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Atypical Presentation of SLE

Authors
Priyanka Thakur, Vijay Karan Singh, Dr. Gunjan Sharma
Corresponding Author
Dr Gunjan Sharma

Abstract
Systemic lupus erythematosus (SLE) presents with a wide array of cutaneous manifestations. The classical cutaneous features are malar rash, discoid rash, annular and papulosquamous lesions. There can also be some atypical manifestations. We present the case report of a patient, which satisfies the ACR/EULAR 2019 criteria for lupus and presented with unusual manifestation of a petechial rash.

Case Report

A 32 year female presented to our subdivisional level hospital with joint pain and swelling in both the wrists for a duration of two weeks. The joint pain was associated with morning stiffness, which used to get resolved in 3-4 hrs. It was associated with low grade fever and generalized myalgia. She took painkillers for the same, but it did not subside. The swelling and pain then progressed to involve other joints like small joints of the feet and ankle over a period of next 2 weeks. Later, she developed petechial lesions along with erythema over the dorsal aspect of hands feet, which gradually progressed to involve legs, arms and chest region. All routine investigation like complete haemogram and biochemistry were normal. ESR and CRP were raised significantly. Antinuclear antibodies (ANA) by Hep 2 cell lines came out to be positive; 1:100, 2+ homogenous pattern. Anti DsDNA antibody was also positive. According to new ACR/ EULAR criteria, both the clinical and immunologic criteria were met with 6 points in arthritis domain, 2 points in constitutional domain and 6 points in highly specific antibodies domain. The patient was started on methylprednisolone 24 mg which was tapered over the period of 2 months. Subsequently, she was started on hydroxychloroquine 300mg daily after seeking clearance from Ophthalmologist. She was also given sunscreen SPF 50 daily over face and limbs and topical corticosteroid fluticasone propionate 0.05% cream over the rash. Subsequent follow-up showed that patient had significant improvement in joint pain and skin lesions.

Discussion

SLE is a chronic multisystem autoimmune disease which runs a remitting and relapsing course. It was first described by Hargraves in 1948, with

discovery of LE cell[1]. It has several phenotypes, with varying clinical presentations from mild mucocutaneous manifestations to multiorgan involvement. Several immunopathogenic pathways play a role in the development of SLE. [2] SLE predominantly affects women of childbearing age, with a female to male ratio of 9 to 1. Several studies have repoted the development of serological abnormalities several years before the onset of clinical lupus. This is known as pre-clinical lupus, where a patient may have serological abnormalities consistent with SLE and may have some clinical features but still does not meet the criteria for SLE. There is evidence that a significant percentage of these patients with pre-clinical lupus, including those with incomplete lupus or undifferentiated connective tissue disease, may transition to clinical lupus and fulfill the SLE criteria later in life. The skin lesions of SLE may be lupus specific, while several nonspecific lesions are also seen. Lupus-specific lesions include (1). Acute cutaneous lupus erythematosus (ACLE) includes localized, malar, and generalized, (2) Subacute cutaneous lupus erythematosus (SCLE) includes annular and papulosquamous, and (3) Chronic cutaneous lupus erythematosus (CCLE) includes classic discoid lupus erythematosus (DLE), hypertrophic/verrucous, lupus panniculitis/ profundus, lupus tumidus, chilblains lupus, mucosal discoid lupus, and lichenoid discoid lupus.[3] Non-specific lesions of SLE are vascular abnormalities, non-scarring alopecia, interstitial granulomatous dermatitis, papulonodular mucinosis, calcinosis cutis, pigmentary abnormalities, nail abnormalities are not included in the diagnostic criteria of LE[4]. However, these are a common element of the clinical picture and provide additional confirmation of the diagnosis of LE. Non-specific skin lesions imply various complaints among patients and significantly reduce the quality of life[4]. Non-specific lesions usually correlate positively with an increased risk of internal organ involvement. Vasculitis is an atypical presentation of SLE. The prevalence of vasculitis in SLE is 11-36%. Dring flare up of SLE, vasculitis often occur and it is accompanied by fever, weight loss and fatigue. The most common type of vasculitis in SLE is leukocytoclastic vasculitis. It has varied clinical presentation is erythematous pin point lesions on fingertips and palms (36%), palpable purpura (25%), ischemic ulcers/lesions (14%), erythematous macules/papules (14%), urticarial lesions (11%) and nodular lesions[5,6,7]. Our patient had erythematous pin point lesions in palms and soles, petechial lesions on the distal extremities and chest. She also had erythema, swelling and tenderness in the joints of the hands and feet. There are several diagnostics challenges of SLE especially when there are atypical presentations. A strong clinical suspicion is required for the diagnosis.

Conclusion

SLE is a multisystem disorder in which specific and non-specific cutaneous manifestations can yield diagnostic and prognostic information. Therefore, proper understanding regarding cutaneous lesions of SLE will be helpful in its diagnosis and management.

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