

Case Report

Neonatal Lupus Erythematosus in a 2-Month-Old Girl: A Case Report

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Abstract: Neonatal lupus erythematosus is a rare acquired disease involving the fetus and the newborn. It is caused by autoantibodies (mainly anti-SSA/Ro and anti-SSB/La antibodies) in the mother entering the fetus through the placenta to cause autoimmune reactions. The most common clinical manifestations of neonatal lupus erythematosus include skin involvement and congenital heart block, as well as blood, respiratory, neurological, and digestive system abnormalities. Patients often go to pediatrics or dermatology with rash as the first symptom. Because the rash is atypical, there is often a risk of misdiagnosis or missed diagnosis. We report a case of 2-month-old baby. Physical examination showed that dark red infiltrating macula was scattered on forehead and plantar. ANA positive, anti-SSA/Ro antibody positive, anti-SSB/La antibody positive, no involvement of the heart or other organ systems was found. Her mother was a patient with lupus erythematosus. The diagnosis of neonatal lupus erythematosus was made according to the history and clinical manifestations, symptomatic treatment was given to the rash site, and after 4 months, the rash disappeared at a follow-up visit. ANA, Anti SSA/Ro antibodies and anti SSB/La antibodies all turned negative. The etiology, clinical manifestation, prevention and treatment of neonatal lupus erythematosus were discussed in combination with existing literature. It was emphasized that autoantibody testing should be carried out for women and pregnant women with reproductive requirements in order to detect and prevent neonatal lupus erythematosus in time. For unexplained skin rashes of infants, if the mother has a history of connective tissue disease, the possibility of neonatal lupus erythematosus should be considered.

Keywords: Erythematosus, Neonatal Lupus, Anti-SSA/Ro, Anti-SSB/La, Prognosis

1. Introduction

Neonatal lupus erythematosus (NLE) is a disease that occurs in the fetus and neonates. Maternal specific autoantibodies make it have unique clinical manifestations. The disease mainly affects the heart and skin, and can also affect organs and systems such as the liver, lungs, blood system, nervous system, etc, which is a passive acquired autoimmune syndrome [1]. Patients often seek medical advice from pediatrics or dermatology with rash as the first manifestation. The rash of the disease can be round or ring-shaped erythema, accompanied by edema or scales. The rash of this disease is atypical, and it is easy to be confused with other skin diseases, so it is easy to be misdiagnosed or missed. In order to make pediatricians and dermatologist

better understand and treat this kind of disease, a case of neonatal lupus erythematosus admitted by our department is reported as follows, and the etiology, clinical manifestations, prevention and treatment of this disease are discussed in combination with literature.

2. Case Report

2.1. Clinical Data

A 2-month-old female infant was treated in our department due to plantar and facial erythema for more than 1 month. More than a month ago, the infant's plantar gradually developed erythema with no obvious inducement. So she was taken to the local hospital for treatment and was diagnosed with dermatitis. Then she was treated with calamine lotion and other

external drugs (the specific medication is unknown), with poor effects. Erythema gradually appeared on the face, next, she was sent to Gucheng County Hospital of traditional Chinese medicine for further diagnosis and treatment. The diagnosis was unknown. She was treated with intravenous infusion (the specific drug was unknown) and topical drugs (bifonazole ointment, other drugs were unknown), and there was no obvious effect. The rash is gradually increasing, and the infant has no other special discomfort during the course of the disease. For further diagnosis and treatment, the patient came to our department for treatment. The patient was a full-term cesarean section baby, and denied the family history of genetic disease, his mother had a history of similar skin diseases on her face one year ago. The immunological examination in our department showed that ANA was positive, the titer was 1:320, anti-SSA/Ro and anti-SSB/La antibodies was positive. Physical examination: vital signs are normal, no obvious abnormality is found in all system examinations, and the superficial lymph nodes of the whole body are not touched and swollen. scattered dark red infiltrative erythema can be seen on the forehead and plantar, without papules, with slightly raised edges, atrophy in the center, and small scales on the surface (Figures 1-2).

Laboratory examination: there were no obvious abnormalities in routine blood, urine and feces, liver and kidney function, rheumatoid factor and antistreptolysin-O. But ANA positive with a titer of 1:320, anti-SSA/Ro and anti-SSB/La antibodies positive, anti-RNP, anti-Sm, anti-ScL-70, anti-Jol-1 and anti-dsDNA antibodies negative; Fungal examination showed that spores and hyphae were negative; No obvious abnormality was found in ECG and color Doppler echocardiography.

2.2. Diagnosis and Treatment

According to the medical history, clinical manifestations and auxiliary examination results, the patient could be diagnosed as neonatal lupus erythematosus. The infant was treated with zinc oxide cream and dexamethasone cream for external use, and is required to avoid light and undergo regular follow-up. 2.3. Follow-up and Outcome.



Figure 1. Dark red infiltrating erythema is scattered on the forehead, the edge is slightly raised, atrophy can be seen in the center, and there are small scales on the surface.



Figure 2. Dark red infiltrating erythema is scattered on the sole of the foot, with fine scales on the surface.

After 4 months of follow-up, the skin lesions of the infant disappeared completely (Figures 3-4), and no obvious abnormalities were found in blood routine, liver and kidney function, ECG. Immunological indicators including ANA, anti-SSA/Ro and anti-SSB/La antibodies turned negative. The patient is still being followed up.



Figure 3. The erythema on the forehead basically subsided.



Figure 4. A. The skin lesions of the right foot basically disappeared. B. The skin lesions of the left foot basically disappeared.

