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Brief report

Neonatal Lupus Erythematosis: A Five-Year Case Review☆

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ABSTRACT

Neonatal lupus erythematosus is an infrequent disease seen in newborns. It is caused by transplacental maternal autoantibody passage. Cutaneous involvement and congenital heart block (CHB) are the most common affections, although it may involve multiple organs such as the liver, lungs, blood, nervous or digestive systems.

This article present a review of the four cases diagnosed in the past five years in a Neonatal Unit, which shows the different clinical spectrum which can develop around this disease (CHB, multisystemic affection and two cutaneous cases), different autoantibodies (specially anti-SSA) with an early negativization during the first year of life and the possibility of future collagen vascular disease as occurred in one case.

Lupus eritematoso neonatal: revisión de casos en los últimos 5 años

Palabras clave:
Lupus eritematoso neonatal
Afectación multisistémica
Bloqueo cardíaco congénito
Autoanticuerpos

RESUMEN

El lupus eritematoso neonatal es una enfermedad rara del recién nacido producida por el paso transplacentario de autoanticuerpos maternos. Las 2 formas de presentación más frecuentes son la dermatológica (lupus eritematoso subagudo) y el bloqueo auriculoventricular completo (BAVC). También puede producir afectación hematológica, hepática, neurológica, respiratoria y digestiva.

Presentamos una revisión de 4 casos diagnosticados en los últimos 5 años en nuestra Unidad de Neonatología, que reflejan el amplio espectro clínico con el que se puede presentar esta enfermedad (un caso de BAVC, uno con afectación multisistémica y 2 casos con expresión cutánea), los diferentes patrones de autoanticuerpos (con un predominio de anticuerpos anti-SSA), la desaparición de autoanticuerpos en todos los casos antes del año de edad y la posibilidad de aparición de colagenopatías en el futuro, como ocurrió en uno de nuestros casos.

Introduction

NL is a rare disease with an estimated incidence of 1/10 000-20 000 in newborns (NB) and predominantly females. It occurs due to transplacental passage of maternal IgG autoantibodies, usually anti-Ro (SSA) (95%), anti-La (SSB) and less frequently anti-U1RNP. A case was recently been described in our country caused by anti-Sm. Although it can affect multiple organs, the 2 most common clinical forms are dermatological involvement (subacute lupus erythematosus), present in 50% of cases, and complete atrioventricular block (CAVB), found in 50% of NB. Both coexist only in 10% of cases.[1,2]

Other cases present with hepatic, hematological, neurological, respiratory, and digestive manifestations, which usually subside before one year of age, coinciding with the disappearance of maternal autoantibodies. Conversely, CAVB becomes chronic in most cases, meriting an early pacemaker.[3–5]

NL diagnosis requires a high index of suspicion and the absence of a family history makes it difficult. Only 50% of mothers present connective tissue disease symptoms at diagnosis, particularly systemic lupus erythematosus (SLE) or Sjögren's syndrome. The rest, though asymptomatic, are at risk of developing it in the future.[1–3]

We present a review of 4 cases diagnosed in the last 5 years in our Neonatal Unit (Table 1).

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Table 1
Clinical and Laboratory Characteristics.

	Case 1	Case 2	Case 3	Case 4
Gender	Female	Male	Female	Female
Maternal history	SLE	Asymptomatic	JIA	Asymptomatic
Dermatological involvement	No	Yes	Yes	Yes
Cardiac involvement	Yes	No	No	No
Other organ involvement	No	Yes	No	No
Autoantibodies in the mother prior to preganancy	Anti-SSA	ANA negative	Anti-SSA, anti-SSA and anti-Sm/RNP	Not investigated
Autoantibodies present at birth	Anti-SSA	Anti-SSA and anti-SSB	Anti-SSA, anti-SSB and anti SM/RNP	AntiU1RNP
Autoantibodies after the first year of life	Negative	Negative	Negative	Loss of follow up
Autoimmune diseases after first birthday	No	No	Anterior uveitis	Loss of follow up

Clinical Observation

Case 1

Female RN diagnosed in the first trimester of CAVB. Mother with SLE and Evans syndrome with positive anti-SSA antibodies in treatment during pregnancy with dexamethasone 4 mg/day.Because of possible complications, she was delivered at a referral hospital, where she underwent a pacemaker placement at birth with good outcome. She had no skin lesions or systemic involvement, although anti-SSA autoantibodies were positive. Follow up was conducted without further connective tissue disease manifestations appearance to date, in which the patient is 4 years old.

Case 2

Black male NB admitted with respiratory distress and skin lesions. Obstetric history: second pregnancy, uneventful. Normal childbirth at week 36+2. Apgar 8/10. Weight: 2.040 g. Length: 44 cm. Head circumference: 31.5 cm. Family history: mother, 26, a native of Senegal, followed for chronic hepatitis B, anemia and mild leukopenia, and polyclonal hypergammaglobulinemia. Antinuclear antibodies (ANA) were negative. Two years before she had presented hyperpigmented skin lesions on the left cheek, self-limited, without receiving a diagnosis.

