

## Neonatal Lupus Erythematosus: Two Case Reports in a Single Centre

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### Abstract

Neonatal lupus erythematosus (NLE) is a rare autoimmune mediated disease that is associated with transplacental autoantibodies. It is characterised by cutaneous manifestations and may involve cardiac, haematologic, neurologic and hepatic abnormalities. We describe two cases with clinical, pathological and immunological findings compatible with neonatal lupus erythematosus, treatment and progress. Both their mothers were asymptomatic but were found to have autoantibodies. In view of the possible serious complications such as complete heart block, a high index of suspicion coupled with a thorough evaluation of both child and mother is required.

**Keywords:** Neonatal lupus erythematosus; antinuclear antibody (ANA); auto antibodies; anti-Ro; anti-La.

### INTRODUCTION

Neonatal lupus erythematosus refers to a clinical spectrum of cutaneous, cardiac and systemic abnormalities observed in newborn infant or neonates whose mothers have autoantibodies against Ro/SSA, La/SSB and less commonly, U1-ribonucleoprotein (U1-RNP)<sup>1,2</sup>. We describe 2 cases here that presented with different cutaneous clinical severity.

### CASE 1

The first case presented with rash on the face and scalp since birth. He was born at term with good Apgar score. Parents are non-consanguineous. Mother has history of 2 first trimester miscarriages but is otherwise asymptomatic. Examinations revealed multiple petechial rash over the face with annular patches over the right temporal region and scalp. He has mild neonatal unconjugated hyperbilirubinaemia but is otherwise clinically well, stable vital signs and no

remarkable systemic findings. Baseline investigations showed no abnormal findings. His erythrocyte sedimentation rate (ESR) was 50, antinuclear antibody (ANA) 1:1280, anti-Ro/ SSA 215, anti-La/SSB >240 and C3, C4 were normal. His mother's connective tissue screening showed ESR 60, ANA 1:2560 (pattern SSA) with both anti-Ro and anti-La detected. Electrocardiography and echocardiography were normal.

He was treated with mild topical steroids, sunscreen protection and advised sun avoidance. Biopsy was not consented. Mother was referred to medical team for treatment and follow up. At 6 weeks of life, he developed multiple photosensitive annular rashes over his face, trunk and back but was otherwise thriving well. His investigations then were ESR 30, ANA 1:320, anti-Ro and anti-La detected. The importance of sun avoidance and sunscreen usage was reemphasized to parents. Rashes subsequently resolved.



**Figure 1.1.** *Annular, discrete erythematous lesions over face at presentation.*



**Figure (1.2, 1.3).** *At 6 weeks old, similar lesions found on trunk after sun exposure.*

## CASE 2

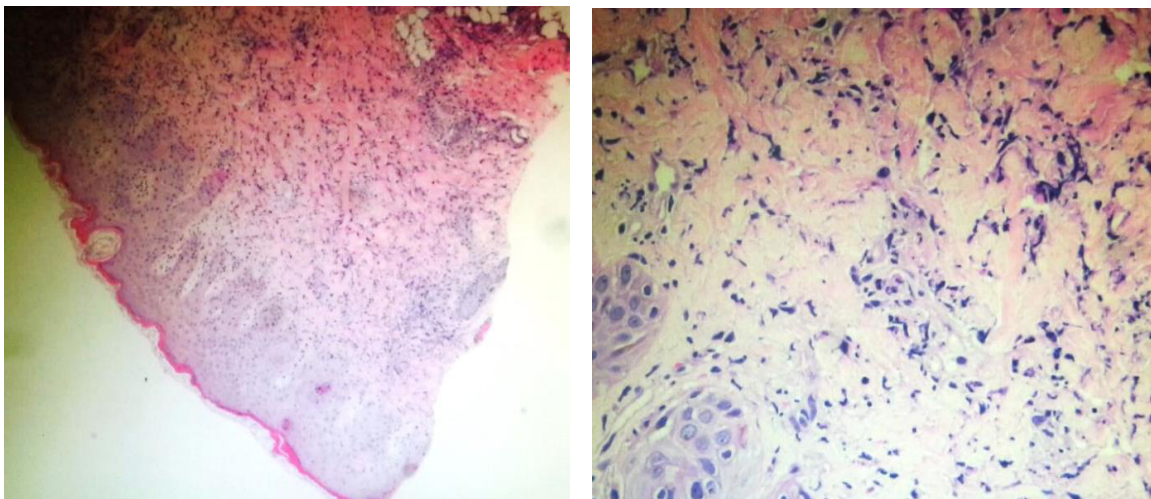
Case 2 is a baby girl presented with rash from 2 days old. It started over face which subsequently generalized to limbs and trunk. Mother has no significant antenatal and past medical history. Child was born term with good Apgar score. Child was treated with various topical medications but did not respond. At 3 weeks old, antibiotic was commenced when she developed fever and irritability. She was referred to our team a week later when she was not responding. On examination she was active, pink, irritable but consolable, normotensive anterior fontanelle with stable vital signs except spiking temperature. There were generalized annular plaques with well-defined borders over face, body and limbs involving the soles but sparing the palms. She has no remarkable systemic findings.

