Case Report

Unusual Skin Manifestations in Neonatal Lupus Erythematosus

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Neonatal lupus erythematous (NLE) is a rare autoimmune disease caused by placental transfer of maternal anti-SSA/Ro or anti-SSB/La antibodies. It usually presents with transient cutaneous lesions, congenital heart block and other systemic symptoms. The authors report a case of neonatal lupus erythematosus who presented with targetoid-like lesions on both feet.

Keywords: *Neonatal lupus erythematosus, Targetoid lesion, Cutaneous symptoms*

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Neonatal lupus erythematous (NLE) is a rare autoimmune disease in neonates first described in 1954 by McCuistion and Schoch[1]. It is characterized by transient cutaneous lesions, congenital heart block and systemic abnormalities. NLE caused by transplacental passage of maternal autoantibodies (anti-Ro/SSA and/or anti-LA/SSB and less common U1 ribonucleoprotein)[2]. The clinical picture of cutaneous NLE is highly variable, with erythematous patches being the most common, especially in the periorbital area[6]. We report a case who manifested with unusual skin lesions on both feet.

Case Report

A full term, one-month-old female infant, presented to our pediatric dermatology clinic for evaluation of an erythematous rash on both feet for one week. She was born via cesarean-section. The patient's past medical history had been revealed as unremarkable. Her mother was healthy. There was no family history of autoimmune disease. On physical examination, she was found of symmetrical erythematous patches in the periorbital area (as shown in Fig. 1), and symmetrical annular plaques with central atrophy and telangiectasia on the plantar surface of both feet (Fig. 2). There was no pallor, no hepatosplenomegaly. Other physical findings were normal. Electrocardiogram (EKG) was normal without heart block. Echocardiography was also normal.

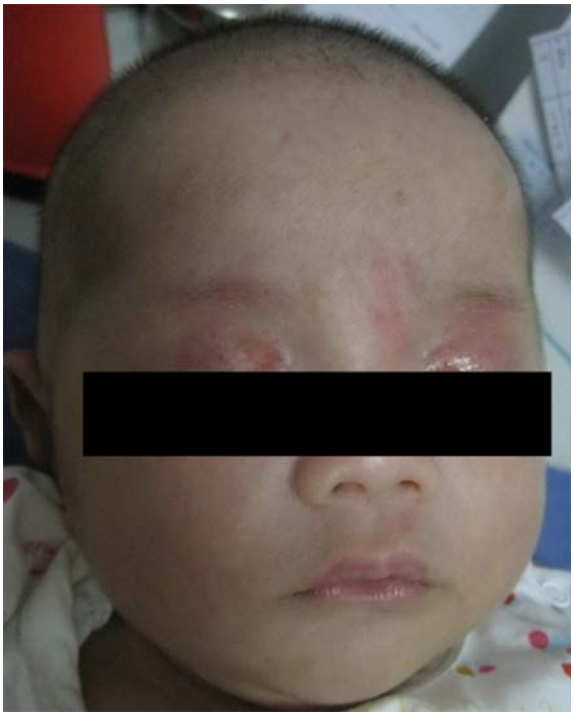

Fig. 1 Symmetrical erythematous patches in the periorbital area.

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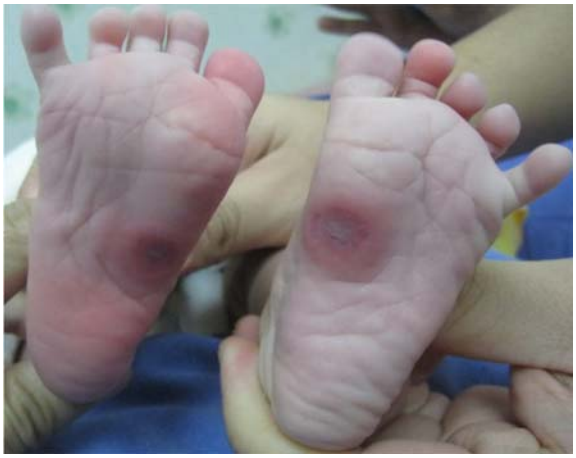

Fig. 2 Symmetrical annular erythematous plaques, with central atrophy and telangiectasia (tagetoid-like lesions) on the plantar surfaces of both feet.

Laboratory testing including liver function test (LFT) (AST 22 U/L, ALT 20 U/L, ALP 350 U/L, albumin 4.07 g/dL, total protein 6.03 g/dL, globulin 2 g/dL, total bilirubin 0.59 mg/dL, direct bilirubin 0.51 mg/dL, indirect bilirubin 0.08 mg/dL), complete blood count (CBC) (Hb 12.4 g/dL, Hct 37%, WBC 8,670/mm^3, neutrophil 15%, lymphocyte 70%, atypical lymphocyte 6%, monocyte 8%, eosinophil 1% platelets 300,000/mm^3), electrolytes, blood urea nitrogen (BUN) and creatinine (Cr) were normal. Erythrocyte sedimentation rate (ESR) was elevated (45 mm/hr). A diagnosis of NLE was confirmed by a positive anti-nuclear antibody (ANA) (speckle pattern 1:2,560 and nuclear pattern 1:2,560). Both anti-Ro/SSA antibody and anti-LA/SSB antibody were positive. Skin biopsy was not performed in this case. Her mother's serum also demonstrated positive anti-Ro/SSA antibody, anti-LA/SSB antibody and ANA.

