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LETTER TO THE EDITOR

A neonatal lupus syndrome with pericardial tamponade due to maternal Sjögren's syndrome

Maternal Sjögren sendromuna bağlı perikardiyal tamponadı olan neonatal lupus sendromu

Hacer Yapıcıoğlu[1], Mustafa Özdemir[1], Tugay Tepe[1], Tolga Akbaş[2], Ferda Özlü[1], Sevcan Erdem[2]

[1]Cukurova University, Faculty of Medicine, Department of Pediatrics, Division of Neonatology, [2]Pediatric Cardiology

To the Editor

Sjögren's syndrome (SS) is a systemic autoimmune disease of unknown etiology characterized by dry eyes and dry mouth[1]. Neonatal lupus syndrome (NLS) is an acquired congenital autoimmune disorder which mostly occurs in fetuses and newborns of mothers with various rheumatic diseasessuch as systemic lupus erythematosus (SLE), SS and rheumatoid arthritis. NLS is a rare disease that develops due to the passage of anti-Ro/SSA and anti-La/SSB antibodies in 12-16 weeks of pregnancy[2]. Manifestations of NLS are congenital heart block, skin lesions, hepatitis, thrombocytopenia, neutropenia, anemia, pneumonia, pulmonary hemorrhage, hypoglycemia and convulsions. The most common manifestations are skin lesions and congenital heart block (CHB)[3].

In this case, we reported a male newborn with NLS who presented with trombocytopenia, convulsion and pericardial tamponade and discussed the management of NLS.

The patient was admitted to NICU for prematurity and respiratory distress. He was the third child of nonconsanguinous parents. Mother was 39 years old. She had preeclampsia and oligohydramnios and an urgent cesarean section was performed at 34th gestational weeks. Ten years ago, she was diagnosed as Sjögren's syndrome while investigating the etiology of maternal neutropenia. As she had also glucose 6 phosphate dehydrogenase (G6PDH) deficiency, hydroxychloroquine (HCQ) was not started. In physical examination, the newborn baby had cutis marmoratus, respiratory distress and acrocyanosis. His laboratory values were normal except for thrombocytopenia (platelet:14.000/ μL). Platelet suspension was transfused once and control platelet counts were within normal limits in the follow-up. Electrocardiography (ECG) showed sinus rhythm. He had myoclonic convulsion on the second day of life and phenobarbital was started and then phenytoin, levetiracetam and pyridoxine were added due to recurrent convulsions. Cranial ultrasound and cerebrospinal fluid (CSF) was normal. Blood ketone, blood ammonia and lactate/pyruvate ratio were normal. Anti-Ro/SSA:305 U/mL (>22: positive), anti-La SSB: <5 U/mL (<15 negative), ANA:137 (>60 highly positive), Anti-ds DNA:6,68 IU/mL(<92,7 negative), AMA:316 U/mL(<20 negative). He had no convulsions on follow-up, however he had sudden cardiac arrest on 9th day of life and two minutes of cardiopulmonary resuscitation (CPR) was performed. He had sinus rhythm before arrest. Echocardiography revealed minimal pericardial effusion. 24-hour holter monitoring was normal. Afterwards his condition was normal. Cardiac arrest developed again on the 19th day of his hospitalization. He was immediately intubated and CPR was started. Although he had several times of adrenalin and 30 minutes of CPR, no cardiac rhythm was revealed. Only after

Address for Correspondence: Mustafa Ozdemir, Çukurova University, Faculty of Medicine, Department of Pediatrics, Division of Neonatology, Adana/Turkey E-mail: drmustafa2013@hotmail.com
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echocardiography showed a massive pericardial effusion (Figure 1), subxiphoid pericardiocentesis was performed and approximately 60 ml serous fluid was aspirated. The rhythm on the monitor returned to normal sinus rhythm and pulse was detected and CPR was terminated. 2 mg/kg/day prednisolone treatment was started. Control echocardiography showed minimal pericardial effusion adjacent to the right atrium and right ventricle (Figure 2). Pericardial effusion was transudate. Blood tests showed: ANA:106 (positive), anti-Ro/SSA:320 U/mL and anti-La/SSB:5 U/mL. aEEG showed low voltage pattern after cardiac arrest. As he had elevated levels of antibodies, breast milk antibodies were evaluated for antibodies: anti-Ro/ SSA:5 U/mL and anti-La/ SSB:5 U/mL. He had no pericardial effusion in control echocardiograms. He was extubated on 24th day of life. Cranial magnetic resonance imaging was normal. EEG showed moderate baseline rhythm irregularity. All cultures (blood, urine and CSF) were negative. He was discharged on 32nd day of life with 1 mg/kg/day prednisolone which was stopped after two weeks. Control EEG showed normal pattern and anticonvulsive drugs were stopped when he was two months old age. He is now 24 months of age, healthy and neurological development is compatible with his peers.

