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NEONATAL LUPUS WITH MACROPHAGE ACTIVATION SYNDROMES PRESENTED WITH RASH AND FEVER ON CAMBODIAN PATIENTS

Rheumatology

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ABSTRACT

Neonatal lupus can occur in infants born to mother with disorders through trans-placental auto – antibodies. Clinical manifestation in neonatal lupus include cutaneous lesion, kidney, hematological or hepatobiliary finding resembling those seen in systemic lupus erythematous. In autoimmune state, macrophage activation syndromes (MAS) represent a critical and potentially fatal complication that can result in mortality if not immediately identified and managed with the appropriate care. Here we preset a 29-day-old-girl diagnosed with neonatal lupus and serious MAS. She was delivered by a primipara mother who did not exhibit any autoimmune symptoms. The patient visited the hospital due to fever and pancytopenia. Laboratory data were compatible with MAS, including pancytopenia, high level of ferritin, soluble interleukin-2 and decreased natural killer cell activity. In addition, autoimmune study showed positive results for ANA, anti-SSA and anti-SSB. The patient recovered after she received high dose steroid and supportive care. Our case indicates that neonatal lupus should be take into consideration when fever, erythematous skin rash and pancytopenia are observed in infants even if their mothers have no prior history of autoimmune condition.

KEYWORDS

Neonatal Lupus, Macrophage Activation syndrome, Anti – Nucleus Antibodies

INTRODUCTION

Clinical manifestation in neonatal lupus include skin rash, hematologic or hepatobiliary events, and occasionally serious congenital heart block [1]. These clinical feature can occur due to trans-placental autoantibodies from mother with autoimmune diseases including systemic lupus erythematous [2]. Neonatal lupus is developed through passively acquired autoimmune diseases. Clinical findings except heart block in neonatal lupus usually disappear at 6months of age [3].

Macrophage activation syndrome (MAS) is a serious inflammatory condition caused by overproduction of cytokines from immune cells including macrophage due to infectious or autoimmune disease. It is fatal unless there is an appropriate treatment after prompt diagnosis [4]. MAS is not common in neonatal lupus. Most cases are related to mothers with pervious autoimmune manifestation [5].

Here, we report serious case of MAS in neonatal lupus, which initially manifestation with fever and skin rash. The baby was born to a mother without any autoantibodies. This data was approved from Rheumatic Diseases Unit, AIR Plus Polyclinic Board in Cambodia.

CASE PRESENTATION:

A 29-day-old girl was admitted due to fever with pancytopenia. She presented erythema marginatum like skin rash on the whole body (Figure 1). Her birth weight was 3.23kg and her gestational age was 38 + 1 weeks from a primipara mother without any autoimmune disorders at a local clinic. She was hospitalized at another hospital at post-natal 5 days due to skin rash and subsequently diagnosed with neonatal lupus, because her mother's autoimmune study showed positive results for ANA, anti-Ro/SSA and anti-La/SSB.

Figure 1: crumb

On admission, the complete blood cell counts (CBC) revealed a hemoglobin level of 5.8g/dL, a white blood cell count of 1.100/uL (absolute neutrophil count of 110/uL), and platelet count of 72,000/uL. In term of inflammatory biomarkers, the C – reactive protein level was elevated at 8.87mg/dL (normal range <0.5mg/dL), procalcitonin was measured at 1.88ng/mL, and ferritin was significant elevated at 1603 ng/mL. the disseminated intravascular coagulation (DIC) profile indicated severe abnormalities, including a prothrombin time INR exceeding 6, an aPTT over 120secounds, antithrombin III levels at 63.9% and elevated fibrinogen elevated fibrinogen level of 410mg/dL. In terms of blood chemistry, the results showed elevated levels of ASAT at 130U/L, ALAT at 90U/L, and lactic dehydrogenase at 310U/L. Heart Ultrasound showed sinus tachycardia. She was administered packed red cell transfusion. We began empirical antibiotics due to neonatal fever with DIC profile and high dose methylprednisolone (30mg/kg/day) therapy for MAS. Her body temperature was normalized at 3days later after methylprednisolone treatment. We obtained results of decreased natural killer cell activity (about 8.8% of normal activity in effector versus target ratio 1:20) and high-level of soluble interleukin 2 (6102U/mL, normal level: 158 – 632 U/mL). Her autoimmune study showed ANA (1:350 with Speckle pattern), anti-Ro/SSA positive (3+), and anti-La/SSB positive (3+).

