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Neonatal Lupus Erythematosus

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We describe case report of a 45 days old male baby with neonatal lupus erythematosus, who presented with 3rd degree congenital heart block and depigmented skin lesions on face and upper part of body. Diagnosis of the baby was confirmed by anti nuclear levels and skin biopsy.

Keywords: *Congenital heart block, Neonatal lupus erythematosus* .

Neonatal lupus erythematosus (NLE) is a rare form of lupus erythematosus first described in 1954(1). It is characterized by the presence of cutaneous lesions or congenital heart block or both, in an infant whose mother has connective tissue disease or auto antibodies to extractable nuclear antigens Anti-Ro (SSA), Anti-La (SSB) or ribonucleoproteins (RNP)(2).

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Manuscript received: October 7, 2003;
Initial review completed: November 13, 2003;
Revision accepted: March 18, 2004.

Case Report

A 45-days-old male delivered to a 22-year-old, primigravida by caesarean section at 35 weeks of gestation due to fetal bradycardia. There was no history of fever, rash, lymphadenopathy or drug intake during pregnancy. The natal and postnatal periods were uneventful. At presentation the infant weighed 3.8 kg, with length of 51.5 cm and head circumference 34.2 cm. The heart rate was 76/min, regular and had no pulse deficit. All peripheral pulses were palpable. There were areas of depigmentation (with no erythema or scaling) over the nose, cheeks, around the eyes, scapular region and lower back. There was no significant lymphadenopathy, bleeding spots, rash or hepatosplenomegaly. Laboratory investigations showed Hg of 12g% (PCV was 40%). Total Leukocyte Count was 6800/mm^3, differential leukocyte count was P60, L40, E0, M0 and platelet count of 22,000/mm^3. Liver function tests were within normal limits. Anti-nuclear antibody (ANA) by immunoflorescence was strongly positive (1:640), Anti Ro and Anti La were negative. Electrocardiogram of the baby revealed type 3 congenital heart block The echocardiography showed a structurally normal heart. Skin biopsy revealed scattered lymphocytic infiltration in the dermis with few lymphocytes seen clustered around dermal appendages features compatible with NLE. Immunoflorescence studies could not be done. ANA was positive at 1:640 with speckled pattern in mother's serum. Anti-Ro

was present but there were no detectable Anti-La or Smith (Sm) antibodies.

Discussion

Neonatal lupus erythematosus (NLE) is characterized by the presence of cutaneous lesions, congenital heart block or both in about 10% cases(3). The reasons why some babies develop skin disease, while others develop heart disease are not clearly known. The incidence of NLE is not known. Although genetically determined autoantibody production occurs in the mother, tissue injury may depend on other factors which are not very clear at present(3). More than 90% infants will have cutaneous lesions, of which 70% are present at birth and the remainder usually develops within 2 months. These lesions are usually transient and last for a few weeks to months(2). Cutaneous lesions characteristically are erythematosus papules, annular discoid or polycyclic plaques with or without fine scaling. NLE lesions are frequently found on the scalp and face, however, unusual presentation with congenital skin atrophy, erosions and alopecia have also been rarely described. On occasion lesions are concentrated in the periorbital or malar region(3,4). The skin biopsy in lupus erythematosus may reveal epidermal atrophy, liquefaction, degeneration of basal keratinocytes, colloid bodies and a perivascular and periappendageal lymphohistocytic infiltrate in the dermis(5).

Amongst the cardiac abnormalities about 50% are conduction defects. In mid to late fetal development anti-Ro antibodies can bind to cardiac conducting cells, alter membrane repolarization and selectively damage AV node(6). Heart block can be detected as early as 16 week of gestation by USG or fetal electrocardiography(3). Other cardiac problems include subendocardial fibroelastosis, fibrinous pericarditis, PDA(6). Other systemic manifestations include hemolytic anemia, thrombocytopenia, liver, lung and CNS involvement(3,6).

Maternal auto antibodies directed against tissue antigens of skin, heart, liver, bowel, lung and blood cells are present in infants with NLE. The IgG class are most frequent (95%) and are directed against the Ro ribonucleoproteins antigen(6).

Antenatal therapy is generally not required. Serial plasmapheresis and systemic steroids have been tried with variable success(3). Systemic steroids may occasionally be required for associated thrombocytopenia or hemolytic anemia. Treatment of heart disease is not always necessary. For children with heart failure due to slow heart rate, pacemaker implantation is the treatment

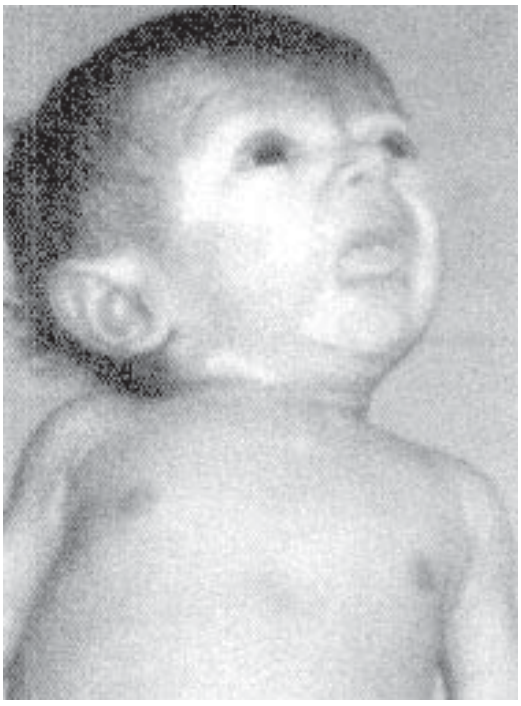

Fig. 1.Photograph showing depigmented macular patches on the face.

of choice. If heart failure persists even after pacemaker implantation and children who have serious internal systemic manifestations, may be treated with systemic steroid(2,3). As many as 8.3% cases of NLE may progress to systemic lupus erythematosus (SLE) in later childhood(6).

Acknowledgement

We thank Dr. Sharmila Mishra, Department of Pathology, ICMR, Delhi for reviewing the skin biopsy.

Contributors: NA, PP worked up the case, reviewed the literature and drafted the article. RM planned the management; PP will act as the guarantor.

Funding: None

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Psychogenic Non-Epileptic Seizures

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Psychogenic non-epileptic seizures (PNES) need to be differentiated from epileptic seizures as the management varies for both. Presence of tongue biting, falling and urinary incontinence favors a diagnosis of epileptic seizures. We report two children with features suggestive of epileptic seizures that were eventually diagnosed as PNES with the help of video-EEG monitoring.

Key words: *Epilepsy, Psychogenic non-epileptic seizures.*

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Manuscript received: January 21, 2004;
Initial review completed: February 27, 2004.
Revision accepted: March 24, 2004.

Psychogenic non-epileptic seizures (PNES) are usually differentiated from epileptic seizures on the basis of absence of tongue biting, falling, incontinence, post-ictal phenomena and concomitant abnormalities on the electroencephalogram (EEG)(1,2). It is important to make an early diagnosis of PNES as a delay in diagnosis leads to overtreatment including polytherapy, repeated hospitalization, poor response to treatment, and mechanical ventilation(3,4). However, PNES are frequently misdiagnosed as epileptic seizures and delays in diagnosis are