Neonatal lupus erythematosus: Report of a case with cutaneous, hematological and hepatobiliary findings

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Neonatal lupus erythematosus is an autoimmune disorder mainly affecting the heart and skin. It is the most common cause of congenital heart block. In addition, hematological, hepatobiliary and neurological involvement may occur. Herein, we report a 23-day-old infant presented with annular, erythematous, and scaly and atrophic lesions on the face and trunk. Based on the clinical, laboratory and histopathological findings, she was diagnosed as neonatal lupus erythematosus. Neonatal lupus eryhtematosus should be considered in infants presenting with annular skin lesions, and we present this case to highlight the value of high index of clinical suspicion in diagnosis.

Key words: cutaneous, hematological, hepatobiliary, neonatal lupus.

Neonatal lupus erythematosus (NLE) is a rare disease caused by transplacental passage of maternal autoantibodies. The disease mostly affects the skin and the heart, but hematological, hepatobiliary and neurologic abnormalities may also be seen[1]. Although most of these findings are transient, cardiac disease may cause significant morbidity and mortality[1,2]. We report a case of NLE with cutaneous, hematological and hepatobiliary findings.

Case Report

A 23-day-old female infant was referred to our outpatient clinic for skin lesions on her face and trunk present since her birth. She was born at 34 weeks' gestation by cesarean section due to oligohydramnios and weighed 1800 g. The mother was 30 years old, gravida 3, para 3 and did not have any known systemic diseases.

Dermatological examination revealed numerous annular, erythematous, slightly scaly and centrally atrophic patches on the trunk, neck and face, particularly on periorbital regions (Figs. 1-3). Laboratory tests showed trombocytopenia (46000/mm^3), leukopenia (3100/mm^3), direct hyperbilirubinemia (3.55 mg/dl) and slightly elevated aspartate aminotransferase levels. Cranial and abdominal ultrasonography were normal.

A skin biopsy was obtained with a presumptive diagnosis of neonatal lupus. Histopathological examination revealed some colloid bodies underneath the epidermis in some focal areas, pointing out a mild interface change (Figs. 4 and 5). Also, there was a prominent accumulation of mucopolysaccharide within the dermis. Further laboratory tests showed positive anti-Ro/SSA in the patient and positive anti-Ro/SSA, anti-La/SSB and low titre anti-nuclear antibody (ANA) (1:100) in the mother. Physical examination of the mother did not reveal any sign or symptom related to connective tissue diseases.

Based on the clinical, histopathological and laboratory findings a diagnosis of neonatal lupus erythematosus was made.

Cardiologic examination with electrocardiography and echocardiography did not show any conduction system abnormality or cardiomyopathy. Treatment with methylprednisolone aceponate cream was

planned for skin lesions and she was referred to pediatrics for hematological and hepatobiliary involvement, but she was lost to follow-up.

Discussion

Neonatal lupus erythematosus is an uncommon autoimmune disease resulting from the passage of maternal autoantibodies through placenta[2]. In addition to maternal antibodies (most commonly anti-Ro/SSA and anti-La/SSB, rarely anti-U1RNP), fetal genetic and environmental factors are also accused in the pathogenesis[1,3]. The incidence of NLE is approximately between 1 in 12,500 to 20,000 live births and slightly higher in girls and premature infants[3].

Most of the mothers are asymptomatic at the time of NLE diagnosis, but the others may have systemic lupus erythematosus, Sjögren's syndrome, rheumatoid arthritis or undifferentiated connective tissue disease[4].

The disease may present with cutaneous, cardiac, hematological, hepatobiliary and rarely neurological findings[1-3]. In a recent review of 17 patients with NLE; cutaneous, cardiac, hepatobiliary and hematological involvement was found in 71%, 65%, 53% and 35% of infants, respectively[3].

Cutaneous findings may be present at birth but usually develops within the first few weeks. Skin lesions consist of erythematous, annular or polycyclic, centrally atrophic plaques resembling the lesions of subacute cutaneous lupus erythematosus (SCLE) and most commonly affect the face and scalp but can involve any part of the body[4,5]. The extensive confluent erythema in the periorbital region is referred to as "raccoon-eye" or "owl-eye". In addition to typical lesions, lesions resembling seborrheic eczema or a fungal infection may also be seen[5]. Photosensitivity may be present but is not requisite for the development of lesions. Histopathologic findings are similar to those seen in SCLE and consist of vacuolar degeneration at the dermal-epidermal junction and a perivascular lymphocytic infiltrate[2,6].

Clinical differential diagnosis of annular, erythematous lesions also includes seborrheic dermatitis, atopic dermatitis, tinea corporis, psoriasis, erythema annulare centrifugum, annular erythema of infancy and congenital syphilis[7].

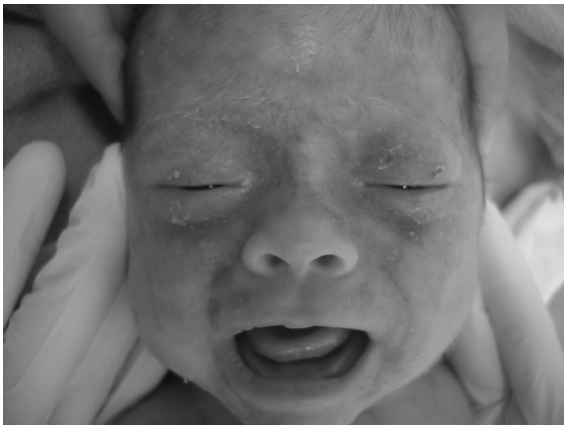

Fig. 1. Periorbital erythematous, scaly patches giving "raccoon-eye" appearance.

