

Neonatal lupus erythematosus: nine cases report

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Abstract: Objectives: To investigate the clinical characteristics of neonatal lupus erythematosus (NLE), which could contribute to study of the pathogenesis mechanism and treatment of NLE. **Methods:** Nine case of NLE were brought into this study in our hospital from August 2005 to June 2011, and the clinical manifestation, auxiliary examination and follow-up status were analyzed. **Results:** One patient died in nine case, two patients had sequelae of nervous system, and other six cured; the children who had mild disease and full-term gestational age was due to the autoimmune disease of the mothers was monitored and controlled stably during pregnancy, while the children whose mother didn't find their autoimmune disease or controlled poorly had low body weight, small gestational age and severe illness; from the clinical manifestation we could see that six patients presented skin rash, five case had peripheral hemogram changes, three patients presented with abnormal electrocardiogram (ECG) caused by non-three degree atrioventricular block and recovered gradually, two patients presented with neurological signs and image changes; two case was anti-U1RNP antibody positive, the remaining seven case was anti-SSA/SSB antibody positive. **Conclusion:** Mother performing autoimmune disease screening and controlling disease actively during pregnancy were beneficial to children; the neonatal ECG changed by non-atrioventricular block could recovery and improve gradually; NLE may leave sequelae of nervous system and need regular inspection.

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Key words: Neonatal; Lupus erythematosus; Clinical characteristics.

1. Introduction

Neonatal lupus erythematosus (NLE) is a multisystem lesion syndrome caused by autoimmune antibodies of the mother entering newborn's body through the placenta, it is a rare disease.(Inzinger et al.,2012)Its main clinical manifestations were skin rash, heart block, blood, liver and other organ involvement. In addition to heart block which is irreversible change, the other clinical manifestation can be spontaneous regression, so the majority of scholars believe NLE is self-limited disease. Nine NLE were included in this study from 2005 to 2011, the clinical analysis will help the further study of the disease.

2.Methods

2.1 Clinical data

2.1.1 Objectives

Nine NLE were included in this study from August 2005 to June 2011, case one and case seven were siblings. Other general data was described in Table 1.

2.1.2 Diagnosis

The mother who had a history of rheumatic diseases presented with related rheumatic antibody positive; newborn presented clinical manifestations, such as skin rash, thrombocytopenia, hepatosplenomegaly and dysfunction, arrhythmia et

al; the children had positive related immune antibody which was consistent with the mother; except these, infectious diseases caused related clinical manifestation. Meanwhile, indirect immunofluorescence was used in detection of antinuclear antibody(ANA), ELISpot test was performed in detection of extractablenuclearantigen (ENA).

3.Results

As shown in Table 1, there were nine neonatus, rate of males and females was 1:2,average gestational weeks were 36.7±0.37 weeks. the average weight was 2481g. There were four mothers with systematic lupus erythematosus(SLE),and one mother was suffered sicca syndrome. Before becoming pregnant,the four mothers had not been treated for SLE, two mothers received a good treatment,two mothers were monitored regularly, treatment result is not good in one mother. For neonatus, six cases made a prompt recovery after treatment,complications were found in two neonatus, only one case dead. For clinical characteristics in here, the NLE onset time in seven neonatus were found less than one week after birth, rashes were detected in six cases, moreover, four cases represented anemia and plaque decrease characters,respectively. Arrhythmia was found in three cases, there were two cases appeared hepatosplenomegaly or neurological disorder.

Table 1. Generally data

case	gestational weeks(weeks)	Birthweight(g)	sex	outcome	Condition of mothers	preconceptional condition before	treatment
1	27 ⁺⁴	1550	F	dead	SLE	Untreatment	
2	39	2450	M	recovery	—	monitor	
3	37 ⁺¹	2230	M	recovery	SLE	untreatment	
4	38 ⁺⁵	2980	F	recovery	—	monitor	
5	37 ⁺⁴	2850	F	recovery	—	untreatment	
6	31 ⁺⁵	1720	F	complication	SLE	treatment ineffective	
7	36 ⁺¹	2760	F	recovery	SLE	better treatment	
8	35 ⁺³	2650	F	recovery	SS	better treatment	
9	40	3140	M	complication	—	untreatment	

F: female; M: male; SLE: systematic lupus erythematis; SS: sicca syndrome

For immunologic test results, anti-SSB positive result was found in two mothers, among one case appeared anti-sm antibody character. Moreover, two mothers represented anti-U1RNP positive character. Other five cases appeared anti-SSA positive result, among three cases represented simultaneously anti-nuclear antibodies positive result, the antibody drop degrees were 1:1000, 1:320 and 1:1000 in three mothers, however, these were 1:640, 1:160 and 1:320 in three neonatus, respectively. Moreover, there one case appeared simultaneously anti-SSA and anti-JO-1 positive results. (Table 2)

