Case report:

Juvenile systemic lupus erythematosus: A rare case report

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Abstract

Juvenile systemic lupus erythematosus (JSLE) is defined as systemic lupus erythematosus (SLE) with onset before 18 years of age having a prevalence of 15%. JSLE is associated with an increased incidence of arthritis, nephritis, hematologic and neurologic manifestations as compared to adult-onset disease. We report a case of 12 years female having mucocutaneous lesions with photosensitivity, joint pain, diffuse alopecia of the scalp and pancytopenia. According to Systemic Lupus International Collaborating Clinics (SLICC) group classification criteria 2012, a diagnosis of JSLE was made. The patient was treated with 5 mg/kg/day hydroxychloroquine, 0.5 mg/kg per day of oral prednisone, topical steroid and sunscreen for six weeks following which the patient improved.

Key words: Juvenile, SLE, alopecia, glucocorticoids

Introduction:

SLE occurring before 18 years of age is defined as Juvenile SLE (JSLE) and has a prevalence of 15%.[1] JSLE is an aggressive multisystem autoimmune disease having lower life expectancy as compared to an adult.[2] The annual incidence of JSLE is estimated to be 0.3–0.9/100,000 and is generally lower in Caucasian children.[1] As JSLE presents having diverse clinical features, its early diagnosis is challenging. In this case, we made an early diagnosis and treated the patient with low dose glucocorticoid and hydroxychloroquine for six weeks which lead to a reversal of the cutaneous lesion. Early diagnosis and treatment can protect from adverse future outcomes, especially in young females.

Case report:

12 years female presented in our outpatient department with a history of scarring skin lesions over the lower back, bilateral upper limbs and both palms for 3 months. It was associated with burning sensation over face on exposure to sunlight, oral mucosal lesions over hard palate with scarring lesions over the both palms (Figure 1) and diffuse alopecia of the scalp.

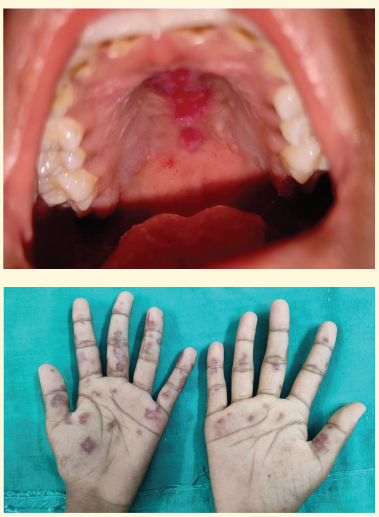

Figure 1 Oral mucosal lesion with scarring lesions of both palms.

She gave history of fever, myalgia, and polyarthritis of small joints of both hands. There was no history of epileptic fits, chest pain, breathlessness, abdominal pain, difficulty in eating, Raynaud's phenomenon and drug intake prior to appearance of the lesion. Laboratory investigations revealed pancytopenia,

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elevated erythrocyte sedimentation rate (ESR), and normal liver and renal function test. Urine examination showed albuminuria without hematuria. Antinuclear antibody (ANA) titers were strongly positive. Anti-dsDNA was normal. Skin biopsy taken from the back showed thinning of the epidermis with loss of rete ridges and vacuolization of basal cell layer. There was sparse lymphohistiocytic infiltrate present focally at the dermo-epidermal junction with perivascular infiltrate in the dermis (Figure 2).

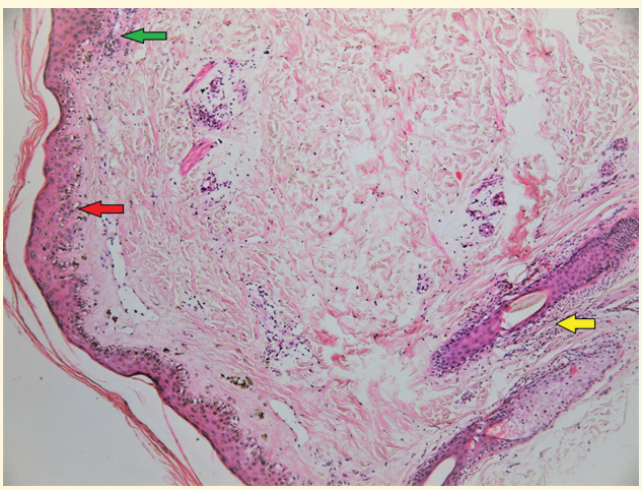
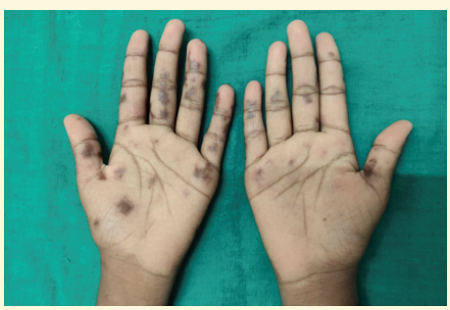


Figure 2 Vacuolisation of basal cell layer (red arrow) with sparse focal lymphohistiocytic infiltrate at dermoepidermal junction (green arrow) with perivascular infiltrate (yellow arrow) (H&E, x100).