3. Discussion

Neonatal lupus erythematosus (NLE) is a rare acquired disease involving fetuses and newborns [1]. Most scholars believe that it is caused by the autoimmune reaction caused by the autoantibodies in the mother (mainly anti-SSA/Ro

antibody and anti-SSB/La antibody) entering the fetus through the placenta [2, 3].

The main clinical manifestation of NLE is annular erythema of the skin or similar to seborrheic dermatitis [4]. Skin lesions often appear 4-6 weeks after birth, mostly on the face, scalp, around the orbit, and a few can occur in the limbs and non exposed parts. The most common skin rash is a circular congestive maculopapular or erythema, with scales and atrophy of the central part, clear and prominent edges, and the skin lesions generally subside spontaneously within 4-6 months after birth. In addition, it can also be accompanied by cardiac involvement. The most common form of cardiac involvement is incomplete or complete congenital heart block, which usually occurs from the second to third months of pregnancy. Once formed, it can last for several years [5-8]. Myocardial lesions have also been reported. Some children will also have thrombocytopenia, leucopenia, hemolytic anemia, jaundice, abnormal liver function, abnormal respiratory function, hyperbilirubinemia, etc., but most of them are transient damage, which can disappear or alleviate to varying degrees within 6-12 months [5].

With the clearance of anti-SSA/Ro and anti-SSB/La antibodies from the mother, the rash of most children with cutaneous NLE can completely disappear. As long as sunlight exposure is avoided, there is little need for drug treatment, and the prognosis is good. Individual children may still progress to systemic lupus erythematosus or other autoimmune diseases. If accompanied by persistent abnormal liver function, severe thrombocytopenia, hemolytic anemia, leucopenia, acute lupus pneumonia, heart block and other changes, a short course of hormone therapy prednisone 1-2mg/ (kg·d) or human gamma globulin can be used, and the course of treatment depends on the condition [4, 5]. At present, there is no effective treatment for newborns with severe heart block and myocardial damage. Newborns who survive eventually need to install cardiac pacemakers [9, 10]. In this study, the children only had annular erythema on the face and the soles of both feet, and the serum autoantibody was positive, which subsided after local topical drug treatment.

About 40% - 60% of the mothers of the newborns have no history of connective tissue disease [11], and some mothers can be diagnosed with lupus erythematosus, Sjogren's syndrome, rheumatoid arthritis, or other connective tissue diseases [10-12]. Antibodies produced by the mother and found in her serum belong to the immunoglobulin G group and enter the fetal blood flow from the time of placenta formation, that is, 12 weeks of gestation [13]. These antibodies may trigger a series of inflammatory reactions in the fetus and lead to neonatal lupus symptoms. In this study, the mother of the child had annular erythema on her face one year ago. The immunological examination in our department showed that ANA was positive, the titer was 1:320, anti-SSA/Ro and anti-SSB/La antibodies were positive, and she was diagnosed as subacute cutaneous lupus erythematosus. It is noteworthy that there are still some asymptomatic or mild mothers who have skin rashes and other discomfort after delivery, and gradually develop into systemic lupus erythematosus,

Sjogren's syndrome or other connective tissue diseases [2].

Therefore, for women with connective tissue disease who have the desire to have children, it is necessary to control their condition smoothly through effective treatment before considering pregnancy. Studies have found that taking antimalarial drugs during pregnancy can effectively reduce the risk of neonatal skin lesions [12] and heart block [14]. For women without connective tissue disease, relevant risk factors, such as ANA, anti-SSA/Ro and anti-SSB/La antibodies, should also be tested before pregnancy; Pregnant patients need to be tested for the above antibodies regularly and the risk situation has been investigated in time [2, 15-17]. Once NLE occurs, it is very important for the long-term follow-up and observation of newborns and mothers. Monitoring the relevant indicators of lupus erythematosus, identifying risk factors and conducting early intervention and treatment can reduce the occurrence of serious complications, thereby improving their quality of life.

4. Conclusion

We report a case of a 2-month-old girl who was diagnosed with neonatal lupus erythematosus. This is an acquired disease that affects both the fetus and the newborn. Autoantibodies in the mother's body are transferred to the fetus through the placenta, resulting in the onset of the disease. The skin manifestations are atypical and are easily misdiagnosed as other skin diseases. However, the disease can affect the heart, leading to cardiac conduction block, and can also lead to cardiomyopathy, thrombocytopenia, leucopenia Hemolytic anemia, jaundice, liver dysfunction, respiratory dysfunction, hyperbilirubinemia, etc. Therefore, clinicians are required to identify suspicious cases, make early diagnosis, and actively treat them; At the same time, detecting autoantibodies during prenatal testing for pregnant women, and early intervention and treatment for abnormal indicators and symptoms can reduce the risk of neonatal lupus erythematosus and the occurrence of late severe complications.

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Conflict of Interest

The authors declare that they have no conflict of interest.

Informed Consent

We have obtained informed consent from patients' families.

Author Contributions

Runchao Wang summarized and wrote the article. Haixia Jing contributed to literature review and editing of the manuscript. And Tingting Chen, Rui Wu and Wentao Liu

collected the case data.

Abbreviations

NLE: Neonatal lupus erythematosus
 ANA: antinuclear antibody
 anti-SSA/Ro: anti-Sjogren's syndrome A/Ro
 anti-SSB/La: anti-Sjogren's syndrome B/La
 RNP: Ribonucleoprotein
 dsDNA: Double stranded deoxyribonucleic acid
 Sm: Smith
 ECG: electrocardiogram

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