Table 2. Clinical characters

case	Onset(after birth)	rash	anemia	Platelets decrease	arrhythmia	hepatosplenomegaly	nervous system
1	5 days		+	+		+	
2	1 week	+			+: I class Atrioventricular block		
3	3 days			+			
4	5 days	+					
5	3 weeks	+					
6	2 days		+		+: sinus bradycardia, abnormal T wave		+
7	2 days	+					
8	2 weeks	+		+	+: abnormal Q wave		
9	1 days	+	+	+		+	+

3.1 Follow up

Eight cases underwent return visit until one year after birth, case nine had referral until 1.5 years after birth. All cases reexamined immune antibody six months after birth and the antibodies were negative. Three patients presented with arrhythmia, but it had recovered three months after birth according to the return visit, the complete atrioventricular block didn't appear. The skull MRI of case six presented with cerebellum high density shadow, skull CT of case nine showed dysplasia of corpus callosum, in case four, bilateral lateral ventricle, frontal, top, temporal lobe white matter tissues presented abnormal density decreased shadow. Both case six and case nine had neonatal sequelae of nervous system, the NBNA score was low (28 points), their motor retardation, the head could erect steadily until 4 month old, they can't sit, climb and turn over when they were 10 month old, no muscle tension increase, no ankle clonus, one-year-old children's head can erect, but they could sit and hold up stand unstable and can't climb, the muscle tension was weak. The disease didn't improve until the end of return visit. (Table 3)

Table 3. Accessory examination

case	Antibody in mother	antibody titer of mother	antibody titer of neonatus
1	Anti-SSA, anti-nuclear antibody	1: 1000	1: 640
2	Anti-SSB, anti-sm antibody		
3	Anti-U1RNP		
4	Anti-SSA		
5	Anti-SSB		
6	Anti-SSA, anti-Jo-1		
7	Anti-SSA, anti-nuclear antibody	1: 320	1: 160
8	Anti-U1RNP		
9	Anti-SSA, anti-nuclear antibody	1: 1000	1: 320

4. Discussion

Neonatal lupus erythematosus (NLE) is a quite a rare disease and more often appeared in girls, which is most common on the first month after birth. However, the mechanism is unclear. Skin lesions is characterized by the erythema, annular erythema, Discoid Lupus Erythematosus(DLE) changing or slight epidermal thinning except keratotic plug or scar formation (Sawant et al.,2007). The most common infection site was around head,neck and eye, however, the other body parts can be involved in this disease.

The pathogenesis of NLE is unclear. That anti-SSA and -SSB antibody from mother adopt to neonatus by placenta play important role in pathogenesis of NLE. It found that 1% of neonatus who had a NLE if the mothers appeared anti-SSA and -SSB-positive. Congenital heart block(CHB) is the most character in neonatus with NLE, whose mothers have risk of anti-SSA/Ro or -SSB/La more than 85% (Buyon et al.,2003). However, the neonatus from mother with anti-SSA-positive have risk of 0.9%-2% suffering CHB, further study shown that these ratio is 3.1% if the mothers appeared the anti-SSB-positive(Gordon et al.,2004).

The ratio of neonatus with NLE appearing anti-SSA-positive is 91.5% in China, about 54.3% cases with NLE appear anti-SSB-positive. In addition, only a small number cases appear anti-ds-DNA, anti-U1-RNP and anti-RNP-positive. It is similar in antibody between mothers and neonatus. It has reported that SLE, sicca syndrome(SS) and other rheumatism occupy 40%,15% and 10% in mothers of cases, respectively. Moreover, there were no any clinical character in 30%-50% mothers in the pre-natal, however, the anti-SSA or -SSB antibody are presented in blood(Ames and Toss 1995). So, it is necessary that the self antibody should be detected frequently.

Skin lesion is the most common clinical appearance. It is reported that skin lesion occupied 50% in patients with NLE in China, which is higher than that in abroad. About 50% patients with NLE suffer from skin lesions at birth or during the first six months of life. It has report that there are strong relation between skin lesions and anti-SSB antibody.

Cardiac damage is prominent clinical appearance, which is the most dead cause for patients with NLE. The cardiac damage include CHB, myocarditis, cardiomyopathy and congestive heart

failure. It is reported that CHB occupied 30%-40% in abroad (Wallace et al.,1997), the ratio of heart disease in NLE patients is 11%, however, in 2005, the ratio increased to 50% in Japan(Sachiko 2005). In here, the ratio is similar to other research results in China. So, the results show that race difference is play important in NLE onset, further researches are need by gene polymorphism. For mothers appearance anti-SSA and -SSB positive, it is suggest that mother during 16 to 32 weeks of gestation period should be detected by echocardiographic and physical examination, so that the doctors can find CHB early.

In conclusion, NLE is a rare disease, which make the neonatus suffer from life risk, and whether there were some complications is unclear. So, further study is need.

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