

CASES FOR DIAGNOSIS

Annular Lesions Following Phototherapy in a Newborn Infant

J. del Boz,^a M. M. Serrano,^b and T. Martín^a

^aServicio de Dermatología and ^bServicio de Pediatría, Complejo Hospitalario Carlos Haya, Málaga, Spain

Patient History

A 20-day-old girl was referred to our department for cutaneous lesions that appeared 1 week earlier and were not associated with any other symptoms (Figures 1 and 2). On the third day of life, the infant had presented jaundice and had been treated by phototherapy, which led to resolution of the symptoms within 48 hours. As relevant family history, the mother was currently being assessed for joint pain and leukopenia that had appeared during the pregnancy.

Physical Examination

Physical examination revealed erythematous papules and annular plaques with a desquamative border and whorl-like growth, located on the cheeks, scalp, retroauricular areas, chest, abdomen, and lower back, with no other skin abnormalities. A complete physical examination revealed no additional abnormalities.

Additional Tests

A complete laboratory workup showed normal blood counts, elevated liver enzymes (glutamic-oxaloacetic transaminase, 197 U/L; glutamic-pyruvic transaminase, 122 U/L; alkaline phosphatase, 231 U/L), and normal bilirubin. Blood tests were positive for antinuclear antibodies (ANA and anti-Ro/SS-A), but normal for complement.

Histopathology

A biopsy showed only inconclusive interstitial and perivascular lymphohistiocytic infiltrate; a second biopsy (Figure 3) revealed minimal vacuolar damage of the basement membrane, isolated necrotic keratinocytes, and interstitial mucin deposition. Direct immunofluorescence was negative.

What is your diagnosis?

Correspondence:
Javier del Boz González
Servicio de Dermatología
Hospital Civil
Plaza del Hospital Civil, s/n
29010 Málaga, Spain
javierdelboz@yahoo.es

Manuscript accepted for publication January 25, 2008



Figure 1.



Figure 2.

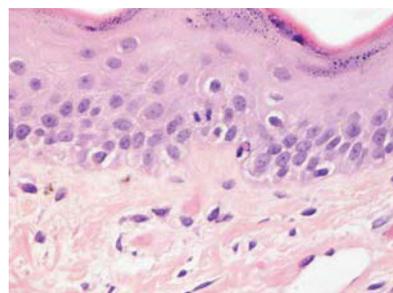


Figure 3. Histology. Hematoxylin-eosin, $\times 40$.

Diagnosis

Neonatal lupus erythematosus (NLE)

Course

An electrocardiogram was normal and transaminase levels returned to normal within a few weeks. The mother was diagnosed with systemic lupus erythematosus and was positive for ANA and anti-Ro/SSA.

Three months later the lesions had disappeared spontaneously, leaving atrophic, hypopigmented macules with superficial telangiectases.

Comment

NLE is characterized by the development of cutaneous lesions or cardiac conduction defects in newborns of mothers with anti-Ro, anti-La, or rarely anti-U1RNP antibodies. This rare entity affects 1 in every 12 000 to 20 000 live newborns.¹⁻³

Around 50% of patients with NLE have cutaneous lesions and the other 50% have a heart condition; 10% of all NLE cases present both cardiac and cutaneous disease. Liver and blood abnormalities may also be present, and pulmonary, neurologic, or gastrointestinal abnormalities may occur occasionally.¹⁻³

Cutaneous lesions may be present at birth, but usually appear within the first 2 months of life.²⁻⁵ The lesions are often triggered by phototherapy,² although UV radiation is not essential for development.^{4,5} The most common skin manifestations are annular erythematous, desquamative plaques, similar to those of subacute cutaneous lupus erythematosus. Another, very common, sign is the appearance of periocular erythema in the form of "raccoon eyes." The presence of purpura, telangiectases, congenital telangiectatic cutis marmorata, erythema multiforme-like, or xanthomatous, atrophic, or cicatricial lesions have also been reported.^{3,5,6}

The cardiac manifestations, which will condition the prognosis, usually consist of complete atrioventricular block and may be evident already in utero. This condition is usually permanent due to the replacement of conduction tissue with fibrous tissue. The mortality among patients is 10%, despite pacemaker implantation.^{2,3}

About 50% to 60% of mothers are asymptomatic when the newborn is diagnosed,^{1,3} although follow-up is essential

because of the risk of developing a connective tissue disease. The probability of having children with the condition in future pregnancies is also 25% higher.^{1,2}

Histology may be nonspecific or resemble the results seen in cutaneous lupus erythematosus. Direct immunofluorescence of skin lesions is negative in half the cases, and therefore, negative histology does not preclude a diagnosis of NLE.³

Both the mother and the newborn should undergo a laboratory workup consisting of complete blood counts, coagulation study, liver and renal function, erythrocyte sedimentation rate, lactate dehydrogenase, C-reactive protein, and urinary sediment. The diagnosis should also be confirmed by an immunologic study that includes ANA, anti-DNA, anti-Sm, anti-Ro, anti-La, anti-U1RNP, anticardiolipin, complement, syphilis serology, and rheumatoid factor.³ Although simultaneous cutaneous and cardiac lesions are rare, an electrocardiogram should be performed.³

Cutaneous lesions are self-limiting within 6 to 8 months (time required to clear maternal antibodies); however, photoprotection is essential. Low-strength topical corticosteroids or other topical immunomodulators can also be used. Persistent telangiectases can be treated with pulsed dye laser.^{1,2,4}

Children with NLE must receive follow-up due to a higher risk of developing an autoimmune disease.¹⁻³

Conflicts of Interest

The authors declare no conflicts of interest.

References

- Peñate Y, Luján D, Rodríguez J, Hernández-Machín B, Montenegro T, Afonso JL, et al. Lupus eritematoso neonatal: 4 casos y revisión clínica. *Actas Dermosifiliogr.* 2005; 96:690-6.
- Requena Caballero C, Navarro Mira M, Sánchez Carazo JL, Febrer Bosch I, Aliaga Boniche A. Lupus eritematoso neonatal. Descripción de un caso y revisión de la literatura. *Actas Dermosifiliogr.* 2001;92:215-9.
- Aparicio Español G, García-Patos Briones V, Castells Rodellas, A. Lupus eritematoso neonatal. *Piel.* 2002;17: 353-9.
- Elish D, Silverberg NB. Neonatal lupus erythematosus. *Cutis.* 2006;77:82-6.
- Diociaiuti A, Paone C, Giraldo L, Paradisi M, El Hachem M. Congenital lupus erythematosus: case report and review of the literature. *Pediatr Dermatol.* 2005;22:240-2.
- See A, Wargon O, Lim A, Sugo E, Le K. Neonatal lupus erythematosus presenting as papules on the feet. *Australas J Derm.* 2005;46:172-6.