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Case Report

Childhood-onset systemic lupus erythematosus – A case report

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Abstract Systemic lupus erythematosus (SLE) is autoimmune multisystem disease associated with various clinical manifestations. Childhood-onset SLE (cSLE) is extremely rare and comprises only 15-20% of lupus erythematosus cases. Most of the children belong to the adolescent group while very few are in the prepubertal age. We herein, report a case of 7-year-old female child diagnosed as systemic lupus erythematosus due to its rare occurrence in pediatric age group.

Key words
Systemic lupus erythematosus, childhood-onset SLE

Introduction

Systemic lupus erythematosus (SLE) is the prototype of systemic autoimmune diseases characterized by the production of autoantibodies and immune complexes leading to protean systemic manifestations.[1] The occurrence of SLE in children is very rare with an incidence about 3-4 per 1,00,000 children.[2,3] The disease is said to be more prevalent in Asian, African-American and Hispanic children.[4,5] The onset of childhood SLE occurs between the ages 3 and 15, with the girls outnumbering boys in the ratio 4:1. The clinical manifestations of childhood-onset SLE (cSLE) are diverse, severe and often atypical as compared to the adults.[5]

Case report

A 7-year-old female child born of non-consanguineous marriage was brought to our skin OPD with the complaint of erythema and rashes present on the face; more marked over her cheeks, bridge of the nose and the forehead since 6 months (**Figure 1**). Gradually, she developed vesicobullous lesions and erythematous scaly-crusted lesions on scalp and around perioral areas, which healed with hypopigmentation. Similar lesions were present over the genitals and the extremities (**Figure 2**). Along with the skin lesions, the child also developed painful oral ulcers, which caused considerable difficulty in eating. There was no similar complaint in the family or any episode of gastrointestinal upset for the past 6 months. But there was a significant history of worsening of facial skin lesions during sun-exposure and discoloration of both hands and feet on prolonged cold exposure. She later developed low-grade fever, which was not associated with chills or rigor along with joint pain over large joints (knee and ankle) since 1½ month. She had undergone some local treatment but was not relieved.

On general examination, the child was thin-built and underweight (18 kg). Generalized pallor and

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cervical lymphadenopathy were present. Her developmental milestones were normal.

Figure 1 Diffuse erythema and scaly lesions (malar rash/butterfly rash) with postinflammatory hypopigmented areas in perioral regions.

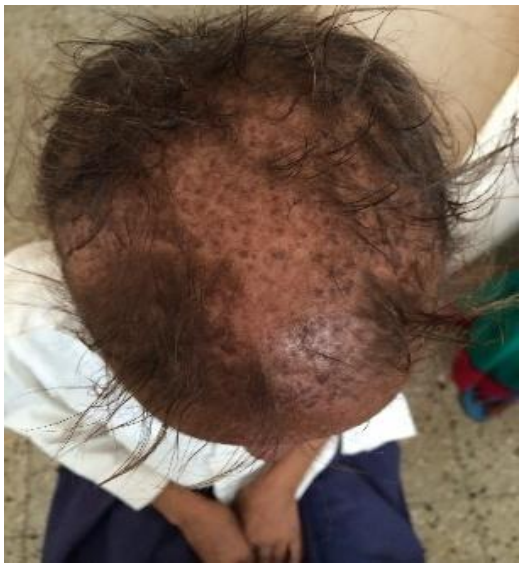

Figure 2 Lupus hair with scarring alopecia.

Cutaneous examination showed diffuse hair loss (lupus hair) and at some places patchy alopecia with few hyperpigmented scaly lesions were seen. Erythematous rash was seen all over the face with (malar rash/butterfly rash) involving both cheeks and bridge of the nose and forehead. Perioral regions showed post inflammatory hypopigmentation marks. Photosensitivity and Raynaud's phenomenon was present. Vasculitic lesions in the form of erythematous macules and

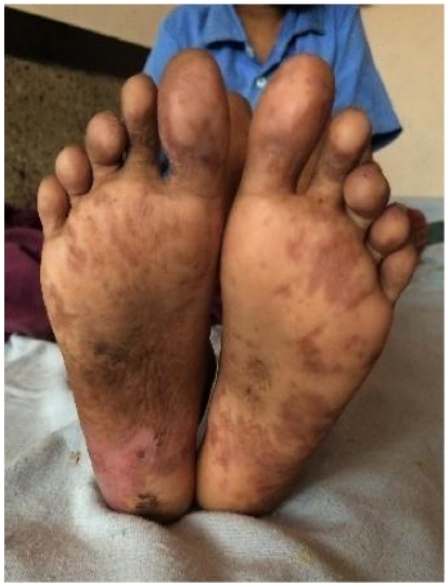

Figure 3 Vasculitic lesions in the sole of both foot with some areas of hypopigmentation.

