

Cutaneous manifestations of neonatal lupus: a case report

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ABSTRACT

Neonatal Lupus Erythematosus (NLE) is a rare disease associated with placental transport of maternal anti-Ro/La and/or anti-U1RNP antibodies into the fetal circulation and characterized by cardiac, cutaneous, hematologic and hepatic manifestations. The most serious complication of NLE is complete heart block and cardiomyopathy. The maternal connective tissue disorder has been systemic lupus erythematosus (SLE) or Sjögren syndrome in most cases, however approximately 50% of mothers are asymptomatic at the time of diagnosis, testing positive only against Ro and/ or U1RNP auto-antibodies. We describe a case of neonatal lupus erythematosus and review the clinical and laboratory manifestations of this rare disease.

Keywords: Anti-Ro antibodies; Anti-La antibodies; Neonatal lupus erythematosus

INTRODUCTION

Neonatal Lupus Erythematosus (NLE) is an uncommon immune-mediated disease associated with placental transport of maternal anti-Ro/La and/or anti-U1RNP antibodies described mainly through isolated case reports and a few published series. These immunoglobulin G (IgG) antibodies cross the placenta and can potentially damage fetal tissue and cause the clinical manifestations of NLE. Cutaneous manifestations, transient hepatitis, thrombocytopenia and anemia can occur but the greatest concern when making the diagnosis of NLE is the risk of congenital heart block.

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CASE REPORT

A five-week-old female baby, born by caesarean section at term without complications, was brought to our clinic for an acute cutaneous eruption that had begun ten days earlier. The clinical presentation was initially a red papule on the chin, which rapidly evolved into several lesions on the scalp, trunk and right arm. The baby had no recent sick contacts, was eating normally and was otherwise doing well. Her mother has suffered from systemic lupus erythematosus (SLE) for three years taking low dose of systemic corticosteroids and hydroxychloroquine 400mg/day and had no exacerbation of the disease during pregnancy. She had made serial dual M-mode echocardiograph during pregnancy because of the presence of anti-SSa/Ro and anti-SSb/La antibodies in serum, which were all normal. Physical examination revealed an interactive baby girl in no acute distress. Her skin examination was notable for the presence of annular erythematosus lesions with raised margins and lighter colored center, on the scalp, chin, right arm and trunk (Figure 1). There was no mucosal involvement. She had no organomegaly and auscultatory findings of her heart and lung examination was normal. A diagnosis of neonatal lupus erythematosus was suspected and appropriate laboratory and heart evaluation was initiated. Mother's serological examination showed an antinuclear antibody titre of more than 1 in 1280; anti SSa/Ro and anti-SSb/La antibodies were also detected. The baby's serum was positive for antinuclear antibody titre of more than 1 in 160, anti SSa/Ro and anti-SSb/La antibodies. Anaemia and enzymatic liver abnormalities were found. The electrocardiogram revealed normal sinus rhythm with normal axis and intervals; an echocardiogram did not identify cardiac mal-

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FIGURE 1. Annular erythematous lesions on the chin, right arm and trunk

formations or cardiomyopathy.

The mother was reassured that the lesions would disappear, by the time when maternal antibodies were absent in the neonatal circulation. A mild topical steroid was prescribed to apply to the lesional skin, and sunscreen as well as sun avoidance were also suggested. The baby's lesion cleared at approximately 4 months of age. At that time, complete blood cell count and liver function studies were all normal. She required no further treatment and continues to do well.

DISCUSSION

Neonatal lupus erythematosus (NLE) is an uncommon autoimmune disorder present in newborn infants of mothers with autoantibodies against Ro, La and, less commonly, U1-ribonucleoprotein (U1-RNP) mainly characterized by the presence of skin lesions and/or cardiac complications. Generally, when patients have skin manifestations, they have no cardiac defects and vice-versa; however, in 10% of cases these manifestations may coexist¹. Other findings may include hematologic, hepatic and neurological abnormalities.

However, only some neonates exposed to these antibodies develop disease and the reasons why some babies develop skin disease, while others develop heart disease are not clearly known². Therefore, other factors (genetic predisposition, viral infection, and other unknown factors)³ exist, that render some infants susceptible in the presence of maternal autoantibodies.

Roughly half of the mothers at the time of childbirth are healthy and do not have signs or symptoms of lupus

erythematosus (LE) or other collagen-vascular disorders^{4,5}; the remainder have some symptoms of LE or Sjögren syndrome or another collagen-vascular disease.

A mother with antiSSa/Ro positive has a 25% risk of subsequent children born with NLE if she had a previous child with NLE⁵. The incidence of NLE is not known and no racial predilection has been observed¹.

