NEONATAL LUPUS ERYTHEMATOSUS: A CASE REPORT

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ABSTRACT

Neonatal lupus erythematosus is a rare autoimmune disorder mainly affecting the skin and heart. The most serious complication of cardiac manifestrations which may cause life - threatening of an infant is congenital heart block. In addition, hematological, hepatobiliary and neurological involvement may occur. We report a 2-day-old infant was diagnosed as neonatal lupus erythematosus via clinical and laboratory findings. She presented with cardiac, hematological, hepatobiliary and neurological manifestrations. Her mother had treated a lupus eryhtematosus systematic syndrome up to pregnancy. Neonatal lupus eryhtematosus should be considered in infants presenting with clinical suspicion and mother suffering from an autoimmune disease.

Keywords: *Neonatal lupus erythematosus, congenital heart block, hematological, hepatobiliary.*

1. INTRODUCTION

Neonatal lupus erythematosus (NLE) is a rare acquired autoimmune disease that is appeared at birth. The disease is caused by maternal autoantibodies (anti-SSA/Ro and/or anti-SSB/La) transmitting across the placenta and affecting the developing fetus. Two affected major organs are heart and skin. The most serious complication of neonatal lupus is a heart condition known as congenital heart block which can potentially be life-threatening. Only about 1-5% of infants born from mothers with that antibodies can get NLE with heart symptoms [1]. Up to 66% of cases with complete and permanent heart block requires a pacemarker [2]. Other heart condition can be cardiomyopathy, Heart symptoms can be detected during gestation or after birth. Skin symptoms can be developed in the first few weeks of birth. That symptoms are usually transient and dissappeared during the next several months. The disease can also impact the hepatobiliary or hematological or neurologic system [3]. Although most of these findings are transient, cardiac abnomalities are potentially high risk causing morbidity and mortality [3,4]. Therefore, patients with NLS should be monitored and followed-up. We report a case of NLE with cardiac, hematological and hepatobiliary findings, hospitalized in the Department of Pediatrics at E hospital.

2. CASE REPORT

A ten-hour-old female infant was transfered from the Obstetric and Gyneconogical department to our department for signs of respiratory distress such as cyanosis. She was born at 38 weeks' gestation by surgery. Her birth weigh was 2600 gram. The mother was 24 years

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old. She has been diagnosed as Lupus erythrema system for 3 years and treated by Medrol 4mg per day during pregnancy. She has not been continuously followed up during pregnancy and did not find any fetal abnormalities.

Admission examination: she was elert, cyanosis, shortness and grunting sounds while breathing, chest retractions, and nasal flaring. Tachypnea with respiratory rate of 67 breaths per minute. Temperature was 36.6oC, and SpO2 was 90-91%. Heart rate was about 100-110 beats per minute and no heart murmur was auscultated. The chest radiograph showed the reduction of lung volume and mild lack of air bronchograms. Cardiologic examination with electrocardiography showed a sinus bradycardia (figure 1), normal T wave. Echocardiography showed a very small patent ductus arteriosus and did not show any other conduction system abnormality or cardiomyopathy. Cranial and abdominal ultrasonography were normal.

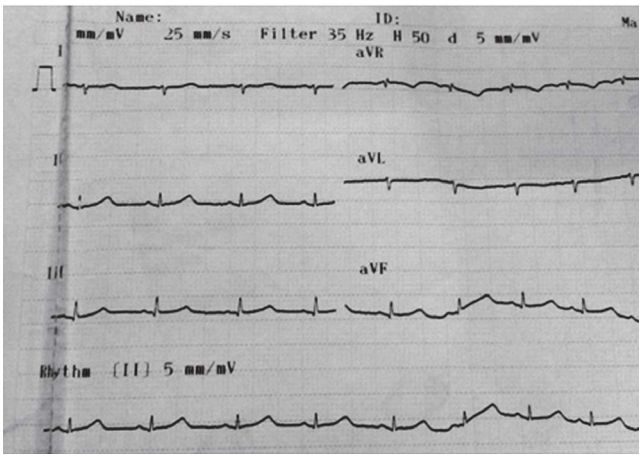

Figure 1. Echocardiogramph at the first day of hospitalization

Laboratory tests showed trombocytopenia (75G/L), normal hematocrit and white blood cells, slight direct hyperbilirubinemia (31.7 U/L), elevated aspartate aminotransferase levels (165.5 U/L) and increased GGT of 298.8 U/L. Blood gases at admission time revealed a mild respiratory acidosis and hypoxemia as follow: pH, 7.2; partial pressure of carbon dioxide (PCO_2), 57; partial pressure of oxygen (PO_2), 78; bicarbonate (HCO_3^-), 23.6, BE, -4.3; oxygen saturation, 89% with FiO_2 of 1.

Further laboratory tests showed positive anti-Ro/SSA in the patient, anti LA (-) at the 5st day of birth.

A diagnosis of neonatal lupus erythematosus was made based on the clinical and laboratory findings.

She required supplemental oxygen via CPAP (5cm PEEP) in 12 hours a fraction of inspired oxygen (FiO2) of 0.4 and then via the nasal cannula of FiO2 gradually decreased to 0.21 for 24 hours. Her respiratory status was stable with respiratory rate was 35 breaths per minute. She then went to room air. Other management was supportive and other newborn cares. She recovered and has been sent back home after 10 days. She was monthly followed up by pediatricians and cardiologists. Laboratory findings at birth included elevated transaminase levels, anemia and trombocytopenia but gradually became normal up to 3 months of birth (figure 2).

