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Case Report

Seizures as an unusual presentation of neonatal lupus erythematosus: a case report

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ABSTRACT

Neonatal lupus erythematosus is an uncommon passive autoimmune disease manifesting as cutaneous lupus lesions and/or congenital heart blocks with rare CNS involvement. A 2months old female infant admitted for respiratory illness was found to have neonatal lupus skin lesions, developed seizures during hospital stay without any metabolic cause or CNS infection. Mother had significantly raised anti-Ro/SSA and anti-La/SSB antibodies. As per classical description, the cutaneous lesions started to fade but the CNS involvement needs follow up. NLE should be suspected on the basis of cutaneous lesions even in absence of heart block and seizures may also rarely manifest.

Keywords: NLE, Hypopigmented-hyperpigmented skin lesion, Seizures.

INTRODUCTION

NLE is an uncommon passive autoimmune disease in which there is transplacental passage of anti-Ro/SSA and anti-La/SSB maternal antibodies.[1-4] Common clinical manifestation includes cutaneous lupus lesions and/or cardiac disease specially congenital heart blocks, in some cases hepatobiliary and haematological involvement but CNS involvement is rare.[5-7]

CASE REPORT

A two month old female infant who was a product of consanguineous marriage, second in birth order, born at term by vaginal delivery at the hospital without any complication and had satisfactory post natal period, was admitted in our hospital with fever and cough for 6 days and rapid breathing for 1day. She was taking breastfeeding and had no noisy breathing, sluggish activity or any abnormal movements. Vital parameters on admission were: temperature 100.1 F, HR 126/min, RR 62/min and SPO2 94% at room air. On general physical examination baby had hypopigmented patches over scalp and forehead which were present since birth and were red to begin with. There were no greasy or scaly lesions suggestive of seborrheic dermatitis and no history of applying topical medication. Her maternal grandmother was diagnosed to have SLE at 30 years age for which she took treatment but no records were available. Mother had no manifestations suggestive of SLE during or before pregnancy and her VDRL and HIV were negative. On respiratory system examination beside tachypnea patient had b/l crackles but no significant intercostal/sternal recession. We started IV cefotaxime and amikacin for pneumonia along with paracetamol for fever. The CVS and CNS examination were unremarkable. The next day she had two episodes of seizures in form of staring look, paddling movements of lower limbs and unresponsiveness to voice. Each episode lasted for four to five minutes with ten hours interval between two episodes. At time of seizures there was no fever, bouts of cough, choking spell and her blood glucose was 104 mg/dl. There was mild microcytic hypochromic anemia (Hb 9.7gm/dl). Samples were taken for serum calcium, sodium and potassium which came normal. IV phenobarbitone was given to control the convulsions. LP was done to rule out meningitis which showed normal CSF. Her TORCH titres were negative and MRI brain

was also normal. ECG and ECHO done to rule out any associated congenital heart block/dysfunction were normal. EEG done on third day of convulsions was normal, though she was on maintenance dose of phenobarbitone during that time. Opinion of skin specialist was sought who suggested neonatal lupus. So, ANA of mother and baby were sent which showed significantly raised activity index in both (4.26 and 3.71 respectively). To confirm NLE Anti-Ro/SSA, anti-La/SSB antibodies of mother and baby were done by ELISA technique, these antibodies were significantly raised in mother (172 IU/ml and 109 IU/ml respectively). The patient's Anti-Ro/SSA antibodies were on the higher side of normal range (29 IU/ml) where as her anti-La/SSB antibodies were in normal range. Topical application of liquid paraffin was started for skin lesions and baby was discharged on seventh day. After two months review skin lesions had started to fade and she had no seizures thereafter.

