Neonatal lupus erythematosus or Sweet syndrome?

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A 2-month-old baby boy was admitted to our hospital with a diffuse rash. Birth history was uneventful. Prenatal screening serology for HIV and syphilis was negative. The mother was known to have lupus nephritis that had been in remission for 5 years with regular monitoring.

On examination, the baby had indurated, erythematous plaques around both eyes, and the conjunctiva and sclera were normal. He had erosions on the hard palate. On the trunk, back, arms, legs, palms, and soles he had striking annular plaques with a raised erythematous border (Figs 1 and 2). He was afebrile, with a normal blood pressure and pulse rate. Respiratory, cerebrovascular, and central nervous system findings were all normal. His developmental milestones were appropriate for age.

His blood profile revealed a thrombocytopenia of 37×10^9L (normal range, 140-350 $\times$ 10^9/L), white cell count of 6.12 $\times$ 10^9/L (normal range, 5.50-18.00 $\times$ 10^9/L), hemoglobin level of 9.6 g/dL (normal range, 9.1-13.1 g/dL), and mean corpuscular volume of 79.3 fL (normal range, 77.0-105.0 fL). Our laboratory measures antinuclear antibodies using the EliA connective tissue diseases (EliA CTD Screen) fluoro enzyme immunoassay, and results are reported in a ratio as either negative (<0.7), equivocal (0.7-1.0), or positive (>1.0). Our patient had positive antinuclear antibodies of 24.0, a positive anti-SS-A (Ro) greater than 240 U/mL, and a positive anti-SS-B (La) greater than 320 U/mL. Anti–double-stranded DNA antibody was not detected, and complement C3 and C4 were normal. Liver function values were elevated, with an increased alanine transaminase of 210 U/L (normal range, 4-35 U/L), aspartate transaminase of 737 U/L (normal range, 0-65 U/L), alkaline phosphatase of 977 U/L (normal range, 82-383 U/L), and gamma-glutamyl transferase of 224 U/L (normal range, 12-122 U/L). The mother's blood profile also revealed a positive anti-(Ro) greater than 240 U/mL and a positive anti-(La) greater than 320 U/mL.

Abbreviations used:

SLE: systemic lupus erythematosus
SLND: Sweet-like neutrophilic dermatosis

Skin punch biopsy of an annular plaque on the trunk found a spongiotic neutrophilic dermatitis, with a focus of interface inflammation. The dermis showed subepidermal edema with diffuse perivascular and periadnexal dense neutrophilic inflammation (Figs 3 and 4). There was abundant karyorrhectic debris present and scattered eosinophils. No lymphocytes were noted. There was no evidence of vasculitis or blistering. No mucin stains were done. The report concluded that this was a neutrophilic dermatosis, consistent with Sweet syndrome.

The patient's clinical presentation and serology were in keeping with neonatal lupus. An electrocardiogram and cardiac echocardiogram were done, both of which showed normal heart function. On discharge, after 4 days of monitoring in the hospital, his thrombocytopenia had self-corrected, and the liver enzymes showed a decreasing trend without any intervention. The skin lesions were treated with 0.025% fluocinolone acetonide ointment. Over the following 3 months, he attended 2 dermatology and

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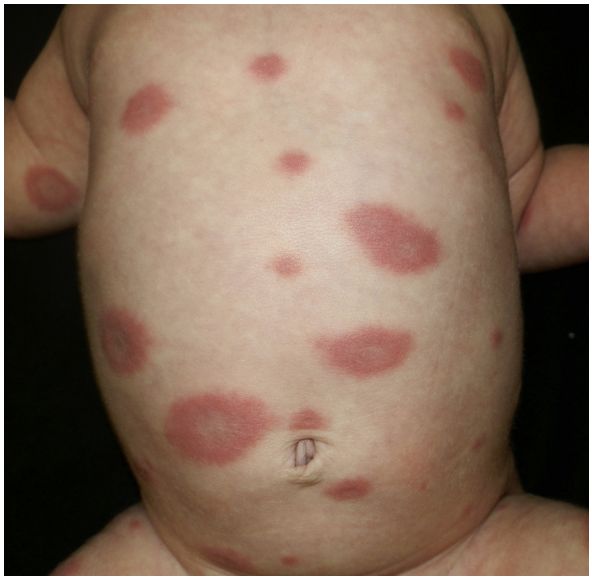

Fig 1. Diffuse erythematous annular and discoid plaques on trunk and arms. A high-resolution version of this slide for use with the Virtual Microscope is available as eSlide: VM04872.

