

Case Reports

Systemic Lupus Erythematosus in an Infant

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Systemic lupus erythematosus (SLE) in young children is a rare occurrence(1,2). Although reported to have occurred in a 3-month old infant(3), onset of SLE is extremely unusual in a child younger than 4 years of age(1,4). We report a case of SLE in a male child, whose disease started at the age of 5 months. This, to the best of our knowledge, is the youngest patient to have presented with characteristic cutaneous manifestations of SLE.

Case Report

A 16-month old male child presented with a history of erythema and swelling of the cheeks, bridge of the nose and forehead since the age of 5 months. Parents noticed an exacerbation of the erythema and swelling within 10-15 minutes of exposure to the sun. Two months later, the child started developing vesiculo-bullous lesions and erythematous scaly crusted

papules and plaques on the face, scalp and extremities. Along with the skin lesions, the child also developed painful oral ulcers which caused considerable difficulty in feeding. A history of continuous low grade fever was present since last 10 months but during episodes of exacerbation of skin and oral lesions, the patient developed high grade fever. The child developed diffuse hair loss as well as had patchy alopecia at the sites of healed skin lesions since 10 months. There was no history of arthritis, Raynaud's phenomenon, seizures, pneumonitis or vasculitis.

The child was born full term after a normal delivery to consanguineous parents (first cousins). Delivery was conducted at home and there was no history of any perinatal complications. There was no history suggestive of SLE in the mother. An elder sibling who was 7 years of age had typical lesions of discoid lupus erythematosus since the age of 1 year. She also had severe painful oral ulcers, but did not complain of any constitutional symptoms and had no systemic involvement. Skin biopsy from the lesion in the sibling showed hyperkeratosis with follicular plugs and moderate upper dermal chronic inflammation. Immunofluorescence studies showed deposition of IgG at basement membrane zone.

General physical examination revealed generalized lymphadenopathy and moderate hepatosplenomegaly. Scaly erythematous plaques on background of bright red erythema were present on the cheeks, bridge of the nose, forehead, scalp and pinna of the ears (*Fig. 1*). Some of the plaques were exudative and crusted. The patient also had small oval, well defined plaques with central atrophy and depigmentation inside the concha of the ears. Discrete as well as confluent scaly oval and annular plaques with central

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Fig.1. Photograph of the patient showing malar rash and discoid lesions on forehead.

atrophy and follicular plugs were present on the trunk and extremities. Erythematous, blanchable, telangiectatic oval plaques were present on palms and soles.

A complete haemogram revealed mild anemia (Hb 11.3 g/dl) while the total leucocyte count and platelet count were within normal limits. The erythrocyte sedimentation rate was 30 mm in first hour. His renal and liver function tests were within normal limits. A routine chest X-ray and electrocardiogram revealed no abnormality. Serology for antinuclear antibody (ANA) was negative and VDRL test was non-reactive. Fundus examination was within normal limits. Histopathological examination of discoid lesion on the face showed an atrophic epidermis with upper dermal chronic inflammation. Immunofluorescence for immunoreactants revealed IgM at basement membrane and capillary walls of the exposed skin and C₃ at basement membrane zone of the unexposed skin.

Discussion

The prevalence of SLE in children has been estimated at 0.6 cases per 100,000(1,2). In children, adolescent females are predominantly affected with the peak age of onset being 12 years(1,3). This female preponderance in teenagers and adults is less evident in the first decade of life where females exceed males by only 3 to 4:1(1,3). However, onset of SLE before the age of 4 years is a rare occurrence (1,4).

None of the 59 cases of childhood SLE who attended Rheumatic Care Center in a General Hospital in Madras between 1972 and 1993 had disease onset before 5 years of age(4). Our patient developed first symptoms of SLE at the age of 5 months. Cumming *et al.*(3) reported a 3-month old premature infant with SLE; this child underwent phototherapy for hyperbilirubinemia, was on artificial ventilation and had multiple surgical interventions and later developed *Escherichia coli* septicemia. Moreover, the diagnosis of SLE was based on laboratory, immunologic and serologic criteria and authors stressed the importance of suspecting SLE in infants with multisystem involvement. Our patient is perhaps the youngest case to have presented with typical skin and oral lesions of SLE.

Lupus like syndromes caused by transplacental passage of maternal IgG antinuclear antibodies are a well known phenomenon(5). Usually these infants present with malar erythematous, annular lesions along with congenital heart block. But these lesions are seen at birth or within the first 2 months of life. Our patient had characteristic malar rash, photosensitivity, discoid lupus-like rash and oral ulcers, a

presentation typical of SLE in older children and adults. Familial cases of SLE have been reported (6). In our case, occurrence of discoid lupus erythematosus in another sibling is an interesting feature. Consanguinity in parents and absence of disease in any of the preceding generations suggests autosomal recessive inheritance pattern.

It is generally stated that children have more severe renal involvement than adults and hepatosplenomegaly and lymphadenopathy are more characteristic of childhood SLE (1). Hepatosplenomegaly and lymphadenopathy were present in our case but there was no renal involvement. ANA's are present in more than 95% of the children with SLE. The absence of ANA in our case is difficult to explain. A minority of patients with SLE like disease, *i.e.*, subacute lupus erythematosus are ANA negative but usually have anti-Ro antibodies(1). We could not test for anti-Ro antibodies due to non availability of the

facility.

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