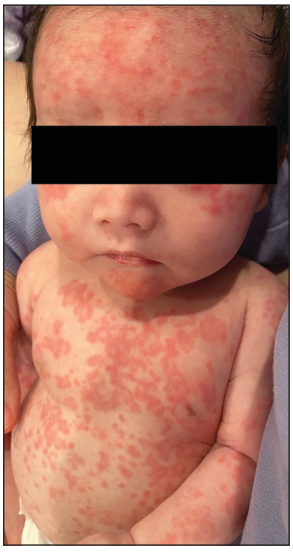

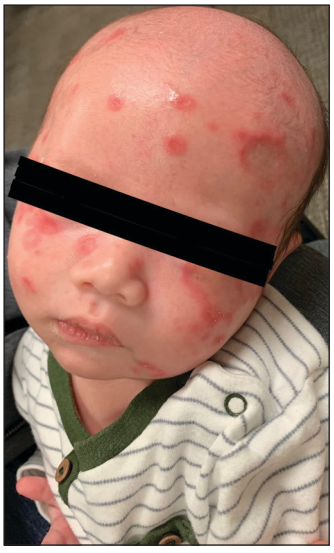

A 5-week-old infant boy presented with a rash at birth (left). The pregnancy was full term without complications, and he was otherwise healthy. A family history revealed that his older brother developed a similar rash 2 weeks after birth (right). Physical examination revealed polycyclic annular patches with an erythematous border and central clearing diffusely located on the trunk, extremities, scalp, and face with periorbital edema.

WHAT'S YOUR **DIAGNOSIS?**

a. annular erythema of infancy
b. erythema multiforme
c. neonatal lupus erythematosus
d. tinea corporis
e. urticaria multiforme

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THE **DIAGNOSIS:** Neonatal Lupus Erythematosus

A review of the medical records of the patient's mother from her first pregnancy revealed positive anti-Ro/SSA (Sjögren syndrome A) (>8.0 U [reference range <1.0 U]) and anti-La/SSB (Sjögren syndrome B) antibodies (>8.0 U [reference range <1.0 U]), which were reconfirmed during her pregnancy with our patient (the second child). The patient's older brother was diagnosed with neonatal lupus erythematosus (NLE) 2 years prior at 1 month of age; therefore, the mother took hydroxychloroquine during the pregnancy with the second child to help prevent heart block if the child was diagnosed with NLE. Given the family history, positive antibodies in the mother, and clinical presentation, our patient was diagnosed with NLE. He was referred to a pediatric cardiologist and pediatrician to continue the workup of systemic manifestations of NLE and to rule out the presence of congenital heart block. The rash resolved 6 months after the initial presentation, and he did not develop any systemic manifestations of NLE.

Neonatal lupus erythematosus is a rare acquired autoimmune disorder caused by the placental transfer of anti-Ro/SSA and anti-La/SSB antibodies and less commonly anti-U1 ribonucleoprotein antinuclear autoantibodies.[1,2] Approximately 1% to 2% of mothers with these positive antibodies will have infants affected with NLE.[2] The annual prevalence of NLE in the United States is approximately 1 in 20,000 live births. Mothers of children with NLE most commonly have clinical Sjögren syndrome; however, anti-Ro/SSA and anti-LA/SSB antibodies may be present in 0.1% to 1.5% of healthy women, and 25% to 60% of women with autoimmune disease may be asymptomatic.[1] As demonstrated in our case, when there is a family history of NLE in an infant from an earlier pregnancy, the risk for NLE increases to 17% to 20% in subsequent pregnancies[1,3] and up to 25% in subsequent pregnancies if the initial child was diagnosed with a congenital heart block in the setting of NLE.[1]

Neonatal lupus erythematosus classically presents as annular erythematous macules and plaques with central scaling, telangictasia, atrophy, and pigmentary changes. It may start on the scalp and face and spread caudally.[1,2] Patients may develop these lesions after UV exposure, and 80% of infants may not have dermatologic findings at birth. Importantly, 40% to 60% of mothers may be asymptomatic at the time of presentation of their child's NLE.[1] The diagnosis can be confirmed via antibody testing in the mother and/or infant. If performed, a punch biopsy shows interface dermatitis, vacuolar degeneration, and possible periadnexal lymphocytic infiltrates on histopathology.[1,2]

Management of cutaneous NLE includes sun protection (eg, application of sunscreen) and topical corticosteroids. Most dermatologic manifestations of NLE are transient, resolving after clearance of maternal IgG antibodies in 6 to 9 months; however, some telangiectasia, dyspigmentation, and atrophic scarring may persist.[1-3]

Neonatal lupus erythematosus also may have hepatobiliary, cardiac, hematologic, and less commonly neurologic manifestations. Hepatobiliary manifestations usually present as hepatomegaly or asymptomatic elevated transaminases or γ-glutamyl transferase.[1,3] Approximately 10% to 20% of infants with NLE may present with transient anemia and thrombocytopenia.[1] Cardiac manifestations are permanent and may require pacemaker implantation.[1,3] The incidence of a congenital heart block in infants with NLE is 15% to 30%.[3] Cardiac NLE most commonly injures the conductive tissue, leading to a congenital atrioventricular block. The development of a congenital heart block develops in the 18th to 24th week of gestation. Manifestations of a more advanced condition can include dilation of the ascending aorta and dilated cardiomyopathy.[1] As such, patients need to be followed by a pediatric cardiologist for monitoring and treatment of any cardiac manifestations.

The overall prognosis of infants affected with NLE varies. Cardiac involvement is associated with a poor prognosis, while isolated cutaneous involvement requires little treatment and portends a favorable prognosis. It is critical for dermatologists to recognize NLE to refer patients to appropriate specialists to investigate and further monitor possible extracutaneous manifestations. With an understanding of the increased risk for a congenital heart block and NLE in subsequent pregnancies, mothers with positive anti-Ro/La antibodies should receive timely counseling and screening. In expectant mothers with suspected autoimmune disease, testing for antinuclear antibodies and SSA and SSB antibodies can be considered, as administration of hydroxychloroquine or prenatal systemic corticosteroids has proven to be effective in preventing a congenital heart block.[1] Our patient was followed by pediatric cardiology and was not found to have a congenital heart block.

The differential diagnosis includes other causes of annular erythema in infants, as NLE can mimic several conditions. Tinea corporis may present as scaly annular plaques with central clearing; however, it rarely is encountered fulminantly in neonates.[4] Erythema multiforme is a mucocutaneous hypersensitivy reaction distinguished by targetoid morphology.[5] It is an exceedingly rare diagnosis in neonates; the average pediatric age of onset is 5.6 years.[6] Erythema multiforme often is associated with an infection, most commonly herpes simplex

virus,[5] and mucosal involvement is common.[6] Urticaria multiforme (also known as acute annular urticaria) is a benign disease that appears between 2 months to 3 years of age with blanchable urticarial plaques that likely are triggered by viral or bacterial infections, antibiotics, or vaccines.[6] Specific lesions usually will resolve within 24 hours. Annular erythema of infancy is a benign and asymptomatic gyrate erythema that presents as annular plaques with palpable borders that spread centrifugally in patients younger than 1 year. Notably, lesions should periodically fade and may reappear cyclically for months to years. Evaluation for underlying disease usually is negative.[6]

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