Although complete or incomplete congenital heart block (CHB) can be found, it is a rare manifestation but it is the greatest concern when making the diagnosis of NLE. CHB can be diagnosed in-utero and once established it is irreversible⁶. It is associated with high morbidity and mortality, and 50% to 70% of patients require pacemakers because sudden death or heart failure may occur⁷. Amongst the cardiac abnormalities about 50% are conduction defects⁶. The heart block is thought to result from the deposition of anti-SSa/Ro antibody at the atrioventricular node, which leads to fibrosis and calcification⁸. A frequent monitoring of the fetal heart rate and ultrasonography are recommended to screen for heart block in pregnant women with known systemic lupus erythematosus or Sjögren syndrome⁶ as we did in our case. Mothers of neonates with NLE, particularly neonates with CHB, have a 2-fold to 3-fold increased risk of subsequent affected neonates⁶.

Approximately half of all patients have cardiac manifestations and the other half cutaneous manifestations. Cutaneous lesions consist of transient non-scarring erythematosus annular plaques with a predilection for head and neck (photoexposed areas) but may also occur on the arms and trunk as in our case. They are present at birth or during the first two months of life and resolve within four to six months when maternal antibodies disappear⁹. While remnant telangiectasias can rarely occur at previously affected site, dyspigmentation is frequent, but with time, spontaneously resolve¹. Differential diagnoses of skin lesions are made with seborrheic dermatitis, tinea corporis, psoriasis, atopic dermatitis, granuloma annulare, neonatal acne, erythema multiforme, Langerhans cell histiocytosis and congenital infections⁴. A skin biopsy is rarely needed to make the diagnosis of neonatal lupus. Histological findings are hyperkeratosis with follicular plugging overlying an interface dermatitis with basal vacuolar changes. Direct immunofluorescence demonstrates granular IgG deposition at the dermal-epidermal junction and, occasionally, IgM and/or C3 deposition¹⁰. Treatment of cutaneous lesions generally requires sun avoidance, sunscreen and mild topical steroids however spontaneous resolution is the natu-

ral course of cutaneous lesions¹¹. Transient hepatitis, thrombocytopenia and anemia can also occur and thus all patient should have a complete blood count and liver function studies should be performed.

Neonatal lupus erythematosus represents a multi-disciplinary challenge involving rheumatologists, obstetricians, neonatologists, pediatric, cardiologists and dermatologists to identify the pregnancy at risk for NLE development and to introduce appropriate in utero or postnatal therapeutic strategies.

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REFERENCES

1. Thorton CM, Eichenfeld LF, Shinall EA, Siegfrie E, Rabinowitz LG, Esterly NB. Cutaneous telangiectasias in neonatal lupus erythematosus. *J Am Acad Dermatol* 1995; 33:19-25.
2. Awasthy N, Paul P, Mehta R. Neonatal lupus erythematosus. *Indian Pediatr* 2004; 41: 1048-50
3. Buyon JP, Kim MY, Copel JA, Friedman DM. Anti-Ro/SSa antibodies and congenital heart block: necessary but not sufficient. *Arthritis Rheum* 2001; 44: 1723
4. Perez MF, Buján MM, Cervini AB, de Torres ME, Lanoel A, Pierini AM. Neonatal Lupus Erythematosus: a report of four cases. *An Bras Dermatol* 2011; 86; 347-351.
5. Neiman AR, Lee LA, Weston WL, Buyon JP. Cutaneous manifestations of neonatal lupus without heart block: characteristics of mothers and children enrolled in a national registry. *J Pediatr* 2000; 137: 674-680.
6. Tincani A, Rebaioli CB, Taglietti M, Schoenfeld Y. Heart involvement in systemic lupus erythematosus, anti-phospholipid syndrome and neonatal lupus. *Rheumatol* 2006; 45: iv8-iv13.
7. Buyon JP. Neonatal lupus: bedside to bench and back. *Scand J Rheumatol* 1996; 25: 271-276.
8. Boh EE. Neonatal lupus erythematosus. *Clin Dermatol* 2004; 22: 125-128.
9. Tincani A, Nuzzo M, Motta M, Zatti S, Lojaco A, Faden D. Autoimmunity and pregnancy : autoantibodies and pregnancy in rheumatic disease. *Ann N Y Acad Sci* 2006; 1069: 346-352.
10. Robles DT, Jaramillo L, Hornung RL. Neonatal lupus. *Dermatol Online J* 2006; 12: 25.
11. McKinlay JR, Cooke LM, Cunningham BB, Gibbs NF. Neonatal lupus erythematosus. *J Am Board Fam Pract* 2001; 14: 68-70.

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