Clinical examination showed scaly erythematous brownish plaques on the skin on the face, with periocular mask distribution, as well as on the ears, scalp, trunk and upper extremities (Figs. 1 and 2). Mild tachypnea without distress, and good bilateral ventilation. Abdomen with hepatomegaly 1 cm and splenomegaly 2 cm below the rib margin. Neurologically, hyperexcitable with trembling and mild generalized hypertonia and hyperreflexia.

Laboratory tests showed: thrombocytopenia (19,000 mm^{-3}), leukopenia (4290 mm^{-3}) and hemolytic anemia: (Hb 7.6 g/dl, hematocrit 23%, reticulocytes 2%, haptoglobin: 6.63 mg/dl). A blood smear presented intense anisocytes, poikylocytes, and some teardrop schistocytes. There was mild hypertransaminasemia mild (glutamicoxalacetic transaminase 119 U/l; glutamic pyruvic transaminase 62 U/l). Normal ECG. Normal chest radiograph. A transfontanelar ultrasound showed a hyperechoic white matter, doubtful microcalcifications and scattered foci of bleeding at the level of the germinal matrix. MRI showed discrete alteration of the white matter, indicating incipient encephalomalacia changes. Skin biopsy showed mild hyperkeratosis, minimal epidermis basal layer vacuolar changes and abundant mucin deposition in the dermis. Positive ANA, speckled pattern, title 1/1280. Anti-SSA and anti-SSB positive, anticardiolipin, anti-native DNA, anti-Sm and anti-Sm/RNP were negative.

With the diagnosis of NL with cutaneous, hematologic, respiratory, hepatic and neurological involvement, treatment was initiated with topical corticosteroids and tacrolimus. For the hematological manifestations, the patient required treatment with gamma globulin at 4 g/kg and intravenous corticosteroids (methylprednisolone 2 mg/kg/day with subsequent reduction, maintained for 53 days) and transfusion of packed red blood cells and irradiated platelets. He required oxygen for 21 days. At discharge the patient presented Hb 10.1 g/dl, leukocytes 8400 mm^{-3} and 307,000 mm^{-3} platelets.

At 2 months, erythematous annular lesions with central necrosis of a discoid lupus type reappeared on the trunk and extremities, with atrophy and scar hyperpigmentation, which resolved at 4 months of age. Hematologic abnormalities were normalized after 2 months. ANA were negative at 8 months of life.

Currently, the patient is 3 years of age and continues monitoring without evidence of connective tissue disease and presenting only residual scars on cutaneous lesions (Fig. 3).

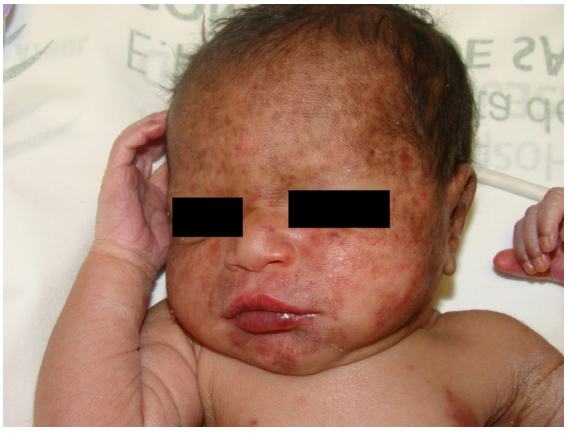

Fig. 1. Clinical case 2. Brownish erythematous scaly lesions on the face, with periocular mask distribution.

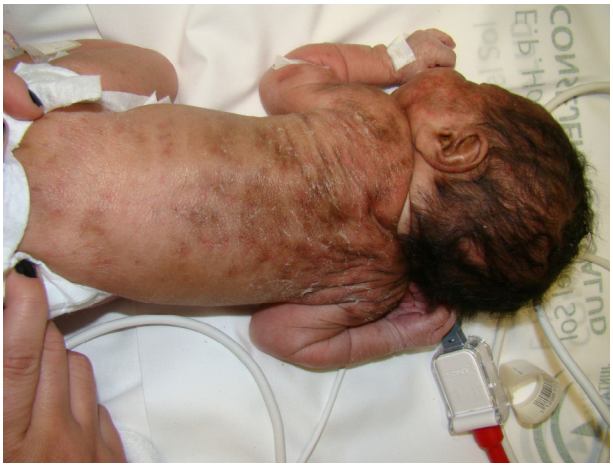

Fig. 2. Clinical case 2. Brownish erythematous lesions on the back.