Neonatal lupus revealed maternal Sjögren's syndrome

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Neonatal lupus erythematosus is a disorder of the fetus or infant caused by certain maternal autoantibodies. Manifestations are usually cutaneous; systemic manifestations are rare. Here, we report a case of neonatal lupus erythematosus, which led to identifying maternal Sjögren's syndrome.

Keywords: neonatal lupus erythematosus, Sjogren's syndrome, autoimmune disorders

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INTRODUCTION

Neonatal lupus erythematosus (NLE) is a disorder of the fetus and neonate that can manifest as cutaneous, cardiac, hepatobiliary, or systemic abnormalities caused by maternal IgG autoantibodies that passively cross the placenta and enter the fetal blood. Although all infants with NLE are born to mothers with autoantibodies, the majority of mothers do not have any diagnosed rheumatic disease, such as Sjögren syndrome or systemic lupus erythematosus (SLE) [1,2]. In this article, we report a case of NLE, which led to a diagnosis of maternal Sjögren's syndrome.

CASE PRESENTATION

A three-month-old Iranian male infant was referred to our clinic with rashes on the cheeks and distal extremities commencing two months earlier, treated with moisturizers and topical corticosteroids without a favorable response. On the physical examination,

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there were annular lesions on the upper and lower extremities, mainly in the extensor areas. Some lesions featured erythema, scaling, and crusting (Figure 1). Also erythematous plaques on the cheeks were detected, which worsened with sunlight exposure (Figure 2).

Initial laboratory tests revealed no abnormal findings. A positive result for the ANA test was reported. The ANA profile of the infant showed strong positivity for both Anti-Ro 60 & Ro 50. The skin biopsy, laboratory findings, and clinical presentation were suggestive of neonatal lupus erythematosus. Electrocardiogram and echocardiography were performed with no abnormal findings. Other laboratory parameters were within normal limits.

The mother had not previously received a diagnosis of an autoimmune disorder. She only mentioned mild arthralgia in both hands and a history of induced therapeutic abortion following a cardiac block of the fetus three years ago. She noted the frequent use of artificial tears for dry eyes. Lab tests of the mother showed positivity for ANA, anti-SSA & anti-SS-B. Serum complement levels were normal (Table 1). A rheumatologist diagnosed her with primary Sjögren's disease.

In the follow-ups, a reduction of her symptoms was observed with suitable treatment. We advised her that any further pregnancies need to be under the supervision of her rheumatologist. The infant was treated with topical steroids and followed up every two months until the age of 12 months, with a lab panel consisting of a complete blood count and liver function tests. The skin lesions gradually disappeared without leaving any scars.

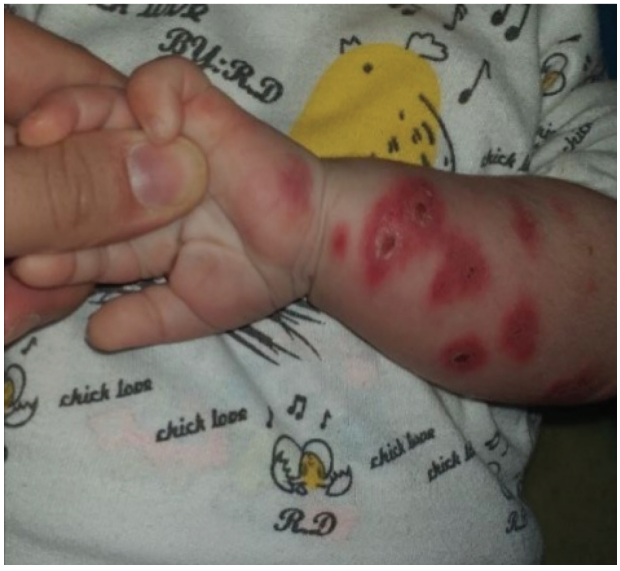

Figure 1. Annular erythematous plaques with some crusting on the upper extremity.

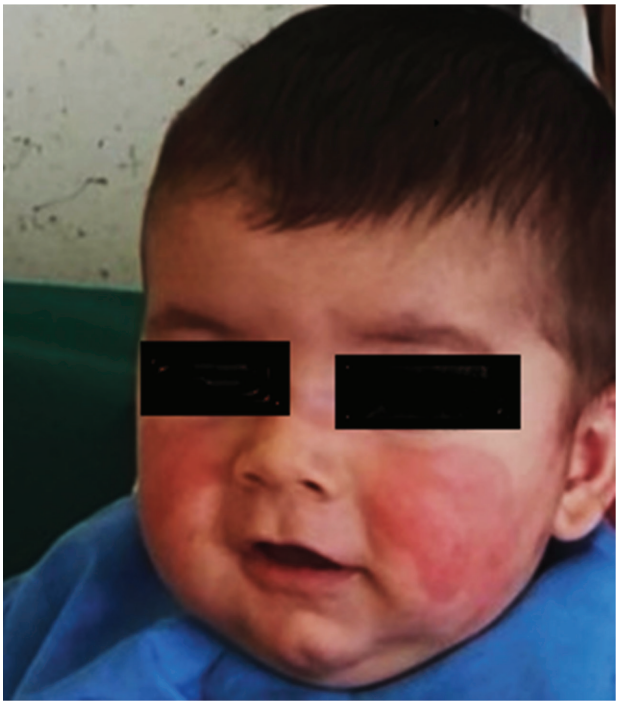

Figure 2. Photosensitive erythematous plaques on both cheeks.

