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Seronegative Neonatal Lupus: Case Report

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Abstract:- Neonatal lupus erythematosus (NEL) is a rare autoimmune disease characterized by transplacental transmission of maternal antibodies, primarily directed against SSA/Ro or SSB/La proteins. Clinical manifestations include cutaneous, hematologic, hepatic, and cardiac symptoms, including congenital atrioventricular block (cAVB). Although cutaneous symptoms are often reversible and treated with topical steroids, cardiac complications, such as cAVB, are considered irreversible and can be fatal. Pregnant women with specific antibodies should be screened, and fetal echocardiography is recommended for prenatal screening. Traditional treatments include corticosteroids, although hydroxychloroquine and intravenous immunoglobulin (IVIG) may be effective alternatives to reduce the risk of cardiac complications in newborns.

I. INTRODUCTION

Neonatal lupus erythematosus can present with cutaneous, hematologic, and hepatic symptoms, as well as possibly congenital atrioventricular block (cAVB) in newborns without preexisting cardiac abnormalities. This rare syndrome is caused by transplacental transfer of maternal antibodies, primarily directed against SSA/Ro proteins or, less commonly, against SSB/La proteins. [1,2,3]

II. OBSERVATION

3 month old infant, born at term, vaginal delivery,from non-consanguineous parentsno family history of lupus. Presented for 1 month with pallor, abdominal distension with erythematous skin lesions. Examination found hepatosplenomegaly, umbilical hernia, mucocutaneous pallor and annular, infiltrated, pale-centered erythematous lesions located on the face, abdomen, upper and lower limbs (Figure 1).

The assessment showed an inflammatory syndrome with an ESR of 45 mm in the first hour and a CRP of 71.6 mg/l. Normochromic normocytic anemia at 6.6 with a negative Coombs test. Lymphopenia at 900/mm3. Thrombocytopenia at 74,000/mm3. Mild hepatic cytolysis. The myelogram showed an absence of blast cells, malignant cells and overload cells. Negative 24-hour proteinuria with a positive urine cytobacteriological examination (ECBU) for E.coli and normal renal function. The dosage of complements C3, C4 and antinuclear and anti-DNA antibodies was negative, TORCH serologies were negative.

Skin biopsy showed a skin covering with few lesions with vacuolar dermatosis in favor of lupus disease. Abdominal ultrasound showed hepatomegaly with a hepatic arrow at 98 mm of homogeneous echo structure, splenomegaly with a splenic arrow at 84 mm and an umbilical hernia with uncomplicated omental content. Skeletal radiography showed slight bone demineralization, metaphyseal widening and flattening, chest radiography and transthoracic ultrasound were normal. The immunological assessment in the mother was negative.

The patient was put on antibiotic therapy for the urinary tract infection and on oral corticosteroid therapy at a dose of 1 mg/kg/day. The outcome was favorable.

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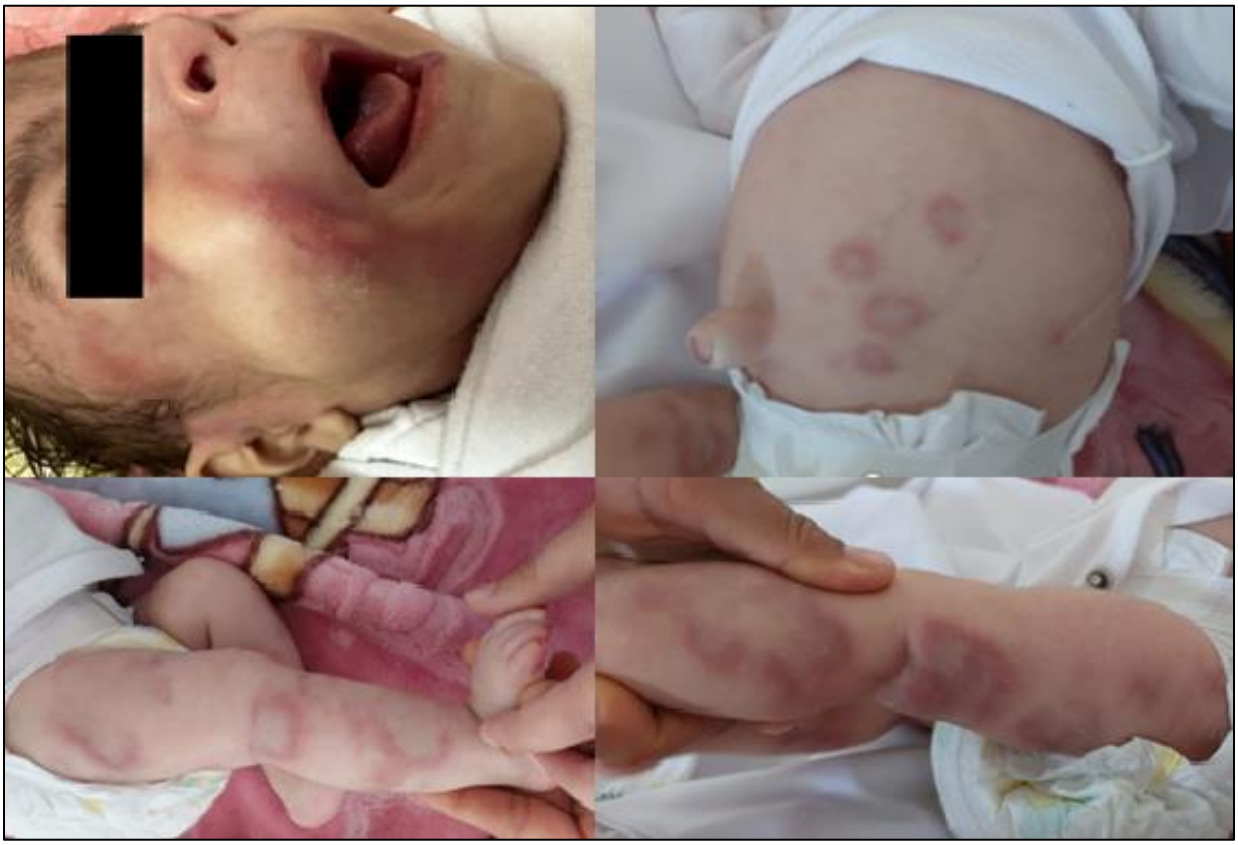