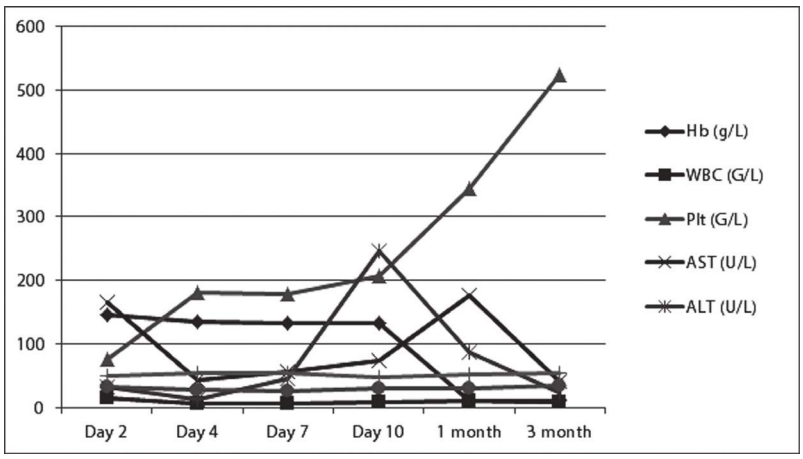

Figure 2. Laboratory tests during hospitalization and follow-up time.

Hb: Hemoglobin; WBC: white blood cells; Plt: platelet; AST: aspartate aminotransferase, ALT: alanine aminotransferase.

3. DISCUSSION

The incidence of the NLE is estimated as 1 in 12,500 to 20,000 live births [5]. It is reported that female and premature infants have lightly higher risk than others. Mothers have Ro or La antibodies suffering from systemic lupus erythematosus or rheumatoid arthritis or Sjogren's syndrome or connective tissue diseases have risk transmitting antibodies to fetus. The diagnosis of NLE is made based on clinical features and autoantibodies in maternal or infant serum [6].

From 1980s, the first report about the link of Ro and/or La antibodies and congenital heart block in infants was published. Moreover, a recent research showed that all cardiac complications in neonates involve in Ro antibody level and are independent to La antibody level. That antibody levels in mother's circulation are directly associated with NLE but not be antibody present. Therefore, it is recommended that echocardiography of fetus should be done in all mothers with high levels of Ro antibodies [6]. Only about 2% infants of mothers with systemic lupus erythematosus may have NLE. It is reported that genetic and environmental factors also play a role to NLE pathogenesis. The clinical onsets of NLE often present at 2 - 4 weeks after birth[7]. In my case, the mother had had systemic lupus erythematosus and had been treated with steroid for four years. That implies a risk of NLE for her baby. Therefore, her delivery had been controlled and the infant had been examined and monitored from birth.

In literature, an infant with NLE may have signs and symptoms of cutaneous, cardiac, hematological, hepatobiliary systems [4,8]. The major finding is red rashes on skin of face, head, body and limbs. This sign is presented in about 71% of NLE cases [5]. Rashes are transient, remain in several first weeks of birth and then disappeared in some months. About 65% of NLE cases have cardiac symptoms including congenital heart block and cardiomyopathy [5].

Congenital heart block is persistent and may cause serious complications, even mortal. A research on the 214 cases of cardiac neonatal lupus syndrome (with congenital heart block) which were detected in utero and neonatal period and followed up showed that 79.1% of alive born children required a pacemarker, 11.8% of them died and 18.8% of them had had dilated cardiomyopathy [9]. In a review of 123 NLE cases in China from 1990 - 2014, Li et al have reported 119/123 NLE cases with cutaneous manifestations.

Cardiac manifestations presented in 15 patients including congenital heart block, patent foramen ovale, flat T-waves, artrial septal defect, left atrial enlargement, and sinus bradycardia. The sinus bradycardia was presented in one NLE patient (1/123) [7]. Our patient had bradycardia with heart rate of about 100 beats per minute, and showed sinus bradycardia in echocardiogram.

Congenital heart block may occur in utero and may be first detected in second trimester of pregnancy. Our patient had not been gradually re-examination during pregnancy and had not found any abnormalities of the fetus. Fluorinated steroids were considered as a factor decreasing regression and increasing survival of cardiac NLE. However, some other research pointed that it had not made [9]. However, ultrasound was reconmended for mothers with autoimmune antibodies to early detected cardiac abnomalities of fetus.

It is reported that a half of NLE cases has hematological and hepatobiliary findings [5,6]. Hepatobiliary manifestations may be direct hyperbilirubinemia, elevated transaminase levels, cholestatic hepatitis and fulminant liver failure [5]. Additionally, hematological abnormalities include trombocytopenia, leukopenia and hemolytic anemia [5,9-11]. Our patient also had elevated transaminase levels, both direct and indirect hyperbilirubinemia, anemia and trombocytopenia. The index gradually had become stable, except for mild anemia keeps going until 3 months of life.

The neurological symptoms are rarely with about 35% of NLE infants [5] and are nearly asymptomatic. Neurological manifestations may be macrocephaly which may also develop hydrocephalus, or leukoencephalopathy [12]. Although that is a rare manifestation, our patient has a tend to enlarge head circumstance (> +2SD) compared to reference of head circumstance of children by age and sex (WHO) from the second month of life (43cm at 2 months and 45 cm at 3 months). Cranial ultrasonography was screened and we have not found any abnormalities. Also, delay motor development has recently been recorded and need to follow-up.

In conclusion, it is recommended that an infant should be considered as an NLE if his/her mother suffered from autoimmune diseases, particularly carrying autoimmune antibodies during pregnancy and clinical suspicion. The diagnosis of NLE requires clinical suspicion and should be warned about a congenital heart block and typical cutaneous lesions. Long-term monitoring of these infants is also important to control life-threatening complications.

Competing interests

The authors declare that they have no competing interests.

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