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Case report

Facial rash in a newborn

Sílvia Mota[1], Carla Sá[2], Nicole Silva[2], Filipa Almeida[3], Ana Paula Vieira[3], Almerinda Pereira[1,2]

[1]Pediatric Service, Hospital de Braga, Braga, Portugal
[2]Neonatal Special Care Unit, Hospital de Braga, Braga, Portugal
[3]Dermatology Service, Hospital de Braga, Braga, Portugal

Abstract

A female newborn, born at 38 weeks from an uncomplicated pregnancy in a healthy 36-years-old mother, presented at birth with desquamative erythematous plaques with irregular borders, distributed bilaterally in the orbital, nasal and malar regions. Although there was no history of maternal autoimmune disease, neonatal lupus (NL) was suspected. Maternal and newborn screenings were positive for Sjögren syndrome type A antigen (anti-Ro/SSA) antibodies. No other alterations of NL were found in the newborn. Rheumatologic consultation on the mother showed no other alterations besides the antibodies. The newborn was discharged on day 3 of life without treatment and recommendations to avoid sun exposure. Outpatient follow-up was ensured in neonatology, dermatology and pediatric cardiology. The rash resolved during the first year of life, leaving slight local skin atrophy.

NL is a rare transferred autoimmune disease with an incidence estimated as 1:20,000 live births. It occurs due to placental transfer of maternal autoantibodies. The major manifestations are cardiac and cutaneous, but hepatic, hematologic or neurologic findings may also be present. The rash usually affects the face and scalp and may be present at delivery but more often develops later, after exposure to ultraviolet light. It usually resolves within the first year of life without sequelae. NL is the leading cause of congenital heart block, but if it is not present at birth it rarely develops. Some mothers do not have a known autoimmune disease at the time of birth but may develop it later in life. Despite NL being a passively acquired autoimmune disease, the child is at increased risk of rheumatologic disease in childhood or adolescence.

Keywords

Neonatal lupus, neonatal autoimmune disease, Sjögren syndrome type A antigen antibodies, congenital heart block, facial rash, skin lesions.

Corresponding author

Sílvia Mota, Pediatric Department, Hospital de Braga, R. das Comunidades Lusíadas 133, Sete Fontes – São Victor, 4710-243 Braga, Portugal; telephone number: +351939040512; e-mail: silviamota.scm@gmail.com.

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Introduction

Neonatal lupus (NL) is a rare condition with an estimated incidence of 1:20,000 live births [1]. Its pathogenesis is based on the transplacental passage of maternal autoantibodies, more frequently Sjögren syndrome type A antigen (anti-Ro/SSA) antibodies and Sjögren syndrome type B antigen (anti-La/SSB) antibodies, but cutaneous manifestations have also been described in the presence of anti-ribonuclear protein (anti-RNP) antibodies [2-4]. Nevertheless, only 1-2% of newborns whose mothers have auto-antibodies develop NL [1, 5].

Case presentation

A female newborn was born via cesarean delivery (the mother had 2 previous caesareans) at 38 weeks of gestational age from an uncomplicated pregnancy with appropriate prenatal care. The mother was a healthy 36-year-old woman with 2 older children whose pregnancies were also uneventful, resulting in 2 healthy babies. At birth, this baby presented with desquamative erythematous plaques with irregular borders, distributed bilaterally in the orbital, nasal and malar regions (**Fig. 1**). Her anthropometric measures were appropriate for gestational age, and the remaining physical examination was normal. Laboratory investigation was performed concerning infection, but the results were all within the normal range (blood count with differential, liver enzymes and C-reactive protein).

Dermatology observation raised the suspicion of NL, and although there was no history of maternal autoimmune disease, investigation was started. Maternal and newborn screenings were positive for anti-Ro/SSA antibodies. A cardiac evaluation was performed on the newborn with electrocardiogram and echocardiogram that were both normal. A rheumatologic consultation on the mother showed no other alterations besides the antibodies. The newborn was discharged home on day 3 of life without pharmacological treatment and recommendations to avoid sun exposure. Outpatient follow-up was ensured in neonatology, dermatology and pediatric cardiology. The rash resolved during the first year of life, leaving slight local skin atrophy.