Figure 2: crumb

Therefore, the diagnosis of MAS in neonatal lupus was established according to MAS criteria [6]. The baby with normal vital sign was showed good activity and feeding and laboratory data including CBC and blood chemistry recovered on the 7th hospital day except a high level of ferritin (Figure 2). She was discharged with oral dexamethasone on the 10th in hospital day and steroid was stopped according to tapering schedule 1 month later. Her mother without any autoimmune manifestations showed ANA (1:1420 with speckle pattern), anti-Ro/SSA positive and anti-Lo/SSB positive. Nine months later, baby's growth and development along with laboratory data were within normal ranges. Also, mother did not exhibit any autoimmune manifestation despite the presence of auto-antibodies. The patient's laboratory data during clinical course are shown in Figure 2.

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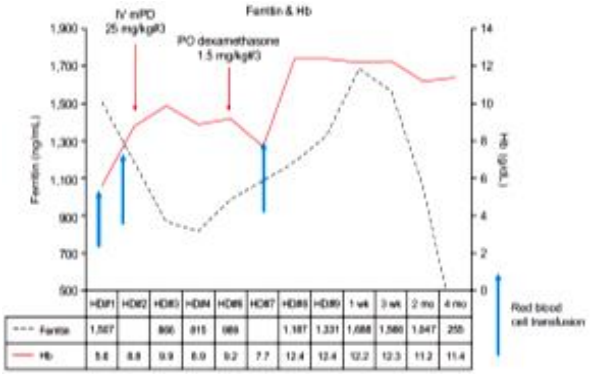

Figure 3 : Crumb

DISCUSSION:

In this case, the patient showed fever with erythema marginatum like skin rash. She developed fever with pancytopenia and skin rash although she was diagnosed with neonatal lupus at another hospital several days ago [1]. At that time, her mother's auto-antibodies were identified although the mother did not have any autoimmune features. The delivery and mother's history were not specific. Pediatricians commonly consider neonatal sepsis for febrile infants because neonatal fever is a very serious clinical presentation. However, this baby was diagnosed with neonatal lupus at another hospital on the basis of autoantibodies. This baby was initially not compatible with hemophagocytic lymphohistiocytosis (HLH) diagnostic criteria. At admission, we identified fever, erythema marginatum like skin rash, pancytopenia, high level of ferritin, and splenomegaly. These findings represented as early prediction for proposed MAS in systemic juvenile idiopathic arthritis and preliminary diagnostic guideline. Finally, the baby was diagnosed with secondary HLH after we obtained results of high soluble IL-2 and decreased natural killer cell activity. Skin lesions are not part of the diagnostic criteria for HLH or MAS. However, skin eruption can be a significant clinical manifestation in HLH or MAS. While we did not conduct a skin biopsy, it is worth noting that in cases of HLH/MAS, a skin biopsy can be clinically relevant, as it may reveal lymphohistiocytic infiltrates in the dermis without any epidermal abnormalities. Therefore, opting for a skin biopsy rather than bone marrow examination can confirm the diagnosis of MAS in neonatal lupus. MAS is a form of secondary HLH that can be triggered by diseases such as infections or rheumatic diseases. In our case, she had been previously diagnosed with neonatal lupus at another hospital, which could be diagnosed as a manifestation of MAS, although clinical findings were compatible with HLH.

In our case, fever and high ferritin with pancytopenia were initial clinical findings, similar to other reported MAS in neonatal lupus. Thrombocytopenia with high levels of ferritin and soluble IL-2 are common features in neonatal lupus with MAS. Neonatal lupus with MAS also shows increased levels of aspartate transferase or alanine transferees. MAS can be treated with steroid therapy as a monotherapy. In the present case, the baby received high dose mPD with oral dexamethasone following the HLH 2004 protocol and recovered, although she required transfusion. Her ferritin level was high three weeks later, which might be related to three times red blood cell transfusions. It normalized at 4 months. Relapsed cases show use of hydrocortisone and delayed diagnosis. Hydrocortisone has lesser anti-inflammatory potency than other steroids. Also, delayed diagnosis could progress to severe inflammation, although intravenous immunoglobulin and methylprednisolone were administered for MAS. Therefore, optimal treatment time and strong anti-inflammatory therapy might be needed for improvement, especially for neonatal lupus with MAS.

In our case, the patient was well after a one-time therapy. These Findings suggested that the MAS was due to neonatal lupus, not due to a genetic problem. We report a case of neonatal lupus with MAS presented with fever and pancytopenia. The baby was born from a primiparous mother without any autoimmune manifestations. Thus, neonatal MAS should be considered when an infant presents fever with pancytopenia, although the baby is born from a healthy mother without any autoimmune findings.

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