

Neonatal lupus erythematosus in dizygotic twins with anti-RNP antibodies

Sirs,

Neonatal lupus erythematosus (NLE) is an acquired neonatal disorder caused by placental transport of maternal IgG autoantibodies. In most infants, anti-SSA/Ro antibodies (95%) or anti-SSB/La antibodies (60–80%) are found (1, 2), but a few cases with only anti-ribonucleoprotein (RNP) antibodies have been reported (3). NLE can develop in children born to mothers with Sjögren's syndrome, systemic lupus erythematosus (SLE), and mixed and undifferentiated connective tissue diseases, but may also occur in asymptomatic mothers who possess the pathogenic autoantibodies (1, 4, 5).

In infants, especially with anti-SSA/SSB antibodies, NLE can present with cardiac involvement (mainly due to atrioventricular block), which can be irreversible, or with transient clinical features, such as cutaneous lesions and hepatobiliary, haematological and neurological abnormalities (1, 6).

The peculiar skin manifestations, that mimic clinically and pathologically subacute lupus erythematosus, have led to naming the disorder NLE (2). In 80% of cases, the cutaneous rash is not present at birth and occurs in the following weeks. It is usually characterised by erythematous annular lesions of the face and scalp, and to a lesser extent, of the trunk and arms, sometimes with central clearing or scaling (2, 6). A typical lesion is the inflammatory rash of the upper eyelids with a raccoon-like appearance (1). The lesions generally clear in a maximum of one year, in parallel with the disappearance of the circulating autoantibodies. In most cases, there is a complete recovery, but in 20% of cases telangiectasia, atrophy or hyperpigmentation can persist.

Herein, we provide the first report of twins with NLE born to a mother with mixed connective tissue disease (MCTD) who had only anti-RNP antibodies (anti SSA/SSB negative). The clinical manifestations of the mother included Raynaud's phenomenon, esophageal dysmotility, arthritis and interstitial lung disease. During pregnancy, she was given hydroxychloroquine and aspirin. Two dichorionic diamniotic twins were born at 31+4-week gestation by caesarean section because of maternal eclampsy. Maternal serology for rubella, CMV, syphilis, HIV and HCV was negative. Because of prematurity and respiratory distress, the twins were transferred to the neonatal ICU, where they received respiratory support with high-flux nasal cannula for a week, together with antibiotic prophylaxis for infectious risk. Due to increased serum bilirubin, both twins underwent phototherapy for 48 hours, with normalisation of bilirubin level. No other laboratory abnormalities were

Fig. 1.



found. EKG and echocardiogram were normal, as were neurological and ophthalmologic investigations. Both infants were discharged from ICU after one month in good general conditions. At 45 days of life, they were brought to our attention because of the simultaneous appearance of multiple annular erythematous lesions with central clearing over the scalp, face and trunk (Fig. 1; the authors acquired the written consent of the family to publish the work and the images).

In both twins, positive speckled antinuclear antibodies at a titre of 1:160 and 1:320 respectively, and anti-RNP antibodies were detected (ELISA confirmed by Western Blot, index 52 and 48), while anti SSA/SSB were negative. The complete blood count and liver function tests were normal, and at EKG no conduction defect was found. Photoprotection and a moisturising cream were prescribed, and 6 months later, the skin lesions had completely disappeared in both twins.

The development of NLE due to the placental transfer of anti-RNP antibodies from a mother negative for anti-SSA/SSB antibodies is rare, as only 19 cases are reported in the literature (3). Only a few instances of NLE have been described in twin neonates, and among them there is just one single report in which anti-RNP antibodies were detected in only one twin (7). Our report strengthens this possible association by describing for the first time the occurrence of NLE in dizygotic twins born from an anti-RNP-positive and anti-SSA/SSB negative mother.

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