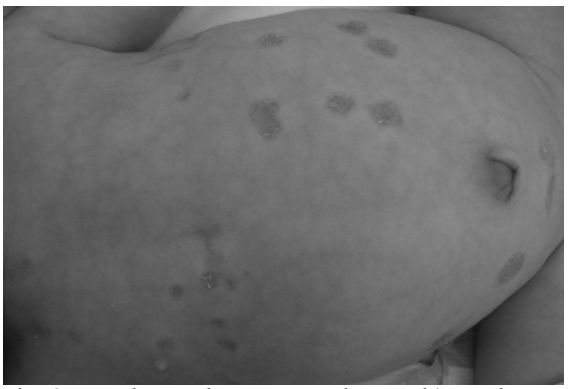

Fig. 2. Annular, erythematous, scaly, atrophic patches on the anterior aspect of the trunk.

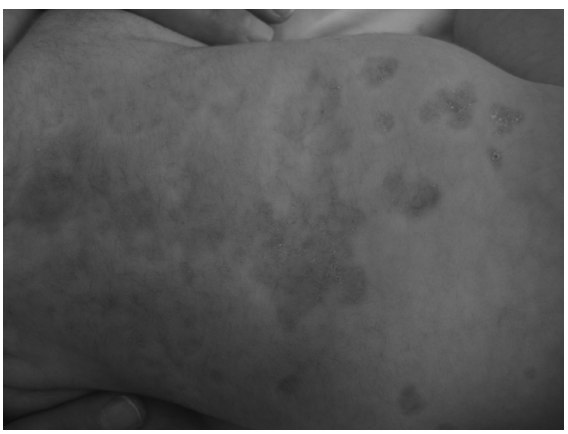

Fig. 3. Annular, erythematous, atrophic lesions on the back.

Cardiac disease typically manifests with congenital heart block (CHB) and cardiomyopathy[6]. Congenital heart block usually occurs in utero and may be first detected in second trimester. It is associated with significant morbidity and mortality[1,2,6]. Early recognition and management of cardiac involvement is critical.

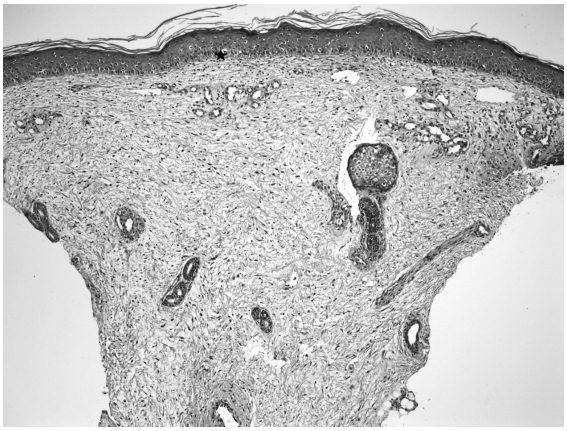

Fig. 4. Focal areas of interface change in the epidermis (star) and a loose dermis due to mucopolysaccharide accumulation (Hematoxylin&Eosin, X40).

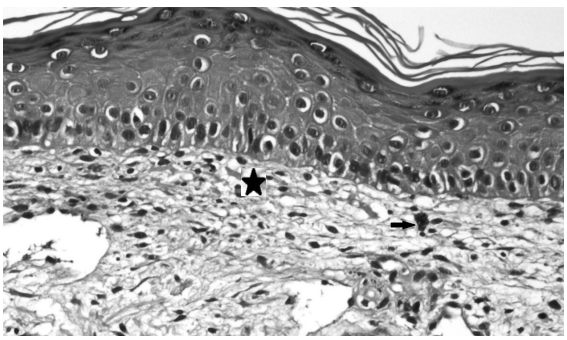

Fig. 5. Colloid bodies (star) and some pigment laden macrophages (arrow) underneath the epidermis (Hematoxylin&Eosin, x200).

Hepatobiliary involvement may cause direct hyperbilirubinemia, elevated transaminase levels, cholestatic hepatitis and fulminant liver failure[3]. Hematological abnormalities include trombocytopenia, leukopenia and hemolytic anemia[3,6].

The diagnosis of NLE is based on the presence of typical clinical features and autoantibodies in maternal or infant serum[3].

Infants with NLE should be followed-up for the subsequent development of autoimmune diseases[5]. Mothers, who were initially asymptomatic, may also develop connective tissue diseases with time[1]. Mothers of an affected child have 25% chance of having other affected infants, therefore monitoring of fetal heart rate is important for early detection of CHB[2].

Cutaneous and other noncardiac manifestations are transient and typically disappear within 6 to 8 months after birth as maternal autoantibodies are cleared from the infant's circulation[3]. For cutaneous lesions photoprotection and low potency topical corticosteroids are the treatment of choice. Skin lesions may sometimes leave residual dispigmentation, telangiectasia and rarely atrophic scars. Congenital heart block is frequently managed with pacemaker implantation. Systemic corticosteroids and intravenous immunoglobulin (IVIG) are occasionally used for the treatment of hepatobiliary and hematological abnormalities[3].

In conclusion, the diagnosis of NLE requires a high level of clinical suspicion and should be kept in mind in infants with CHB and typical cutaneous lesions. Long-term monitoring of these infants and their mothers is also important for possible detection of autoimmune diseases.

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