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CASE REPORT

Neonatal lupus erythematosus in a Nigerian infant

Ezeudu CE
Ezejiofor OI
Igwe WC
Ezechukwu CC
Nwankwo HM
Ezejiofor IF

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Ezeudu CE (✉)
Igwe WC, Ezechukwu CC
Department of Paediatrics,

Ezejiofor OI
Department of Medicine
Nnamdi Azikiwe University
Awka, Anambra state, Nigeria
Email: ezeuducj@yahoo.com

Nwankwo HM
Department of Medicine

Ezejiofor IF
Department of Histopathology,
Nnamdi Azikiwe University
Teaching Hospital, Nnewi,
Anambra state, Nigeria

Abstract: Systemic lupus erythematosus is a chronic inflammatory autoimmune disease that affects all organ systems and follows a relapsing and remitting course with presentation ranging from indolent to fulminant. It is an uncommon condition in children and usually manifests as the cutaneous form in the paediatric age group. We report a case of neonatal lupus erythematosus in a six-week-old Nigerian infant seen at NAUTH, Nnewi. The objective of this report is to highlight the existence of this very rare condition in infants. A 6-week-old female infant presented at the paediatric unit of our facility with a history of a pre-auricular skin eruptions of five weeks and poor suckling of three weeks duration. The skin lesions persisted unchanged after 14 days of antibiotics. A skin snip was taken for histology and a diagnosis of neonatal lupus erythematosus was made.

Key words: neonatal lupsus erythematosus, infant, Nnewi.

Introduction

Systemic lupus erythematosus is a chronic inflammatory autoimmune disease that affects all organ systems and follows a relapsing and remitting course with presentation ranging from indolent to fulminant. It is an uncommon condition in children and usually manifests as the cutaneous form in paediatric age group.[1]The exact pathogenesis of neonatal lupus erythematosus (NLE) is not well understood, but it has been attributed to a lot of factors, mainly genetic and environmental. Some drug induced cases have been reported.[2.3]Immunoglobulin G (IgG) against R_o (SSA), La(SSB) and/or U1- ribonucleoprotein produced by the mother are passively transferred transplacentally, predisposing the infant to the development of neonatal lupus erythematosus. Inspite of this, only about 2% of babies with positive maternal transplacental antibodies develop neonatal lupus erythematosus.

The major clinical manifestations of neonatal lupus erythematosus are dermatologic, cardiac and hepatic. Some infants may also manifestcentral nervous system, and haematologic abnormalities. Conduction cardiac abnormalities, hepatomagaly, seizures, anaemia and thrombocytopenia can all occur. Cutaneous manifestations of NLE occur in about 70% of patients with neonatal lupus erythematosus and lesions are seen predominantly in the face, neck, and scalp.[4]These lesions appear typically as erythematous plaques that may persists as alopecia over a period of time. We report a case of neonatal lupus erythematosis in a six- week-old female infant seen at the paediatric unit of our facility.

Case Report

OM is a six-week-old female infant who presented at the paediatric unit of Nnamdi Azikiwe University Teaching Hospital, Nnewi with a rash of five weeks and poor suckling of three weeks duration. She was well until the second week of life when she developed skin eruptions around the left ear but later spread to involve the left parietal area. The lesions started as small vesicles containing clear fluid, ruptured spontaneously, and then ulcerated. (Figure 1).Similar lesions also developed on the occipital region of the scalp. Two weeks into the illness she was noticed to be suckling poorly and to be losing weight. She was given some drugs purchased from a patent medicine dealer and also received treatment from a private hospital with no significant improvement. She was then brought to our facility for further management.

She was the first of a set for twins delivered at term to a thirty-one-year old primiparous woman. Her pregnancy

and delivery were normal and there was no significant illness in the early neonatal period. She weighed 3kg at birth.

Physical examination revealed a weak infant with a normal body temperature, mildly pale, anicteric with no sign of dehydration. She weighed 2.5kg with a head circumference of36.5cm. She was semiconscious and globally hypertonic. There were erythematous, ulcerative lesions around the left ear extending to the parietal area with a significant hair loss on the affected areas. The occipital area also has similar lesion but with less ulceration. Abdomen was full and soft and moved with respiration. Liver was palpable 2cm below the right coastal margin in the mid-clavicular line. Other findings on systemic examinations were essentially normal. Complete blood count showed marked neutrophil leukocytocysis with toxic granulations. Erythrocyte sedimentation rate was 85mm/hr.

Diagnosis of failure to thrive due to poor nutritional intake and sepsis were made. She was placed on standard treatment for sepsis and nutritional rehabilitation.

Two weeks into admission the lesionspersisted with some scarring leaving some areas of hypopigmentation/ hyperpigmentation and alopecia. The dermatologist was invited after alopecia had persisted for several weeks. The dermatological review found multiple erythematous, hypopigmented/hyperpigmented atrophic plaques on the left ear, left parietal and occipital areas, and raised a suspicion of lupus erythematosis.