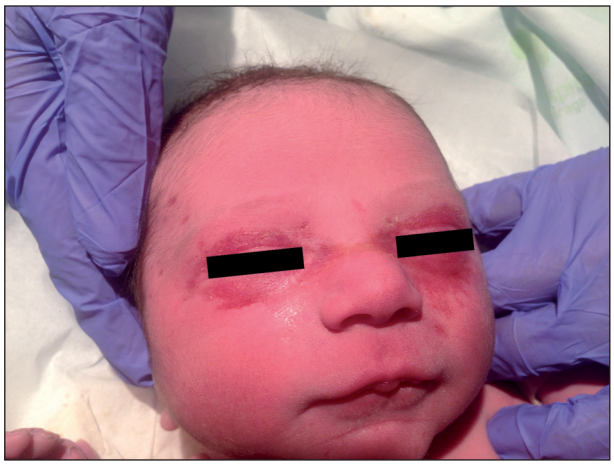

Figure 1. At birth, this baby presented with desquamative erythematous plaques with irregular borders, distributed bilaterally in the orbital, nasal and malar regions.

Discussion

The most frequent manifestations of NL are cutaneous (rash) and cardiac (various degrees of heart block or cardiomyopathy) [2], but other organ systems can be affected as hepatobiliary (elevated liver enzymes, cholestasis or hepatomegaly), hematological (more frequently thrombocytopenia but any kind of cytopenia may occur) and central nervous system [2, 3, 6]. NL is responsible for more than 80% of complete congenital heart block. [5] Although most of the manifestations of NL are benign and temporary, the cardiac disease is permanent and has a mortality rate of 20% [2, 3, 7].

The cutaneous manifestations are more common in girls with a proportion to 2:1 [1]. The same child can have more than one manifestation of NL [1, 7-10].

The body areas more commonly affected in cutaneous NL are the face and scalp (the peri-orbital region seems to be particularly prone to the rash); nevertheless, any skin area can be affected [1, 5, 7, 8]. The lesions may be present at birth but more frequently appear in the first weeks of life, after some sun/ultraviolet light exposure [1, 2, 9]. The skin lesions usually disappear spontaneously and without sequelae, but residual scaring or telangiectasias may occur [1, 5, 7-9]. Its disappearance seems to

be correlated with the loss of the passively acquired auto-antibodies [5]. As cutaneous NL is mostly a self-limited condition, treatment with topical agents such as low potency corticosteroids is generally unnecessary, but may be considered [1, 5]. Although exposure to ultraviolet light is not mandatory to the development of the skin lesions, avoidance of sun exposure should always be recommended [11].

A cutaneous NL diagnosis is made when there are a combination of the typical cutaneous findings and the presence of antibodies in the mother and the newborn [1, 7]. Skin biopsy is not usually performed since it does not contribute to the diagnosis but, if it is performed, the histological findings are similar to those in the cutaneous manifestations of subacute cutaneous lupus of the adult [1, 9].

Although the cutaneous findings of NL are self-limited, follow-up of the child must be assured due to the increased risk of developing an autoimmune disease later in life [1, 8, 12].

The mother of a child with NL has a risk of recurrence up to 25% to any NL manifestation in a following child, so adequate prenatal counseling and pregnancy surveillance should be guaranteed [8]. Two affected siblings, including twins, can have different manifestations of the disease, suggesting that other factors besides the auto-antibodies play a role in the development of NL [10].

In this case, the mother did not have an autoimmune disease and studies show that it happens in up to 28% of mothers of a child with NL, but half of them will develop it in a 3 to 5-year time period [8, 13].

Conclusion

NL is a passively acquired neonatal autoimmune disease. This newborn presented only with cutaneous manifestations, and besides slight residual skin atrophy, she has no other lasting effects of the disease up to this date. Both mother and child with NL should maintain follow-up because of the risk of developing autoimmune disease in the future.

Ethics and patient consent

This case was written in accord to all ethical considerations. Written patient consent was obtained from the mother.

Declaration of interest

The Authors declare that there is no conflict of interest. The Authors received no specific funding for this work.

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