A one-day-old preterm female child was admitted in Neonatal Intensive Care Unit with cutaneous lesions over face, scalp and back since birth. Baby was having bradycardia at birth. Antenatally on ultrasonography there was fetal pericardial effusion and oligohydramnios. Mother was gravida two, para one with no live births. Her first pregnancy was terminated at 16th week of gestation as the baby was anencephalic. Mother had no other systemic signs and symptoms of any disease, no photosensitivity, no history of genital infections during pregnancy and no history of any drug intake during pregnancy.

Dermatological examination revealed ill defined, erythematous slightly scaly rash around both eyes with telangiectasia and multiple triangular, atrophic scars of variable size distributed over face particularly forehead, scalp and trunk [Figures 1-3].

The neonate’s electrocardiography showed complete atroventricular dissociation and two-dimensional echocardiography showed patent foramen ovale with mild pericardial effusion. Serology of both mother and baby showed ANA and anti-Ro/SS-A positive with ANA titres of 5.20 and 4.60 for mother and child respectively using ANA Blot assay.

**WHAT IS THE DIAGNOSIS?**
ANSWER

Diagnosis- Congenital Lupus Erythematosus.

DISCUSSION

Neonatal lupus erythematosus (NLE) is an uncommon disease, first described by McCuiston and Schoch in 1954.\(^1\) It is caused by the passage of maternal antinuclear antibodies (ANA) and extractable nuclear antigen (ENA) antibodies through the placenta.\(^1\) The ENA are anti-Ro/SSA, anti-La/SSB, and less frequently anti-RNP. Its congenital variant, known as congenital lupus erythematosus (CLE), is very rare. At the moment of diagnosis mothers are asymptomatic in 40-60% of cases. The most common clinical findings include cutaneous lesions and third-degree congenital heart block. Other extracutaneous manifestations include thrombocytopenia, aplastic anemia, transient hepatic involvement and central nervous system vasculopathy. However, multisystem involvement is less common.\(^3-5\) Both mother and the child are at an increased risk of developing some rheumatologic or autoimmune disease in future.\(^3,5-7\)

NLE dermatitis is a transitory benign condition disappearing in approximately seven months coinciding with the disappearance of maternal IgG in fetal circulation.\(^5\) The rash is typically annular or polycyclic, erythematous and scaly. It commonly appears weeks after birth and is distributed usually on sun exposed areas, mainly face, scalp and neck. It typically affects the peri-orbital region giving a spectacle like distribution of skin lesions around the eyes which is characteristic.\(^8\) This is known as "Raccoon" sign. CLE is very unusual and cutaneous lesions can differ from NLE, presenting with atrophy or scarring.

NLE is the main cause of isolated congenital heart block which typically starts during the second or third trimester of gestation, and is responsible for the high mortality (20- 30%) and morbidity in these patients.\(^2\) Later cardiomyopathy can also occur.

Differential diagnoses include intrauterine infections (Congenital syphilis, congenital varicella syndrome, herpes neonatorum), as well as syndromes with cutaneous atrophy or cutaneous aplasia (Goltz syndrome, MIDAS syndrome).\(^2\)

Thus diagnosis of NLE and CLE can be confirmed through history, physical examination and the presence of specific antibodies in fetal and maternal circulation. Skin biopsy is useful and supportive in making diagnosis.\(^5\) Skin biopsy will show interface vacuolar dermatitis with mucin deposition in the dermis and perivascular lymphocytic infiltrate. Direct immunofluorescence will show granular deposits of IgG and C3 at dermoepidermal junction.

The prognosis of children with cutaneous lesions is excellent and the majority of these lesions disappear during the first six months of life, even without treatment. However, photoprotection is almost always recommended.\(^6\) Patients with heart block usually require pacemaker implants.

All pregnant women with anti-SSA/Ro-SSB/La antibodies should have serial fetal echocardiography done by an experienced pediatric cardiologist weekly from 16 to 26 weeks and every other week until about 34 weeks. The initiation of dexamethasone or plasmapheresis can be considered as prophylactic therapy of the high-risk mother (documentation of high titre anti-SSA/Ro and anti-SSB/La antibodies, and a previous child with NLE/CLE).\(^6\)

REFERENCES

1. McCuiston CH, Schoch Jr EP. Possible discoid lupus erythematosus in newborn infant. Arch Dermatol 1954;70:781-5.
2. Diociaiuti A, Paone C, Giraldi L, Paradisi M, El Hachem M. Congenital lupus erythematosus: Case report and review of the literature. Pediatr Dermatol 2005;22:240-2.
3. Carvalho JF, Viana VS, Cruz RB, Bonfá E. Síndrome do lupus neonatal. Rev Bras Reumatol 2005;45:153-60.
4. Peñate Y, Guillermo N, Rodríguez J, Hernández-Machín B, Montenegro T, Afonso JL, et al. Histopathological characteristics of neonatal cutaneous lupus erythematosus: Description of five cases and literature review. J Cutan Pathol 2009;36:660-7.
5. Lynn Cheng C, Galbraith S, Holland K. Congenital lupus erythematosus presenting at birth with widespread erosions, pancytopenia, and subsequent hepatobiliary disease: Case report. Pediatr Dermatol 2010;27:109-11.
6. Lee LA. Transient autoimmunity related to maternal autoantibodies: Neonatal lupus. Autoimmun Rev 2005;4:207-13.
7. Perez MF, Torres ME, Buján MM, Lanoël A, Cervini AB, Pierini AM. Neonatal lupus erythematosus: A report of four cases. An Bras Dermatol 2011;86:347-51.
8. Frey MN, Ioppi AE, Garbin GC, Furian RD, Bau AE. Congenital and neonatal lupus erythematosus: Two case reports. An Bras Dermatol 2012;87:625-8.

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