Neonatal Lupus Erythematosus

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Neonatal lupus erythematosus is an immune-mediated disease that rarely is associated with congenital heart block. Transient hepatitis, thrombocytopenia and anemia can also occur.

We describe a case of neonatal lupus erythematosus and review the clinical and laboratory manifestations of this rare disease. Its only clinical sign might be an annular facial rash. The rash is usually diagnostic, but can be confused with other annular lesions. Because of the possible serious complications of undiagnosed lupus, a thorough evaluation of both child and mother is required when this diagnosis is entertained.

Case Report

A 5-month-old, full-term, male infant, whose birth was without complications, was brought to the clinic with a 4-month history of erythematous, atrophic, annular plaques distributed about the forehead, periorbital ridge, and temples. There was no associated scale or telangiectasia (Figure 1). He had no organomegaly, and auscultatory findings of his heart and lung examinations were normal. Tinea faciei had been diagnosed and treated accordingly for several months, but the infant then was referred for a second opinion because of persistent symptoms. A diagnosis of neonatal lupus erythematosus was suspected, and appropriate laboratory evaluation was initiated.

Maternal serologic examination showed an antinuclear antibody titer of more than 1:1280; anti-Ro (SS-A), anti-La (SS-B), and anti-U_1RNP antibodies were also detected. The infant had a positive antinuclear antibody titer and a negative anti-U_1RNP test. No tests for anti-Ro (SS-A) and anti-La (SS-B) antibodies were conducted because of laboratory error. His transaminase levels and white cell count were elevated. Findings on an echocardiogram were unremarkable. His medical history was notable only for recent otitis media. There was no family history of lupus or connective tissue disease. The mother was in good health and had no other medical problems, and she took no medications. When examined, she had no abnormal findings, and she denied joint aches, photosensitivity, or any other symptoms related to lupus or Sjögren syndrome. She was referred for an evaluation with an internal medicine consultant, and the child was treated with a low-potency topical steroid. The mother was told to keep the child out of the sun and to apply sunscreen. His rash cleared completely within 6 weeks.

Comments

Neonatal lupus erythematosus is an uncommon immune-mediated disease associated with the transplacental transfer of maternal immunoglobulin G autoantibodies. In 95% of cases the autoantibody is anti-Ro (SS-A), but it can be anti-La (SS-B) or anti-U_1RNP.[1] Any of these autoantibodies can be found alone or in combination. There are even reports of histologically documented cases of neonatal lupus erythematosus where none of the above-listed antibodies were found.[2] This finding suggests that some cases can be mediated by antibodies other than anti-Ro (SS-A), anti-La (SS-B) or anti-U_1RNP or perhaps by some factor or cofactor yet to be determined. In cases where only anti-U_1RNP antibodies are found, only cutaneous disease has been reported.[3]

A child will develop neonatal lupus erythematosus simply because its pregnant mother has anti-Ro (SS-A) antibodies. Neonatal lupus erythematosus occurs only in about 1% of these neonates.[4] If a anti-Ro (SS-A)-positive mother has one child with neonatal lupus erythematosus, only 25% of subsequent children born to this mother will be so af-

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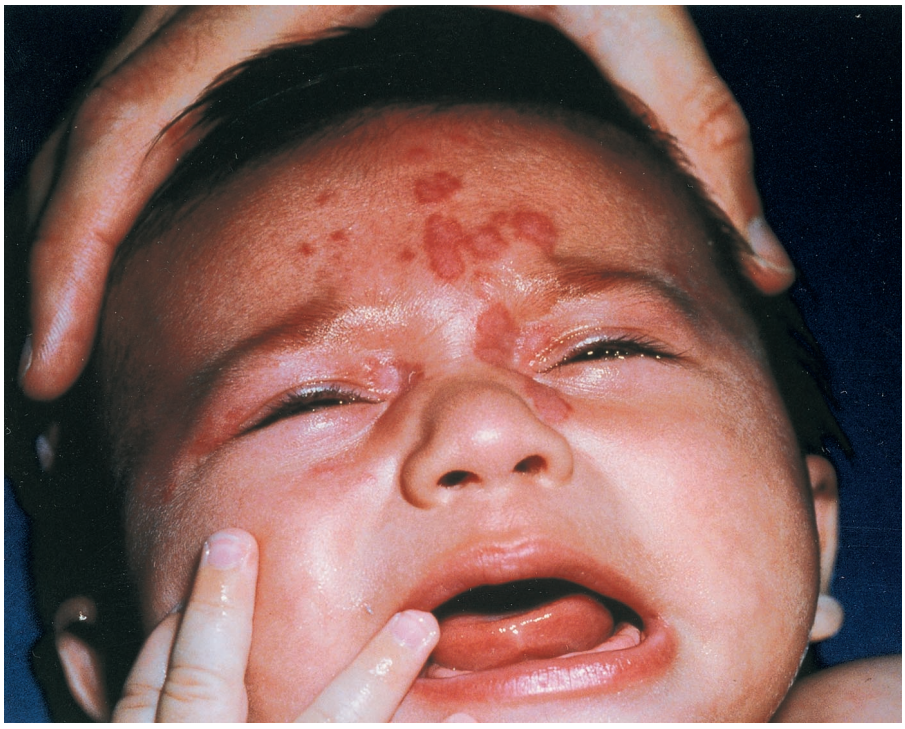