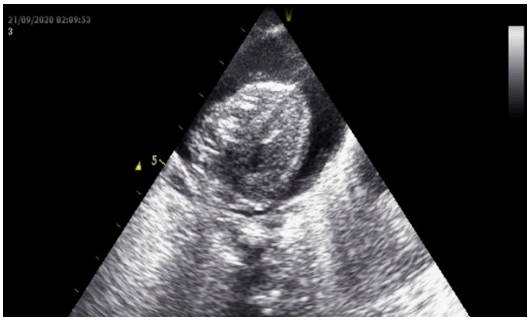

Figure 1. Massive pericardial effusion.

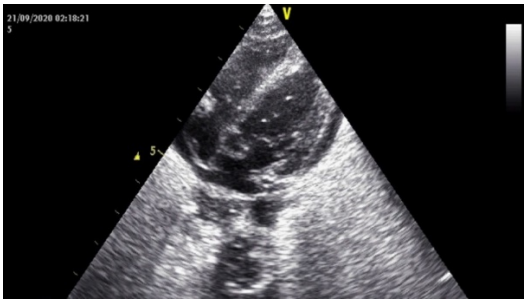

Figure 2. Minimal pericardial effusion after subxiphoid pericardiocentesis

Neonatal lupus syndrome is an autoimmune disease gained during fetal life as a result of intrauterine passage of maternal antibodies. Only 1-2% of women with anti-SSA/Ro or Anti-SSB/La antibodies give birth to a newborn with NLS[2,4]. Some of the clinical situations such as skin lesions and hematological abnormalities are reversible, however CHB is a permanent symptom. Thrombocytopenia is present in 10% of newborns with NLS with or without other cytopenia. Typically, it worsens in the first days after birth and then begins to improve[5]. We have detected thrombocytopenia in the present case which was supplemented with a platelet suspension. Afterwards, platelets increased gradually and he did not need another transfusion.

Anti-SSA/Ro and Anti-SSB/ La antinuclear antibodies have been detected in breast milk of mothers with positive serum anti-SSA/ Ro and anti-SSA/ La antibodies. In the present case, blood anti-SSA/Ro antinuclear antibodies were increased from 305 to 320 U/mL. Since there was no significant decrease in autoantibody titers in our case, we have measured antibody titers in breastmilk for transport via breastmilk and result was negative.

Although NLS may develop in children born to anti-SSA/Ro or anti SSB/La positive women with systemic diseases; mothers of affected children by NLS may have an active autoimmune disease or 25-60% may be asymptomatic[2]. As mothers may be asymptomatic, maternal autoimmune diseases should be in mind if her infant has NLS clinic. Early identification of a CHB in fetus may prevent potential complications such as miyocarditis, pericardial effusion and fetal heart disfailure[6]. Hydroxychloroquine is one of the standard medications in SLE and Sjögren's syndrome, it is safe and compatible with pregnancy and breastfeeding[7]. The recent studies showed HCQ during pregnancy may decrease the risk of CHB in fetus. However she had no HCQ treatment as she had G6PDH deficiency.

90-95% of heart blocks seen in the intrauterine or neonatal period are due to NLS. The most common heart block in NLS is irreversible 3rd degree (complete atrioventricular) block. Postnatally, the baby may develop congestive heart failure, hepatomegaly, and metabolic acidosis requiring a pacemaker. Low heart rate may lead fetal hydrops.

Pacemaker implantation in newborns at high risk for CHB potentially reduces the negative consequences of severe bradycardia and asystole in the immediate postnatal period[8]. According to a study conducted by Glatz et al, maternal steroid use, close follow-up, and early placement of a temporary epicardial pacemaker are recommended in newborns with congenital CHB[9]. We have not detected any heart block in the present case, however he had cardiac arrest once and we could not find the etiology; then a pericardial effusion was revealed which caused cardiac tamponade later. To our knowledge pericardial effusion in NLS were reported before [9-13], however in these cases, the reasons were hydrops fetalis secondary to heart block[10], dilated cardiomyopathy[11] or congestive heart failure[9,12,13]. Our patient had none of them and this case is the first report in the literature. Although pericardial effusion was minimal, a cardiac tamponade occured in a week. So, any pericardial effusion should be followed closely.

In conclusion, NLS is an autoimmune disease and should be in mind in children born to women with Sjögren's syndrome. As half of the mothers are asymptomatic, any infant with multisystemic involvement such as congenital heart block, thrombocytopenia, pericardial effusion or convulsion should be evaluated for NLS.

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