Fig 1:Annular, Infiltrated Erythematous Lesion with a Pale Center Located on the Face, Abdomen and Lower Limb.

III. DISCUSSION

Neonatal lupus erythematosus (NEL) is a rare autoimmune disease with a prevalence of <1/20,000 births [4]. In 1954, Bridge and Foley noted that maternal "lupus erythematosus factor" could be transmitted to the baby. That same year, a lupus rash was observed in a 6-week-old infant whose mother was diagnosed with systemic lupus erythematosus a few months later. In 1957, congenital heart disease was described in an infant whose mother also had the condition. Finally, in infants with neonatal lupus erythematosus, episodes of transient cytopenia and abnormally elevated aminotransferase levels were first reported a few years later. [5,6]

NLE develops when maternal antinuclear antibodies, mainly IgG antibodies such as anti-SSA/Ro, anti-SSB/La, or anti-RNP, cross the placenta from the 12th week of gestation. Although most women carrying these antibodies have healthy children, the risk of neonatal lupus is estimated at 1 to 2, suggesting the involvement of genetic and environmental factors. This disease, like other autoimmune diseases, is characterized by the presence of autoantibodies targeting the heart and other molecules, causing a variety of symptoms ranging from bradycardia to serious complications such as complete atrioventricular block or fetal death. However, the low prevalence of the disease in mothers carrying anti-SSA/Ro and/or anti-SSB/La autoantibodies remains unexplained, possibly due to the specificity of the maternal autoantibodies and the timing of their production during pregnancy. Furthermore, it is clear that the fetus is not only a passive target of the autoimmune attack, but can also defend itself effectively, as evidenced by the spontaneous recovery to normal of cardiac anomalies in some fetuses. [4,7]The clinical manifestations of LENN are divided into two categories: reversible and irreversible. Among the reversible symptoms, cutaneous manifestations are the most frequent, while hepatobiliary, hematological or neurological involvement are less common. In contrast, cardiac manifestations such as atrioventricular block are considered irreversible.

The cutaneous manifestations of neonatal lupus erythematosus usually appear by 6 weeks of age, but they may also be present at birth or appear later, by 5 months of age. Sensitivity to sunlight is observed in 94% of children with skin lesions, and sun exposure appears to accelerate their development. Skin lesions have also been observed after neonatal phototherapy for hyperbilirubinemia. They present as rounded erythematous macules or papules, sometimes covered with fine scales. Although the face, particularly the periorbital region, is commonly affected, lesions can also occur anywhere on the body, including the scalp, without affecting the mucous membranes. The main features of the cutaneous form of neonatal lupus erythematosus usually include the presence of

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multiple annular erythematous lesions or arcuate macules. This rash is often described as having a similar appearance to a raccoon or owl eye. Although the face is the most commonly affected area, the palms, soles of the feet, or diaper area may also be affected. [8,9,10,11]