Figure 1. Erythematous, atrophic, annular plaques distributed on forehead, periorbital ridge, and temples, with no associated scale or telangiectasia.

fected.[4] These numbers are low, which is another reason to suspect a cofactor is involved in the pathogenesis of neonatal lupus erythematosus. Although congenital heart block can be found with this condition, it is rare. Its incidence ranges from 15% to 30% of affected infants.[1] This form of congenital heart block is associated with high morbidity; 50% to 70% of patients require pacemakers.[1,5] Transient forms of hepatitis, thrombocytopenia, and anemia can also occur.[6–9]

Skin lesions and heart block are seen simultaneously in less than 10% of patients with neonatal lupus erythematosus.[10] Cutaneous lesions consist of transient nonscarring erythematous annular plaques with a predilection for periorbital and photodistributed areas. Lesions generally appear within the first 2 months of life and resolve within 4 to 6 months when maternal antibodies disappear.[11] Rarely, remnant telangiectasias can occur at previously affected sites.[10]

A considerable proportion of mothers of affected infants are asymptomatic (40%). They might have Sjögren's syndrome, systemic lupus erythematosus, rheumatoid arthritis, overlap syndrome, or even leukocytoclastic vasculitis.[4,12] Asymptomatic mothers do not invariably become ill, and if they do develop lupus erythematosus, it is not likely to be life threatening.[3] Although neonatal lupus erythematosus has a very characteristic appearance, tinea faciei, a photodistributed drug eruption, urticaria, and annular erythemas are in the differential diagnosis. The diagnosis of neonatal lupus erythematosus is generally based on clinical findings when there are maternal and or neonatal autoantibodies. Determination of titers for antinuclear antibody, anti-Ro (SS-A), anti-La (SS-B) and anti-U_1RNP antibody is recommended for diagnosis. Liver function tests and a complete blood count should also be obtained. Skin biopsy is usually not required to establish the diagnosis. Histologic findings are similar to those of subacute cutaneous lupus erythematosus.[11] All patients suspected of having neonatal lupus should have a thorough cardiac examination.

Management of the cutaneous lesions of neonatal lupus erythematosus requires sun avoidance, sunscreen, and low-potency topical corticosteroids to hasten resolution.

Conclusion

Neonatal lupus erythematosus is a rare disease that can have a characteristic erythematous annular fa-

cial rash. The rash can be confused other annular rashes, including tinea faciei, as it was in this patient. In 95% of the cases, the diagnosis can be confirmed by a positive anti-Ro (SS-A) antibody test. Other laboratory studies that can aid in the diagnosis are anti-La (SS-B) or anti-U_1RNP antibody tests. Cardiac complications occur in up to 75% of cases, and congenital heart block is reported in 15% to 30% of affected patients.[1] An electrocardiogram is recommended to screen for heart block. In our patient, an echocardiogram was part of inclusive screening for other possible cardiac problems, such as transposition of the great vessels, patent ductus arteriosus, septal defects, and endocardial fibroelastosis.[1] Also, a complete blood count with platelet count and liver function tests are recommended for infants suspected of having neonatal lupus.

Spontaneous resolution is the natural course of the cutaneous lesions; however, sun avoidance and low- to mid-potency topical steroids are appropriate treatments. Treatment of the heart block is not necessary unless the child has heart failure.

Early recognition, evaluation, and treatment by the family physician could help to avoid the possible serious complications of neonatal lupus.

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