



Lúpus eritematoso neonatal: Resumo expandido

Neonatal lupus erythematosus: Expanded summary

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ABSTRACT

Neonatal Lupus Erythematosus (ENL) is a rare autoimmune disease in which immunoglobulin G is transported through the placenta into the fetal circulation. It affects both males and females. It has been noticed that the most common clinical manifestations are: skin changes, hematological, hepatobiliary and cardiac involvement such as atrioventricular block. However, some of these symptoms tend to disappear naturally after a few months. The diagnosis can be made by biopsy, which will show histopathological mechanisms similar to those of discoid lupus and direct immunofluorescence will show the presence of IgG at the dermal-epidermal junction. The aim of this article was to improve knowledge through an integrative literature review, which used a synthesis of results obtained through already published research in order to argue the results found. It was found that the use of sunscreen, healthy habits and lifestyle and palliative treatment, when applied appropriately, can control and even make the symptoms of this disease disappear.

Keywords: Lupus, Pregnancy, Autoimmune disease.



1 INTRODUCTION

The first description of Neonatal Lupus Erythematosus (NLE) was made by Bridge and Foley in 1954, who noticed and reported the maternal-fetal transmission of the antibodies responsible for this condition. Currently, neonatal lupus erythematosus (NLE) can be classified as an autoimmune disease belonging to a group of medical conditions in which immunoglobulin G is transported through the placenta into the fetal circulation and directed against self antigens, causing clinical manifestations in the newborn.¹ Thus, ENL is a rare disease and an example of passively acquired autoimmunization. It can be defined as a multiform clinical picture observed in the children of mothers with anti-SS-A and/or anti-SS-B antibodies.²

Currently, the incidence of neonatal lupus erythematosus is close to 2% in mothers with Sjögren's syndrome type A (Ro/SSA) or B (La/SSB) autoantibodies, a rate that rises to 18% to 20% in subsequent pregnancies of mothers who have already had newborns affected by this condition.¹ However, although there are estimates in numbers, the true prevalence of ENL has not been established due to the high proportion of unrecognized cases. In terms of its manifestation, ENL does not show a significant difference between the sexes, affecting men and women equally. Among the cases of ENL, it is estimated that less than 5% of children with this condition develop systemic lupus erythematosus in late adolescence or early adulthood.^{1,2} Given the difficulty in diagnosing the pathology, it is essential to approach and understand ENL in order to enable proper diagnosis and treatment of this disease, avoiding errors and ensuring a good prognosis for patients with this condition.

2 OBJECTIVE

The general aim of this article is to seek out relevant information on the medical spectrum in relation to the pathology discussed. In addition to addressing the main characteristics of the study presented, definition, pathophysiology, symptoms, diagnosis and treatment, through analysis and interpretation of scientific articles documented on the subject.

3 METHODOLOGY

The current study was produced using bibliographic studies referenced in the PubMed, Scientific Electronic Library Online (SciELO) and UpToDate databases. Neonatal lupus erythematosus was chosen as the topic. From this perspective, articles published in the following languages were used: Portuguese, Spanish and English. In addition, the inclusion criteria were articles published from 2017 onwards that were related to the topic mentioned, as well as exclusion



criteria for publications published less than 2017, with no direct relation to the subject. In this way, the analysis was carried out on a total of 6 articles published between 2017 and 2022.

4 DEVELOPMENT AND RESULTS

Neonatal lupus erythematosus is a set of symptoms that is presented in newborns in which the mother has antibodies to soluble cell nucleus antigens. Anti-Sjögren's Syndrome B (anti-SSB) antibodies, also known as anti-La, and anti-Sjögren's Syndrome A (anti-SS-A) antibodies are the main factors contributing to neonatal lupus. From the 12th week of pregnancy, the mother's antibodies begin to be transferred to the child via the placenta, but these antibodies trigger a cascade of inflammatory reactions in the fetus, leading to the symptoms of neonatal lupus.³

The symptoms of neonatal lupus can be reversible or irreversible, the cutaneous and hematological presentations are usually reversible in most cases, however, some children evolve to disorders of the conduction system and cardiac stimulation, generating a first or second degree atrioventricular block, mainly third degree atrioventricular block, which are irreversible symptoms, about 2% of these children have irreversible symptoms.⁴

The pathophysiological mechanism that explains the cardiovascular block in these children is the fact that the antigens translocate to the surface of the cardiomyocytes, the maternal antibodies that have been transmitted to the fetus trigger fibrosis in the atrioventricular node, which causes the atrioventricular block.^{1,4}

Skin lesions, on the other hand, are often mistaken for birth trauma, rash or skin infection and are often misdiagnosed. These newborns present with photosensitivity within the first 2 days of birth, as well as skin lesions characterized by an inflammatory rash, papular or plaque lesions, classic exanthema, persistent rash characterized by telangiectasia, atrophy and hyperpigmented areas. Histological examination identifies granular deposits of immunoglobulin G at the dermoepidermal junction and vascular alterations at the interface and adnexal structures.¹

The hematological and hepatobiliary clinical manifestations are mostly volatile and generally do not present in isolation in ENL. Newborns with this syndrome may present with anemia, thrombocytopenia and neutropenia, and in 20% of cases they may present with aplastic anemia. On the hepatobiliary side, these newborns present with an asymptomatic elevation of aminotransferases that occurs in the first few months of life; cholestasis or hepatomegaly, which can reach 1/4 of cases. In some cases, severe liver failure can also occur in the neonatal period, which may be related to neonatal hemochromatosis.¹



In order to diagnose ENL, there must be a compatible clinical picture associated with the presence of autoantibodies in the mother's and child's serum, with the aim of finding anti-Ro/SSA, anti-La/SSB and anti-U1RNP antibodies. If the skin picture is uncertain, the diagnosis can be made by biopsy, direct immunofluorescence will show the presence of IgG at the dermal-epidermal junction. In addition, a complete blood count, liver function tests and an electrocardiogram plus echocardiogram should be ordered to assess the cardiovascular system.^{5,6}

In general, the appropriate therapeutic approach is to monitor the patient's progress, as most of the manifestations of neonatal lupus are self-limiting and resolve with the disappearance of maternal antibodies from the neonate's bloodstream. Sun protection is recommended and topical corticosteroids are often indicated for skin lesions. Treatment of BAV is necessary when the presence of fetal bradyarrhythmias or myocarditis is detected, in which case the mother should be treated with dexamethasone to prevent heart damage.⁶

5 CONCLUSION

Given the above, we consider the study of the literature on ENL to be of great value, due to the low dissemination of the subject in the medical community, as it is often underdiagnosed. Bearing in mind that some symptoms soon disappear and others, such as changes in the cardiovascular system, can remain for longer. In this way, disseminating information on the subject helps to diagnose ENL more quickly.



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