Liver damage associated with neonatal lupus erythematosus (NLE) typically presents as an asymptomatic increase in alanine aminotransferase and/or aspartate aminotransferase, which may be accompanied by signs of cholestasis. These elevations are usually transient and resolve spontaneously within the first few months of life, leaving no sequelae. Some infants may have mild hepatomegaly and, less commonly, splenomegaly. [12]

In approximately 10–20% of infants, typical hematologic symptoms occur, including anemia, thrombocytopenia, and, less commonly, neutropenia. Less common manifestations include immune thrombocytopenic purpura, microangiopathic hemolytic anemia, disseminated intravascular coagulation, and thrombosis. [9,13]

Complete atrioventricular block (AVB) is considered the most severe form of neonatal lupus because of its irreversible nature and its association with a high risk of serious or even fatal complications. Other cardiac complications, such as incomplete blocks and more recently sinus bradycardia, have also been observed.

These cardiac manifestations include conduction abnormalities such as first-, second-, and third-degree atrioventricular block, as well as cardiomyopathies. In addition, conduction abnormalities may manifest as irregular heart rhythm and QT interval prolongation. [11,14] Our case presents cutaneous, hepatic, and hematological involvement, without cardiac involvement.

Skin lesions are usually treated with topical steroids, and they usually heal within eight months without leaving any consequences. LENN has manifestations that can be detected after birth, including serious cardiac complications such as complete atrioventricular block (AVB). All pregnant women should be tested for antinuclear antibodies, and fetal echocardiography is recommended for prenatal screening in women with specific antibodies. Traditional treatments involve the use of corticosteroids, although their effectiveness is questionable. A possible alternative is hydroxychloroquine, which may reduce the risk of cardiac manifestations in newborns. Studies have also shown positive results with the use of intravenous immunoglobulin (IVIG) to prevent AVB in fetuses. [9,10]

IV. CONCLUSION

Neonatal lupus erythematosus is characterized mainly by dermatological and cardiac symptoms. However, clinical manifestations can vary, which can complicate diagnosis and cause delays. Its diagnosis requires clinical evaluation, since lupus-specific tests are not part of routine examinations for rashes in newborns. Regular cardiac monitoring is recommended in women who have had a child with this disease, as well as careful monitoring of their future children in subsequent pregnancies.

REFERENCES

[1]. N. Costedoat-Chalumeau et al. “Neonatal lupus”: review of the literature. The journal of internal medicine 24 (2003) 659–671
[2]. Lela A. Lee. Neonatal Lupus Clinical Characteristics and Management. Pediatr Drugs 2004; 6 (2)
[3]. Morel N, et al. “Neonatal lupus”: literature review. Rev Med Interne (2014),
[4]. J. Blanc et al. Polymorphism and differential diagnoses of neonatal lupus erythematosus. Annals of Dermatology and Venereology (2020) 147, 258—264.
[5]. Bridge RG, Foley FE (1954) Placental transmission of the lupus erythematosus factor. Am J Med Sci 227:1–8
[6]. Federica Vanoni. Sebastiano AG Lava. Emilio F. Fossali. Riccardo Cavalli. Giacomo D. Simonetti. Mario G. Bianchetti. Marie-Ange Bozzini. Carlo Agostoni. Gregorio P. Milani. Neonatal Systemic Lupus Erythematosus Syndrome: a Comprehensive Review. Clinic Rev Allerg Immunol. 2017.
[7]. J. Hoebeke, S. Muller. Neonatal lupus: new hypotheses concerning immunopathogenesis. Revue du Rhumatisme 72 (2005) 159–161
[8]. N. Costedoat-Chalumeau et al. “Neonatal lupus”: review of the literature. The journal of internal medicine 24 (2003) 659–671
[9]. Derdulska et al. Neonatal lupus erythematosus. J. Perinat. Med. 2021; 49(5): 529–538
[10]. Ramphul et al. Cutaneous Neonatal Lupus Erythematosus: A Case Report 2018. Cureus 10(2): e2212.
[11]. Kam Lun Hon and Alexander KC Leung. Neonatal Lupus Erythematosus. Hindawi Publishing Corporation Autoimmune Diseases Volume 2012, 6 pages
[12]. E. Silverman & E. Jaeggi. Neonatal Lupus Erythematosus. 2010 Scandinavian Journal of Immunology 72, 223–225.

ISSN No:-2456-2165 https://doi.org/10.38124/ijisrt/IJISRT24OCT257

[13]. Benay Johnson. Overview of Neonatal Lupus. Journal of Pediatric Health Care. 2013
[14]. R. Cimaz, A. Duquesne. Neonatal lupus. Archives of Pediatrics 13 (2006) 473–478
[15]. Jérôme Coulombe, Julie Barsalou. Neonatal lupus manifesting as a butterfly-shaped rash. CMAJ 2021.