These findings were suggestive of JSLE. According to SLICC criteria, 7 out of 11 clinical criteria and 1 out of 6 immunological criteria were positive. Diagnosis of juvenile SLE was made. The patient weighed 31kg and was treated with 5 mg/kg/day hydroxychloroquine, 0.5 mg/kg per day of oral prednisone with proton pump inhibitor, topical mometasone furoate 0.1% cream twice a day for 10 days on skin lesions, topical triamcinolone acetonide 0.1% gel over oral lesions with broad spectrum sunscreen for six weeks following which patient showed improvement of both skin and oral lesion (Figure 3).



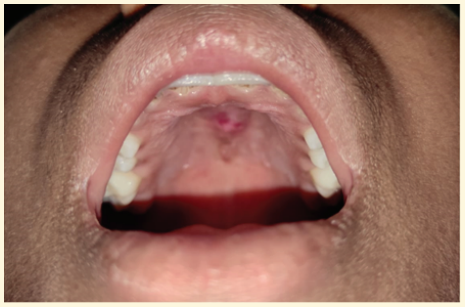


Figure 3 Improvement of both skin and oral lesions after six weeks of treatment

Discussion:

SLE is a chronic autoimmune inflammatory disease of multifactorial etiology most frequently affecting the skin, joints, kidneys, nervous, hematologic, and cardiovascular systems. Although SLE in children present clinically similar to adult onset SLE, there are some differences in frequency and severity of certain clinical manifestations. Many children and adolescents have an insidious onset of persistent fever, weight loss, fatigue, and arthralgia with general deterioration over weeks to months. Major organ involvement typically occurs within the first two to three years of disease onset. Some patients have acute or even life-threatening symptoms at presentation due to concomitant macrophage activation syndrome (MAS), severe renal disease, severe neuropsychiatric manifestations and increased risk of cardiovascular disease.[2] Patients with JSLE more often have renal involvement and encephalopathy as compared to adult-onset SLE.[3] The classic malar rash may be absent.

The etiology of SLE remains unknown, but genetic, hormonal, immunologic and environmental factors play a role. Identification of monogenic forms of SLE, mainly due to defects in the complement system, type I interferon pathway, aberrant nucleic acid repair and abnormal B cell development are some of the known etiological factors of SLE.[4] ANA, Anti-ribosomal P, anti-dsDNA and antihistone antibodies are more often found in patients with juvenile-onset SLE.[3,5]

SLE have variable signs and symptoms, therefore for diagnosis three classification criteria exist, one from the American College of Rheumatology (ACR) 1997, Systemic Lupus International Collaborating Clinics (SLICC) group classification criteria 2012, and European League Against Rheumatism (EULAR)/ACR classification criteria2019.[6-7] SLICC criteria are signifi-

cantly more sensitive than ACR criteria for JSLE classification.[8] For the SLICC criteria, to make a diagnosis of SLE four or more of the criteria are required including at least one clinical and one immunologic criterion.

The goals of therapy for patients with SLE are to ensure long-term survival, achieve the lowest possible disease activity, prevent organ damage, minimize drug toxicity, and improve quality of life. The agent that best combines safety and efficacy for the treatment of SLE is hydroxychloroquine (≤5 mg/kg per day) and low dose of glucocorticoid (less than 0.35 mg/kg per day of prednisone).[9] High-dose glucocorticoids have a significant impact on growth and appearance in children and adolescents so judicious use of corticosteroids should be advised. Steroid-sparing agents like mycophenolate mofetil or azathioprine can be used in patients who do not respond to the above drugs. We report this case as SLE being a female-prone, multisystem disorder when picked up and treated early can prevent lifelong crippling complications.

Conclusion:

Since JSLE manifests with a variety of clinical symptoms, early identification is difficult. In this instance, we were able to make an early diagnosis and treat the patient for six weeks with a modest dose of glucocorticoid and hydroxychloroquine, which caused the cutaneous lesion to reverse. Particularly in young girls, early diagnosis and treatment can prevent negative future outcomes.

Acknowledgement

None

Conflict of interest:

Photographs are published with the permission of parents.

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