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Research Paper

Delay In Diagnosis Of Systemic Lupus Erythematosus (SLE): A Case Report

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Abstract: *An autoimmune disease with multi-system manifestations, Systemic lupus erythematosus (SLE) has several clinical presentations varying from mild muco-cutaneous involvement to severe multi-organ involvement. We present the case 19-year old girl with a 4-year history of recurrent skin rashes along with recurrent painless oral ulcers, non-scaring alopecia, joint pains and low grade intermittent fever. There was wide-spread macula-papular rash over whole face involving naso-labial folds, arms, legs and most of the back. Investigations revealed normochromic normocytic anemia, low serum albumin, positive ANA, positive Anti-DsDNA and Anti-RNP antibodies and positive RA Factor and an excisional skin biopsy was done. She was diagnosed as having Systemic Lupus Erythematosus and started on hydroxycholoroquine and a short course of oral prednisolone resulting in marked improvement. This case highlights the difficulties and delays faced in diagnosis of SLE especially when the disease activity is mild and the patient has non-specific complaints.*
Keywords: *Systemic Lupus Erythematosus, Skin Biopsy, ANA.*

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I. Introduction

An autoimmune disease with multi-system manifestations, Systemic lupus erythematosus (SLE) has several clinical presentations varying from mild muco-cutaneous involvement to severe multi-organ involvement including renal, cardiac, musculo-skeletal and CNS disease.[1] Numerous immune-pathogenic pathways herald the development and progression of SLE and various pathogenic auto-antibodies such as ANA, Anti-DsDNA antibodies have been identified.[2] In spite of recent technologic advancements and understanding of etiologic factors of SLE, the specific pathogenesis is still not established. Diagnosing SLE is challenging. Several classification and diagnosis criteria for SLE have been postulated but their utility is a matter of debate in clinical setting.[3] Treatment of SLE is dictated by disease severity and organ system involvement.[4] Due to early detection of SLE and proper management, morbidity and mortality has improved dramatically. The 10-year survival rate of SLE was 63.2% in 1950s which has now improved to 95% in 2000s.[5] However SLE still poses a significant risk in morbidity and mortality if not timely diagnosed and adequately treated.

II. Case Report

We report the case of a previously healthy 19-year old girl who presented with a 4-year history of recurrent skin rashes, currently having generalized erythematous pruritic rash over her face, arms, legs and back for last 3 months along with photosensitivity over sun-exposed areas. She had consulted various GPs and dermatologists multiple times and had been prescribed various topical steroids which resulted in improvement of the rash. However, the rash recurred when she stopped applying the topical medicines. On exploration of history, there was history of recurrent painless oral ulcers, non-scaring alopecia, joint pains involving small

joints of hands without swelling or AM stiffness and low grade intermittent fever. She was unmarried and a student of A levels. She did not smoke or use illicit drugs. There was no family history of psoriasis, scabies or skin disorders. On examination, she was vitally stable with a temperature of 99.8^{o}F. There was wide-spread macula-papular rash over whole face involving naso-labial folds, arms, legs and most of the back. Joint examination revealed mild tenderness with no joint swelling or limitation of movements. There was no focal sensory, motor or cerebellar neurological deficit with power 5/5 in all four limbs. Cardiovascular and respiratory examinations were normal.

On investigation, CBC revealed normochromic normocytic anemia with hemoglobin of 8.5 g/dl with normal TLC and platelet counts. ESR was raised at 95 mm/hour with normal CRP. Serum albumin was low at 2.8 g/dl but urinalysis, LFTs and RFTs were normal. Serologies for TPHA, HBV, HCV and HIV were negative. Blood and urine cultures were negative. An excisional skin biopsy was performed which showed epidermis with mild basal cell vacuolization, mild follicular plugging, focal melanin incontinence and mild perivascular inflammation. An autoimmune profile was done which showed positive ANA, positive Anti-DsDNA and Anti-RNP antibodies, positive RA Factor with normal serum C3 and serum C4 and negative Anti-Ro, Anti-La, Anti-CCP and Anti-Phospholipid antibodies. Her serum CPK and serum Aldolase were normal. Her Echocardiography and abdomino-pelvic Ultrasound were within normal parameters. She was diagnosed as having Systemic Lupus Erythematosus and started on hydroxycholoroquine 300mg (5mg/kg body weight) per day. She was given a short course of oral prednisolone, starting from 30mg (0.5mg/kg body weight) per day with tapering and stopping in 4 weeks. In addition, she was prescribed topical steroids, Sunblock SPF 60, oral omerprazole, iron, calcium and Vitamin D supplements. She showed marked improvement after initiation of therapy with resolution of rash within 3 weeks. At 6 months follow up, she was in remission and tolerating hydroxycholoroquine without any adverse effects.