Biopsy and histology of the lesion showed hyperkeratotosis with follicular plugging. There was dermoepidermal mononuclear inflammatory cell infiltration extending to the adnexal structures and subcutaneous tissues. There was basal layer vacuolation with melanin incontinence and few colloidal bodies. (Fig 2). ***The overall picture was consistent with a diagnosis of neonatal lupus erythematosus.***

The patient showed marked improvement and was discharged home after three months of conservative management. She was however, subsequently lost to follow-up.

Fig 1: Ulceration involving the left pinna and the parietal scalp with loss of hair, marked erythema, patches of hypopigmentation and hyperpigmentation

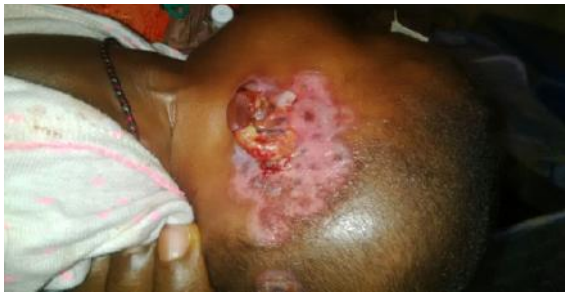
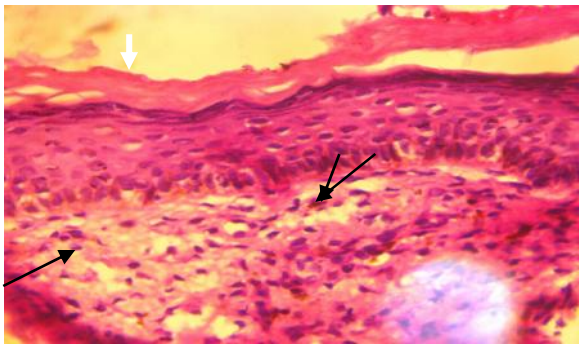


Fig 2: Photomicrograph of Neonatal lupus erythematosus showing hyperkeratosis (white arrow) and moderate mononuclear inflammatory infiltrate in the upper dermis associated with dermal oedema (an arrow). There is also focal basal cell vacuolization and melanin pigment incontinence (double arrows).



Discussion

Neonatal Lupus erythromatosus is usually seen in infants born to mothers with auto- antibodies against R_o (SSA), La(SSB) and/or U1- ribonucleoprotein. These auto- antibodies gain access to the fetal circulation through the placenta. The circulating auto-antibodies produce signs and symptoms in the infant that are similar to adult Lupus erythematosus. The mothers of these infants with NLE are usually diagnosed of systemic lupus erythematosus (SLE) or other autoimmune conditions such as Rheumathoid arthritis or Sjogren's syndrome. The commonest abnormalities seen in the newborn babies with NLE are dermatological, cardiac, and occasionally other organs such as the liver may be involved. The disease is rare, affecting only approximately 1 in 20,000 live born infants.[5]

Such dermatological abnormalities include the cutaneous manifestations of lupus erythematosus seen in adults such as; Chronic Discoid Lupus, Subacute Lupus, and Acute Lupus rashes. Moretti *et al*[6] reported a two month old infant with such dermatological lesions on the left cheek, scalp, retroauricular area and abdominal skin. These rashes were present in the index patient in the head and neck region which are usually common sites because of their photosensitive nature. The formation of multiple atrophic scars and dyspigmentation on the scalp with scarring alopecia was very much in keeping with the clinical appearance of chronic discoid lupus erythematosus. These characteristic skin lesions with other clinical features such as poor suckling, failure to gain weight and generalized hypertonia strengthened the clinical suspicion of Neonatal lupus erythematosus.

The elevated Erythrocyte sedimentation rate (ESR) of 85mm/hr and skin biopsy histology findings consistent with chronic discoid lupus erythematosus were all supportive of the diagnosis of NLE. The absence of clinical features suggestive of any auto-immune condition in the mother does not exclude the diagnosis.[7] In all such cases, it will be very informative to screen the mother

for evidence of circulating auto-antibodies. This was however not possible in the index case because of financial constraints and unavailability of the auto-antibodies screening tests in our centre.

The exact pathogenesis of the condition is not very clear but the transplacental passage of auto-antibodies such as anti-Rho (SSA) antibody which is present in 95% of cases of Neonatal lupus is fundamental.[8,9] The significant improvement made before discharge on conservative management is likely because of the disappearance of the circulating antibodies within several months of management.

Conclusion

Lupus erythematosus is rare in paediatric age group but should be suspected in infants with persistent skin lesions and alopecia around the head and neck region.

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