

Neonatal Lupus

What is it about?

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Two interesting cases of patients whose diagnoses turned out to be a challenge are presented.

Introduction

- Neonatal Lupus (NL) is an autoimmune disease passively acquired and related to transplacental transmission of maternal autoantibodies Anti SSA/Ro and Anti SSB/La.
- It can be presented in 1 out of 20 thousand born alive but only 1-2% of newborn with positive maternal autoantibodies could develop NL.
- Cutaneous affection is temporal with spontaneous resolution in the 7-12th month of life together with brighten of maternal autoantibodies.
- The cardiac involvement is permanent, which darkens the disease prognosis.

Case 1

- 2-year-old girl comes from city area, mother with systemic lupus erythematosus (SLE) as antecedents.
- Obstetrics history: full-term born by normal delivery, with right weight, APGAR 9/9.
- She has had mild annular erythematous cutaneous lesions on her face, neck and thorax since birth, photosensitive. At the time of evaluation she presented light malar rash, resolving skin neck injuries with hypopigmented area, ocular conjunctive edema and oral ulcers (figures 1-2).
- With this personal antecedent of mucocutaneous lesions, a cardiological examination without alterations, a positive immunological profile (qualitative ANA positive, anti SS/Ro 32/ 25 U/ml) and degeneration of the basal lamina, edema patchy mononuclear infiltration of the dermis at damage skin biopsy, the NL was considered in a retroactive way.
- Actually the patient is followed as SLE minor manifestations when lymphopenia was added to the clinical picture, under treatment just hydroxychloroquine.



Figures 1-2: light malar rash, resolving skin neck injuries with hypopigmented area, ocular conjunctive edema and oral ulcers.

Case 2

- 3-month-old girl comes from rural area, seemingly healthy mother.
- Obstetrics history: full-term born by caesarean section due to acute fetal distress, with right weight, APGAR 9/9, interatrial and ventricular septal defect without hemodynamic involvement, no heart block.
- She had been evaluated initially as an infectious skin disease, showing erythematous maculopapular and annular cutaneous lesions from birth on face, scalp, and neck (figures 3-7).
- Despite antibiotic treatment, the lesions were exacerbated and spread to the thorax and limbs.
- According to the characteristic skin involvement, the absence of germs in cultures that were performed, real time- PCR negative for herpes viruses and the immunological profiles of the mother and girl (table 1), the NL was considered.
- Damage skin biopsy: inconclusive

Table 1. Immunological profiles mother and girl case 2

Autoantibodies	Baby	Mother	Reference (by ELISA)
ANA	Positive		Qualitative
Anti DNA (DC)	≤20		0-20 U/ml
Anti SSA/Ro	64	46	0-20 U/ml
Anti SSB/La	43	35	0-20 U/ml

Source: patient's medical file

After treatment with general skin care and steroids (1mg/ kg/ day) the patient has progressed favorably.



Figures 3-7: erythematous and maculopapular cutaneous lesions on face, scalp, and neck, spread to the thorax and limbs, with a favorably evolution after treatment.

Discussion

- (NL) refers to a clinical spectrum of cutaneous, cardiac and others systemic abnormalities, is frequent to observe the skin manifestations only.
- The lesions may be rarely present at birth, but it could, and develops after the first month of life. The cutaneous hallmark of neonatal lupus erythematosus is a superficial inflammatory rash affecting the upper eyelids and the scalp.
- It usually presents as macular annular or elliptic erythema. The eruptions are highly characteristic, however they are sometimes misdiagnosed. Sometimes, some atypical findings persist 12 months after onset of rash in 20% of the cases.
- Histologic examination reveals atrophic epidermis and dermal mononuclear infiltrate.
- The incidence of skin rash is very variable in the different studies, this finding is difficult to explain, but the different UV exposure of the newborns at birth could be an important factor, as already mentioned.
- Further cardiac manifestations sometimes also occur as structural and valvular abnormalities including ventricular septal defect, ostium secundum type atrial septal defect.
- The management depends on the type and the severity of manifestations. Glucocorticoids or intravenous immunoglobulin may also be effective in selected cases.
- Mothers may have connective tissue disease, or may be completely healthy at the time of giving birth. They were also counselled about the increased risk of disease in future pregnancies.
- Slightly less than 5% of infants with NL develop SLE in late adolescence or in early adulthood

Conclusions

- NL is usually difficult to diagnose and should be suspected in a neonate with periorbital erythematous lesions even without a maternal history of collagenosis.
- The cutaneous manifestations were the most frequent in the presented cases, those that did not suffer from cardiac, hematological or hepatic involvement and evolved favorably.
- Prolonged and multidisciplinary follow-up should be carried out for both children with NL and mothers.

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