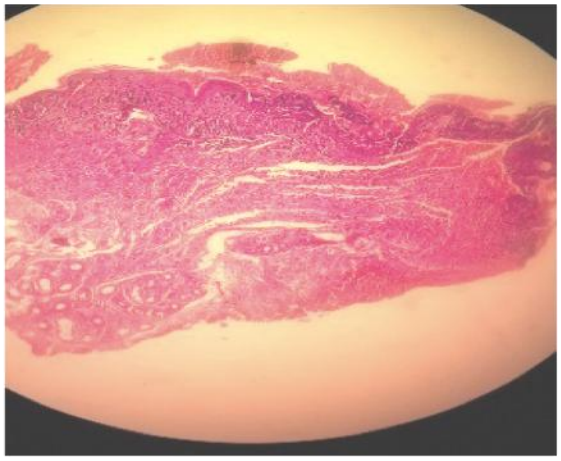

Figure 4 Superficial perivascular and interstitial infiltrate composed of neutrophils and lymphocytes, basal layer vacuolization and interface dermatitis with abundant mucin in dermis (H and E 10X).

patches were seen on the both hands and foot (**Figure 3**).

Laboratory investigations revealed WBC = 5,800 cells/mm3 (58% neutrophils, 28% lymphocytes), hemoglobin = 7.9 g/dL, platelets = 1,67,000/mm3, erythrocyte sedimentation rate

(ESR) = 30 mm/h; specific tests – ANA test was positive with ANA titer = 1:520 though anti- double stranded DNA (dsDNA) was negative. Immunofluorescence study could not be done due to lack of availability in our centre. Liver and renal function tests and urinalysis were within normal limits. Chest X-ray and electrocardiography revealed no abnormality. Ophthalmologic tests were also normal. Systemic examinations were within normal limits. The histopathological examination of skin biopsy specimen showed sparse superficial perivascular and interstitial infiltrate predominantly of neutrophils and lymphocytes more seen on the papillary dermis and also at all levels of epidermis along with mild spongiosis. Basal layer showed vacuolization and interface infiltration by neutrophils. Reticular dermis showed abundant mucin (**Figure 4**).

On the basis of clinical presentation, laboratory findings and histopathology, diagnosis of childhood-onset SLE was done. In treatment plan, proper counseling was done to the child's parents with strict avoidance of sun exposure and cold exposure. Blood transfusions were given to the patient to overcome anemia. As for specific treatment, a broad spectrum sunscreen lotion, emollients along with oral corticosteroid (prednisolone) 1 mg/kg/d, hydroxychloroquine (6.5mg/kg/d) and vitamin D supplements were further added to the patient.

Discussion

Childhood-onset systemic lupus erythematosus (cSLE) is one of the most common systemic autoimmune disease in children. In children, adolescent females are predominantly affected with the peak age of onset being 12 years; although lupus is uncommon before 10 years of age.[6,7]

The exact etiology is unknown but the interaction between immune complexes, autoantibodies, genetic, drugs and environmental factors do play a significant role in causing inflammation and eventually damage to the organs and systems.[8]

cSLE patients have a less favourable prognosis as compared with adult counterparts resulting in two to three times higher mortality. The clinical presentation of cSLE is frequently more severe than adult onset SLE with multiple organ involvement, particularly the kidney and central nervous system.[9,10]

Janwityanujit *et al.* and Font *et al*. suggested that cutaneous changes (photosensitivity, malar rash, Raynaud phenomenon, vasculitic lesions) and nephritis is more common in cSLE.[11,12,13] In our case, skin findings were similar; although there was no renal involvement. Neuropsychiatric manifestations have been reported in 29-44 percent of pediatric patients with SLE[14] but in our case, there were no neurological or psychiatric findings. The most common neurologic symptoms in children include headaches, coma, psychosis and depression.

The diagnosis can be confirmed by histopathology and serology. Serology showed high titre of ANA though Anti-dsDNA was absent in our case. Anti-ds DNA antibodies are highly specific for SLE, and are present in about 61-93% children with active disease, especially active nephritis. However, they may be absent in about 40% children with active lupus, especially if nephritis is not present.[15] We could not do immunofluorescence study due to non availability in our centre.