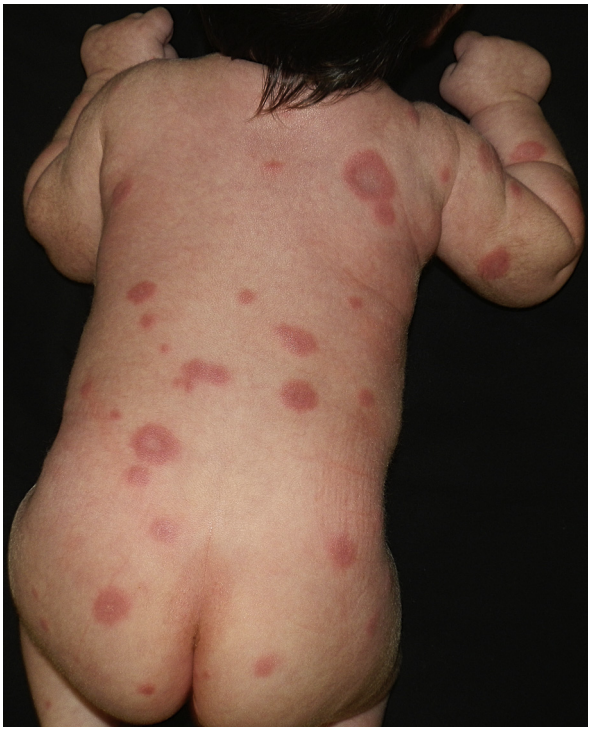

Fig 2. Erythematous annular plaques on back. A high-resolution version of this slide for use with the Virtual Microscope is available as eSlide: VM05092.

3 rheumatology outpatient clinics, and the lesions resolved with postinflammatory hyperpigmentation (Fig 5). Three months after admission, his liver enzymes had normalized. Heart function remained normal at his 6-month repeat electrocardiogram and cardiac echocardiogram investigations.

DISCUSSION

Neonatal systemic lupus erythematosus (SLE) is a rare acquired autoimmune disease. It affects approximately 10% to 20% of infants born to mothers with anti-Ro and anti-La autoantibodies who either have known or undiagnosed SLE, Sjögren syndrome, or undifferentiated autoimmune syndrome with circulating autoantibodies.[1] Maternal autoantibodies anti-Ro and anti-La are transferred from the maternal circulation across the placenta into the fetal circulation.[2] Anti-Ro autoantibodies are usually found in Sjögren syndrome, but they can also be found in 30% of patients with SLE with cutaneous involvement.[3] Cutaneous clinical presentation of neonatal lupus typically affects sun-exposed areas but may occur on the trunk, palms, and soles.[1] The lesions are erythematous patches or plaques, annular or discoid in shape. There may be atrophic macules or patches with or without telangiectasia.[1]

Extracutaneous manifestations of neonatal lupus include heartblock, cardiomyopathy, abnormal liver function tests with or without cholestatic features, hematologic abnormalities including anemia, thrombocytopenia, leukopenia, and central nervous system findings with hydrocephalus.[1] Congenital heart block develops in 15% to 30% of babies with neonatal lupus with 10% going on to develop cardiomyopathy.[1] An electrocardiogram is recommended at the time of diagnosis of neonatal lupus.[4] Any abnormality detected warrants referral to a cardiologist for further management. Our patient had cardiac investigations done at diagnosis of neonatal lupus at 7 weeks and at 6 months, which showed normal functioning of his heart. The Research Registry for Neonatal Lupus (United States) reports the mortality rate of cardiac neonatal lupus at approximately 20%.[5] Treatment of neonatal lupus is organ specific and depends on the severity of the presentation. Infants with neonatal lupus who present with cutaneous, abnormal liver function tests and hematologic abnormalities usually have a resolution of signs and symptoms within 4 to 6 months, with the clearing of maternal autoantibodies.[1,6] This was true for our patient. Supportive measures such as avoidance of sun exposure and sunscreen application are encouraged. Low- to mid-potency topical steroids and topical calcineurin inhibitors can be prescribed in some patients with cutaneous lesions, although they usually heal with no sequelae.[1] However, there have been reports of atrophy and hyperpigmentation.[6]