Based on these clinical manifestations and laboratory tests, NLE was diagnosed. The patient was treated with Mometasone furoate 0.1% at both feet and a mild potency topical corticosteroid (0.02% triamcinolone acetonide) on her face. Sun protection was prescribed. The skin lesions gradually disappeared in 4 months. Her mother developed symptomatic SLE 9 months after the infant's diagnosis, presented as vasculitis in both hands.

Discussion

Neonatal lupus erythematosus (NLE) is an uncommon autoimmune disease in neonates. Pathogenesis is due to transplacental passage of maternal antibodies (anti-Ro/SSA and/or anti-LA/SSB and less common U1 ribonucleoprotein). It has been estimated that 1 to 2 percent of women with these autoantibodies will have seen in the baby with neonatal lupus erythematosus[2]. The most common clinical manifestations are skin, cardiac, hepatobiliary and hematologic systems. Cutaneous involvement occurs in roughly 70% in NLE patients[3]. It usually appears by 6 weeks after birth and generally disappears by 8 months of age. The "owl's eyes", which is characterized by annular erythematous or polycystic plaques with or without fine scales around the periorbital area, are the pathognomonic sign for diagnosis[4]. One study by Wisuthsarewong found various cutaneous manifestations as followed erythematous patches (91.7%), subacute cutaneous lupus erythematosus lesions (50%), petechiae (41.7%), persistent cutis marmorata (16.7%), and discoid rash (8.3%)[3].

Bullous or atypical targetoid-like lesions are less commonly found especially on the soles of the feet[5]. Periorbital erythematous patches were found in this patient, therefore, helping physicians to suspect the diagnosis of NLE, but without maternal history of SLE, confirmation with ANA, anti-Ro/SSA, anti-La/SSB and anti-U1RNP antibodies would lead to the diagnosis. Skin biopsy can be performed if the diagnosis is suspected but it is not mandatory. Interface dermatitis, keratinocyte damage, moderate hyperkeratosis, follicular plugging, vacuolar degeneration in the basal cell layer, and epidermal atrophy can be found from histologic examination. These findings are not pathognomonic and can also be found in other inflammatory diseases[6].

Topical corticosteroids and avoidance of exposure to sunlight are recommended for the management of skin lesions.

Cardiac manifestations include congenital heart block (first, second or third-degree heart block) and cardiomyopathy[6]. The pathogenesis of heart block is due to both anti-Ro/SSA antibody and anti-LA/SSB antibody damaging endocardial and myocardial tissue. The treatment with corticosteroid in fetal NLE with cardiac involvement may improve the heart block[7].

Hepatobiliary presentation includes elevation of liver enzymes and/or conjugated bilirubin. Hepatomegaly and splenomegaly may also present in some infants. Liver function usually normalizes within 6 months[8].

Abnormalities in the hematologic system can present widely with hemolytic anemia, thrombocytopenia, or neutropenia. Symptoms usually appear

by 2 weeks of life and disappear by 2 months of age[8]. Mortality and morbidity in NLE patients depend on organ involvement. Cutaneous NLE itself has good prognosis but it is associated with 6-10 fold risk of having next baby with cardiac NLE[6]. Long term follow-up is needed because children with NLE are prone to develop SLE or other autoimmune diseases later in life due to their genetic predisposition[8].

Conclusion

NLE is a rare autoimmune disease in neonates. In the presence of the atypical cutaneous lesions without maternal symptoms of SLE, Annular erythematous patches or plaques around the periorbital area are the clues to early diagnosis and appropriate treatment of the disease, thus preventing further morbidity and mortality later in life. Furthermore, careful planning and counselling for subsequent pregnancies would help prevent or reduce the risk of NLE in the future.

What is already known on this topic ?

Cutaneous manifestations are the most common presenting symptoms in patient with neonatal lupus erythematosus. Owl's eyes are the pathognomonic cutaneous sign for diagnosis.

What this study adds ?

This study reports a case presenting with atypical targetoid-like lesions which are unusual skin manifestation in neonatal lupus erythematosus.

Acknowledgement

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Potential conflicts of interest

None.

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อาการทางผิวหนังที่ไม่พบบ่อยในผู้ป่วยโรคลูปัสในทารกแรกเกิด

ธิติวัฒน์ ชัยกุล, สุธิดา ชัยธีระยานนท์

โรคลูปัสในทารกแรกเกิดเป็นโรคที่เกิดจากภูมิคุ้มกันทำลายตัวเองที่พบได้ไม่บ่อยสาเหตุเกิดจากการส่งผ่านของ anti-SSA/Ro หรือ anti-SSB/La แอนติบอดีจากแม่ไปสู่ลูก โดยทั่วไปมักมาด้วยอาการทางระบบผิวหนัง ระบบหัวใจ และระบบต่างๆ ทั่วร่างกาย คณะผู้นิพนธ์รายงานผู้ป่วยโรคลูปัสในทารกแรกเกิดที่มาด้วยอาการของผื่น target ที่เท้า