Both parents provided written informed consent for the publication of this case report.

Ethical consideration

An informed consent form was obtained from the child's parents for reporting the case. As this was a case report, ethical code is not applicable.

DISCUSSION

Neonatal lupus erythematosus (NLE) is a rare condition caused by the passive transfer of autoantibodies from the mother to the fetus. Autoantibodies (most commonly anti-Ro/SSA and anti-La/SSB; rarely anti-U1RNP) present in the mother's blood cross the placenta and enter the fetal circulation. The presence of autoantibodies is necessary but not sufficient to cause the disease; genetic and environmental triggers are also important [1]. The condition was first described in 1954 with a case report of an infant with cutaneous lesions whose mother was diagnosed with SLE. The incidence of the disease is estimated at between 1 in 12,500 to 20,000 live births [2].

Table 1. Laboratory test results of neonate's mother

Test	Result	Unit	Reference range
Serology			
CRP	3	mg/L	< 6
RF	39.14	IU/ml	Po: > 14
Direct coombs	negative	---------	-------
Indirect coombs	negative	---------	-------
Immunoassays - Immunity markers			
C3	118	mg/dL	89-187
C4	24.3	mg/dL	16-40
CH50	100	mm	70-130
Immunoassays - Autoimmune disease			
Anti SCL70	1.4	U/ml	NL: < 15, B:15-25, E: > 25
ANA	> 9	U/ml	Neg: < 0.8, B:0.8-1.2, Po: > 1.2
Anti-dsDNA	15.7	IU/ml	Neg: < 50, B:50-60, Po: > 60
Anti RNP-sm	3.0	U/ml	NL: < 15, B:15-25, E: > 25
Anti Jo	0.5	U/ml	Neg: < 15, B:15-25, Po: > 25
Anti SSA-RO	> 200	U/ml	NL: < 15, B:15-25, E: > 25
Anti SSB-LA	52.3	U/ml	NL: < 15, B:15-25, E: > 25
Anti-centromere	0.9	U/ml	NL: < 10, E: ≥10
Anti-dsDNA (ELISA) (eight months later)	0.35	index	Neg: ≤0.9, Equi: 0.91-1.09, Po: ≥1.1

Cutaneous lesions, a common clinical manifestation of NLE, usually appear in the first weeks of life, but may also be present at birth. The most common age for the appearance of skin lesions is six weeks, but it may not be recognized until 12 to 14 weeks. Characteristic annular or macular rashes usually appear on the face and scalp but may involve any part of the body including the trunk and extremities. 'Owl eyes', a distinctive facial involvement, typically in the periorbital area, may also be present. New lesions may appear for several months, but they rarely develop beyond six months, consistent with the time of disappearance of most maternal antibodies from the infant's circulation. The mean duration of the rash is 17 weeks. The lesions may be induced or exacerbated by sun exposure. Rashes appearing after phototherapy for neonatal jaundice have also been reported. The lesions usually resolve without scarring, although some mild epidermal atrophy and/or hyper/hypopigmentation may persist in the long term [3-4].

Biopsies of cutaneous lesions usually show vacuolar degeneration at the dermal-epidermal junction and perivascular lymphocytic infiltration, as in our patient. Histopathologic findings of skin lesions are similar to those seen in a nonscarring type of cutaneous SLE called subacute cutaneous lupus (SCLE) [5].

Cardiac manifestations of NLE include conduction system abnormalities (ranging from first-degree to complete heart block), cardiomyopathy, valvular dysfunction, and endocardial fibroelastosis. Although a complete heart block is irreversible, other manifestations of NLE are usually transient and self-limiting. Cardiac conduction disorders, especially complete heart block, are treated with cardiac pacing and occasionally with intravenous immunoglobulin and corticosteroids [6].

Hepatobiliary involvement can present as transient conjugated hyperbilirubinemia, transient transaminase elevations, hepatomegaly, and/or liver failure. Hematological abnormalities include transient thrombocytopenia, neutropenia, and anemia [7].

Mothers of infants with NLE have a 25% chance of having other affected infants, so monitoring of fetal heart rate in later pregnancies is important for the early detection of any conduction abnormalities. To prevent and manage cutaneous and fetal cardiac conduction disorders in mothers with positive autoantibodies, maternal treatment with hydroxychloroquine has been suggested [8]. Babies with NLE will also be at some increased risk of developing connective tissue diseases. Long-term follow-up of both the asymptomatic mother and the infant is usually recommended [9].

CONCLUSION

We suggest if fetal heart disease especially

conduction disturbance is found in pregnancy assessments, the mother should be screened for autoantibodies. Also, in an infant with NLE, a detailed history should be taken from the mother in search of subtle signs of rheumatic diseases.

Acknowledgment

None.

Authors' contributions

Molood Safarirad and Ahmad Vosughi Motlagh proposed the case to be reported.

Sadegh Alavi Reza Paya drafted the manuscript.

Navid Namazi and Nastaran Namzi critically revised the manuscript.

All authors gave final approval and agreed to be accountable for all aspects of the work ensuring integrity and accuracy.

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