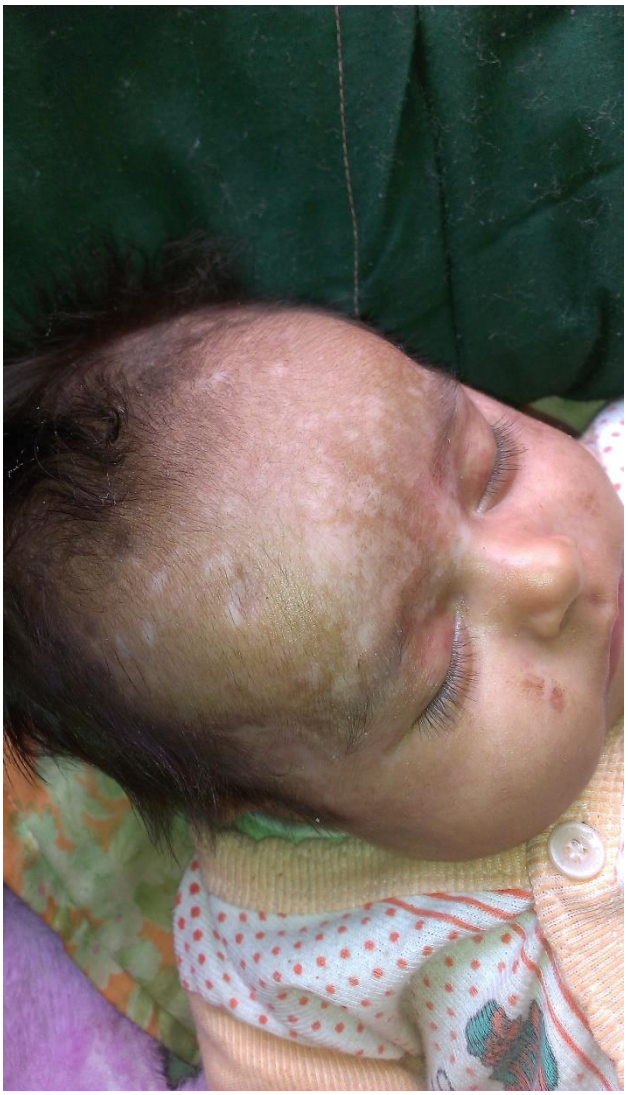

Figure 1: Skin lesions of NLE.

DISCUSSION

The incidence of NLE is about 1 in 20,000 live births and has been reported in a number of different ethnic groups[1], and is a distinct syndrome unlike lupus in children and adults. The overwhelming majority of NLE infants are born to mothers who have anti-Ro(52kd and 60 kd/SSA) antibodies and approximately 40% of mothers will also have anti-LA/SSB antibodies.[2,5-7] About half of the new-born infants with NLE demonstrate isolated congenital heart block, 40% have cutaneous lesion and 10% have both.[5-7]In addition to these major features, about 10% of NLE infants also have jaundice due to neonatal cholestatic hepatitis and approximately 4% have hematologic features, such as thrombocytopenia or aplastic anemia. Clinically the cutaneous lesions may be present at birth, but most babies have the onset in the first few weeks after birth. The characteristic skin lesions are multiple rounds, pink to red macules involving the scalp, face and the extremities. Some skin lesions partially clear in the centre giving an annular configuration that mimics tinea. Skin lesion generally clear without scarring but in dark infants a transient hypopigmentation or hyperpigmentation has been reported at the site of lesions. [2] Our patient had hypopigmented lesions which have begun to fade indicating its transient nature.

The earliest report of apparent CNS involvement in NLE was a myelopathy with an abnormal gait noted by Kaye and Butler in 1987.[8] Though the overall CNS involvement has been reported in 1.4% cases of NLE, there are only few case reports of seizures in the early infancy.[8-11] Mothers of NLE infants may or may not show features of connective tissue diseases at the time of birth of the affected infants.[5-7] In at least half of the mothers symptoms are entirely absent or subtle which are missed on routine maternal history and physical examination. In our patient also mother had no apparent manifestation of SLE.

Our diagnosis of NLE in this patient was based on skin lesions, history suggestive of SLE in the family and presence of significantly raised ANA activity index in baby and mother which was confirmed by the serological markers specific for NLE (Anti-Ro/SSA, anti-La/SSB antibodies) in mother. In the infant these antibodies are passively transferred and decrease in due course of time, thereby there values in this patient were not significantly raised. Only few cases of seizures are reported in NLE albeit due to family history, sero-positivity in mother and skin features suggestive of it, we attributed the seizures to NLE. Also, no other possible cause of seizures was found.

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