The provisional diagnosis was Sweet's syndrome, to exclude underlying neonatal lupus. She was covered

with Cefuroxime and started on Methylprednisolone, then continued with high dose oral prednisolone 2mg/kg/day. She was extensively investigated for the underlying cause(s). Her condition improved gradually, temperature settled after 2 days and clinically less irritable. Initial laboratory investigations showed raised alanine transaminase of 73U/L which subsequently normalized to 35U/L after 5 days. Her ESR was 64. Histopathology examination (HPE) from skin biopsy was in keeping with Sweet's syndrome. The laboratory results later confirmed that she has neonatal lupus erythematosus with low C3, C4, ANA 1:640 and anti-La detected. Mother's C3, C4 were normal, ANA 1:2560 with both anti-Ro and anti-La detected. Electrocardiography and echocardiography were normal. Prednisolone was tapered down and off. Mother was referred to medical team for treatment and follow up. Patient continued to improve and fully recovered thereafter.



**Figure (2.1, 2.2, 2.3).** Generalized discrete annular plaques involving trunk & limbs



**Figure (3.1, 3.2).** HPE showed dense neutrophilic infiltrations & papillary dermal oedema; X10 magnification (Figure 3.1) and X100 magnification (Figure 3.2)



### DISCUSSION

Neonatal lupus erythematosus is a condition first described almost 60 years ago by McCuiston and Schoch, who noted the presence of characteristic skin lesions on a baby born to a mother who suffered from lupus<sup>1</sup>. By the late 1970s, NLE was found to be caused by maternal transplacental passage of the anti-SSA/Ro antibodies from mother to fetus<sup>1</sup>. NLE is a syndrome characterized by skin, cardiac and systemic abnormalities, seen in newborn infants whose mothers have autoantibodies against Ro/SSA and/or La/SSB<sup>1</sup>.

Clinical manifestations that present in the fetus/neonate or infant result from trans-placental passage of maternal anti-Ro/SSA and/or anti-La/SSB and/or rarely anti-U1-RNP antibodies are termed NLE<sup>1</sup>. Approximately 98% of affected infants have maternal transfer of 1 or more of these autoantibodies<sup>3</sup>. However only 1-2% of mothers with these autoantibodies have neonates with NLE, regardless of whether the mothers are asymptomatic or not<sup>3</sup>. There had been reported cases that clinical features of NLE confirmed histologically but none of the antibodies were found<sup>1,2</sup>. Both of our cases had autoantibodies, case 1 had both anti-Ro and anti-La while case 2 had only anti-La. The precise mechanism of injury to specific tissues, eg skin and heart, is still not clearly understood. The antibodies associated with heart block and with cutaneous disease are believed to be different; antibodies against the Ro/SSA and La/SSB ribonucleoproteins are associated with heart block, whereas antibodies against the La/SSB ribonucleoprotein are associated with cutaneous disease<sup>3</sup>. On the other hand, anti-U1-RNP autoantibodies are usually associated with atypical cutaneous lesions without cardiac or systemic abnormalities in a small number of NLE cases<sup>3</sup>.

The most common clinical manifestations of NLE are dermatological followed by cardiac and liver<sup>3</sup>. Some infants may also have haematological, neurological or splenic abnormalities<sup>3</sup>. Cutaneous lesions most often appear within the first few weeks of life and resolves by 4-6 months old<sup>2</sup>. Typically rashes are erythematous or polycystic plaques with or without fine scales, predominantly on the scalp, neck and face (with periorbital distribution), but similar lesions can appear on trunk and extremities. The lesions can be urticarial, desquamative, ulcerative or crusted<sup>3</sup>. The severity of cutaneous manifestation can vary considerably as illustrated in both the cases here.

Cardiac disease, commonly being third degree heart block is the most common cardiac finding, affecting 15-30% NLE infants<sup>3</sup>. They are associated with high morbidity with 50-70% of them requiring pacemakers. Cardiac manifestations usually dictate the prognosis of NLE. Cutaneous and cardiac manifestations are seen simultaneously in less than 10% of NLE patients<sup>2</sup>. Fortunately, both our babies only had cutaneous manifestation and did not have cardiac complications. Liver involvement with raised liver enzymes may be observed but they are usually fairly asymptomatic and spontaneous resolution is seen as well<sup>3</sup>. The infant illustrated in our second case had mild transient elevated alanine transaminase, but resolved rapidly with no complication. Both cases illustrated above did not have haematological, neurological or splenic abnormalities.

Management of cutaneous lesions includes sun avoidance, sunscreen and low potency corticosteroids to hasten resolution. Spontaneous resolution is the natural course of the cutaneous lesions by 4-6 months of age when the maternal antibodies disappear<sup>2</sup>. Rarely parenteral or enteral corticosteroids are needed as seen in our case 2. However the decision was made in view of her irritability and spiking temperature; and the results of the autoantibodies were not available yet. Child responded to the treatment and when the results were known, the oral prednisolone was tapered off rapidly. The management for cardiac complications is more complex and should ideally be managed by the paediatric cardiologist. Cardiac complications management is not discussed here as they were not experienced by both the illustrated cases above.

### CONCLUSION

Neonatal lupus erythematosus is a rare disease that has a characteristic rash and commonly affects infants of asymptomatic mother. High index of suspicion is required to diagnose through antibodies screening of both infants and mothers. Cardiac complications have to be actively sought for and managed accordingly.

### ACKNOWLEDGEMENT

Special thanks to Dr Lee Bang Rom for the histology photos.

### CONFLICT OF INTEREST DECLARATION

The authors have no conflict of interest to declare.

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**Citation:** Dr. Wee Ai Leen, Dr. Leong Kin Fon, Dr. Sabeera Begum. *Neonatal Lupus Erythematosus: Two Cases Reports in a Single Centre. Archives of Dermatology and Skin Care. 2018; 1(2): 15-19.*

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