III. Discussion

Although the prognosis of SLE has improved substantially over the years, SLE can still cause a life-threatening disease and premature mortality. According to Tselios et al.[6] lupus patients were three times more likely to die from any cause as compared to patients without lupus especially lupus patients who had onset of SLE before 40 years of age. Infections (24.5%), atherosclerosis (15.7%), lupus flare (13.3%) and malignancies (9.6%) were the leading causes of mortality in lupus patients.[6] In China, Mu et al.[7] reported substantial mortality in SLE especially female patients with the leading mortality cause being infections. Therefore it is of pertinent importance that SLE be diagnosed timely and early treatment be initiated. With a sensitivity of 97% and specificity of 92%, the Systemic Lupus International Collaborating Clinics (SLICC) criteria is commonly used for diagnosing Systemic Lupus Erythematosus (SLE).[8] Requirement for diagnosis of SLE is $\geq$4 criteria (at least 1 clinical and 1 immunological criteria) as shown in Table I.[8] Based on the SLICC criteria, our patient had a score of 5 namely chronic cutaneous lupus, non-scarring alopecia, oral ulcers, positive ANA and positive Anti-DsDNA antibodies.

Table 1: SLICC criteria for diagnosis of Systemic Lupus Erythematosus

SLICC criteria for diagnosis of SLE	
CLINCAL CRITERIA	**IMMUNOLOGICAL CRITERIA**
1. Acute Cutaneous lupus	**12.** Positive ANA
2. Chronic Cutaneous lupus	**13.** Positive Anti-DNA Antibodies
3. Oral or nasal ulcers	**14.** Positive Anti-Smith Antibodies
4. Non-scaring alopecia	**15.** Positive Anti-Phospholipid Antibodies
5. Arthritis	**16.** Low Complement Levels (C3/C4)
6. Serositis	**17.** Positive Direct Coomb's Test
7. Renal Involvement	
8. Neurological Involvement	
9. Hemolytic anemia	
10. Leucopenia	
11. Thrombocytopenia	
Requirement for diagnosis of SLE: $\geq$4 criteria (at least 1 clinical and 1 immunological criteria) **OR** Biopsy-proven lupus nephritis with positive ANA or Anti-DNA	

Management of SLE is complex and variability in clinical practice exists as the treatment is dictated by disease severity and organ system involvement. Corticosteroids are often used as first-line agent because of their rapid onset of action and powerful anti-inflammatory actions.[9] The dose and duration of steroids depend on the disease severity. Hydroxychloroquine is especially useful to treat fatigue, muco-cutaneous and musculoskeletal

manifestations in lupus and to prevent flares.[10] Hydroxychloroquinine is safe during all trimesters of pregnancy and also improves long-term survival by protecting against irreversible organ damage, thrombosis, and bone mass loss in SLE.[11] Even with long-term hydroxycholoroquine use, retinal toxicity is rare but should be monitored with Ocular Coherence Tomography (OCT).[10] Other agents used in SLE include simple analgesics (paracetamol and NSAIDs) for pain relief, conventional DMARDs (methotrexate, azathioprine, cyclosporine, cyclophosphamide) and biological DMARDs (rituximab, belimumab, etanercept, abatacept) with increasing disease severity and involvement of major organ systems especially renal system.[12,13,14] Our patient had predominant muco-cutaneous involvement without any major system involvement. She responded well to treatment with hydroxychloroquine and a short course of oral prenisolone. At 6 months of follow up, her disease was in remission with hydroxycholoroquine and she did not suffer any adverse effects. In conclusion, this case highlights the difficulties and delays faced in diagnosis of SLE especially when the disease activity is mild and the patient has non-specific complaints. Therefore it is necessary that the treating physician keep SLE in his differentials when managing such patients so that timely diagnosis and early treatment may lead to reduction in morbidity and mortality.

Consent: Informed consent was taken from the patient.
Conflict of Interest: None declared.

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