CASE REPORT Prof-802

NEONATAL LUPUS

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ABSTRACT... buttsahib100@yahoo.com. The case presented here is of a newborn having cutaneous lesion which were noticed soon after birth. Cutaneous lesion included well demarcated macular erythematous rash over face. The cutaneous lesion was associated with positive anti Ro Ab in the newborn as well as in the mother. There was no evidence of hepatic, cardiac and hematological involvement in this newborn.

Key Word: Neonatal lupus Erythematosus (NLE)

INTRODUCTION

Neonatal lupus erythematosus (NLE) is a rare disorder caused by the transplacental passage of maternal autoantibodies. Only 1% of infants with positive maternal autoantibodies develop NLE. The most common clinical manifestations are cardiac, dermatologic and hepatic. Some infants may also have hematologic abnormalities. Most mothers at the time of child birth are healthy and without signs or symptoms of lupus erythematosus or other collagen vascular disorders. Mothers of children with NLE may later develop an atypical rather than classical picture of systemic lupus erythematosus or other connective tissue disorder. If a mother with anti Ro autoantibodies has 1 child with NLE, the incidence in subsequent pregnancies is approximately 25% . The incidence of congenital heart block is 15-30% in infants with NLE.

CASE REPORT

A case of one day old female newborn is presented who was born by LSCS. She presented with rash over her face noticed at birth. Baby was otherwise healthy, full term and taking feed normally. There was no jaundice, bleeding tendencies, cyanosis or respiratory distress.

On examination, her weight was 2.5 kg, length 49cm and head circumference was 32.5cm. She was having an erythematous, macular rash with well defined

margins distributed over her face especially on the forehead, peri-orbital region and bridge of the nose. Rest of the systemic and regional examination was normal.

Mother had bad obstetric history with previous two first trimester abortions and one IUD but without any evidence of well defined autoimmune disorder. Considering this and peculiar rash, provisional diagnosis of neonatal lupus was made and relevant investigations were sent. Results of which showed that test for anti Ro (SSA) Ab was positive (8.0 unit/ml normal range is 0-2 units/ml) whereas test for anti La (SSB) Ab was negative.

Other investigation revealed Hb 18.1 g/dl, hematocrit 55.1%, total leucocytes count 16.2 x10 E 9/L, and differential leucocytes count showing neutrophils 35.4%, lymphocytes 56.7%, monocytes 5.3% and basophils 0.7% . Platelet count was 1, 50,000/mm^3.Serum ALT was 20 IU/L ECG was normal.

Maternal serum was positive for anti Ro Ab both qualitatively & quantitative (17 units/ml) whereas tests for anti La Ab, anti nuclear Ab, antidouble stranded DNA, Anti Sm Ab, Anti Scl-70 and Sm/RNP were all negative.

On the basis of positive maternal anti Ro Ab and positive neonatal Anti Ro Ab, and peculiar cutaneous manifestation, diagnosis of neonatal lupus was confirmed.

DISCUSSION

NLE is an uncommon disease. It occurs in 1 in 20,000 live births[1]. NLE has been reported more in females. With the heart disease, female to male ratio is 2:1 and with skin disease ratio is 3:1[2]. The age of onset is from birth to 6 months of age[3,4,5].

The mother produces IgG auto-antibodies against Ro (SSA), La (SSB) and / or U 1-ribonucleoprotein and they are passively transported across the placenta. The presence of maternal anti-SSA/Ro and SSB/La antibodies increases the risk of bearing infants with NLE. These autoantibodies can be found alone or in combination, however anti Ro Ab is present in almost 95% of patients[6] .

Mother of patients with NLE may have differentiated or undifferentiated autoimmune disorder such as SLE, Sjogren Syndrome, Undifferentiated autoimmune syndrome or Rheumatoid arthritis[7].

The skin manifestations of NLE occur in the first month or later in life and are mainly due to the presence of anti La/SSB antibodies[8].

The cutaneous findings are transient. Two third of patients with the skin finding have them at birth, with the remainder developing them within first 2-6 month of life[9].

Cutaneous finding include well demarcated erythematous, mild, scaling plaque that is often annular and appears predominately on the scalps, neck or face. This plaque is typically periorbital in distribution. Similar plaques may appear on the trunk or extremities[10,11].

Healing tends to occur within a year, with mild cutaneous atrophy with or without associated telangiectasia[12].

Mother with primary sjogren syndrome or undifferentiated autoimmune syndrome have a greater risk of delivering an infant with congenital heart block than those with SLE[7] . NLE have 15-30% incidence of congenital complete heart block developing between 18th and 20th week of gestational age[6,13,14,15].

Mother with autoimmune disease and anti Ro Antibodies are at risk of developing a child with NLE but at low risk of developing congenital complete heart block[16].

Other disturbances such as sinus bradycardia, prolongation of QT interval and irregular heart beat may also be present. In some cases myocarditis and

pericarditis can develop. Hematological abnormalities like hemolytic anemia, thromobocytopenia and neutropenia may occur in the first 2 weeks of life and disappear by the end of 2nd month[17].

Hepatic manifestations include hepatomegaly with elevated transaminase level[18].

Laboratory studies include checking neonatal serum for anti-Ro, anti- La and anti, u1-RNP antibodies. Complete blood counts with platelet count may reveal pancytopenia or isolated anemia, neutropenia or thrombocytopenia. Liver function tests and ECG should be done in all cases.

Maternal serum should also be screened for antinuclear, antidouble stranded DNA, anti Ro, anti La and anti U1-RNP antibodies. Skin biopsy reveals interface dermatitis with moderate hyperkeratosis, follicular plugging and vascular degeneration in the basal cell layer.

Treatment of skin lesions include mild steroids and possibly laser treatment for residual telangiectasia. Photoprotection is desirable because skin lesions may be precipitated by sun exposure. Most patients with NLE of the skin, liver or blood have transient disease that resolves spontaneously in 4-6 months. Only rare reports exist of these children developing systemic lupus later in life. The presence of cardiac abnormality is associated with congestive cardiac failure. Neonatal mortality rate in the presence of heart disease is 20-30%[19].

The mother of a child with NLE should be aware that the rate of recurrence in future pregnancies is about 25% and subsequent pregnancies should be closely monitored[20].

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