The disease severity varies from mild to severe, and requires long-term and often aggressive treatment. Strict avoidance to sun is to be advised the patients with use of broad-spectrum

sunscreens. Corticosteroids (1-3mg/kg/d) and hydroxychloroquine (4-6mg/kg/d) have shown excellent results in control of disease. Other options include azathioprine (0.5-2.5 mg/kg/d), cyclophosphamide (0.5-2.5 mg/kg/d), intravenous immunoglobulins 2 g per kg per dose and plasmapheresis.

To conclude, childhood SLE is a challenging disease both difficult to diagnose and to treat. It is less often observed in children than adults. The clinicians should be aware of the greater risk of systemic complications in children with systemic lupus erythematosus. Henceforth, pediatric SLE patients should be continually followed up and appropriate therapy should be initiated depending upon the disease activity to reduce morbidity and mortality.

References

1. Cassidy JT. SLE, Juvenile dermatomyositis, Scleroderma, and Vasculitis. In: Kelly WN, Harris ED, Ruddy S, Sledge CB, editors. *Textbook of Rheumatology, 4th edn*. Philadelphia, WB Saunders; 1993. P. 1224-47.
2. Ali VS, Dalvi AS, Merchant RH, Mehta KP, Chadlani AT, Badakere SS *et al*. Systemic lupus erythematosus in Indian children. *Indian J Pediatr*. 1989;**26**:868-73.
3. Chandrashekharan AN, Rajendra CP, Ramakrishnan S, Madhavan R, Pratibhan M. Childhood systemic lupus erythematosus in south India. *Indian J Pediatr*. 1994;**61**:223-9.
4. Lehman TJ, McCurdy D, Spencer C. Prognostic value of antibodies to Ro/SSA, SSB/La, and RNP in children with systemic lupus erythematosus (abstract). *Arthritis Rheum*. 1990;**33**(Suppl):S154.
5. Segel M, Lee SL. The epidemiology of systemic lupus erythematosus. *Semin Arthritis Rheum*. 1973;**3**:154.
6. Tucker LB, Menon S, Schaller JG, Isenberg DA. Adult- and childhood-onset systemic lupus erythematosus: a comparison of onset, clinical features, serology, and outcome. *Br J Rheumatol.* 1994;34:866-72.
7. Jimenez S, Cervera R, Font J, Ingelmo M. The epidemiology of systemic lupus erythematosus. *Clin Rev Allergy Immunol*. 2003;**25**:3-12.
8. Von Feldt JM. Systemic lupus erythematosus. Recognizing its various presentations. *Postgrad Med.* 1995;**97**: 79, 83, 86.
9. Pande J, Sekharan NG, Kailash S, Uppal SS, Singh RR, Kumar A *et al.* Analysis of clinical and laboratory profile in Indian childhood systemic lupus erythematosus and its comparison with SLE in adults. *Lupus*. 1993;**2**:83-7.
10. Tucker LB, Menon S, Schaller JG, Isenberg DA. Adult- and childhood-onset systemic lupus erythematosus. A comparison of onset, clinical features, serology and outcome. *Br J Rheumatol*. 1995;**34**:866-72.
11. Janwityanujit S, Totemchokchyakarn K, Verasertniyom O, Vanichapuntu M, Vatanasuk M. Age related differences on clinical and immunological manifestations of SLE. *Asian Pac J Allergy Immunol.* 1995;**13**:145-9.
12. Font J, Cervera R, Espinosa G, Pallarés L, Ramos-Casals M, Jiménez S *et al.* Systemic lupus erythematosus (SLE) in childhood: analysis of clinical and immunological in 35 patients and comparison with SLE characteristics in adults. *Ann Rheum Dis*. 1998;**57**:456-9.
13. Livingston B, Bonner A, Pope J. Differences in clinical manifestations between childhood-onset lupus and adult-onset lupus: a meta-analysis. *Lupus*. 2011;**15**:1345-55. doi: 10.1177/0961203311416694.
14. Quintero-Del-Rio Al, Van Miller. Neurologic symptoms in children with systemic lupus erythematosus. *J Child Neurol*. 2000;**15**:803-7.
15. Carreno L, Lopez-Longo FJ, Monteagudo I *et al.* Immunological and clinical differences between juvenile and adult onset systemic lupus erythematosus. *Lupus.* 1999;**8**:287-92.