Case Report

Acute flare-up of systemic lupus erythematosus by unknown insect bite: A case report

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ABSTRACT

Systemic lupus erythematosus (SLE) is a rare chronic inflammatory autoimmune disease. In genetically susceptible cases, environmental factor plays a major role as triggering factor. In this case study, a 21-year-old female with known case of SLE on treatment presented with fever, joint pain, rashes over the face after the bite of unknown insect, which was reduced immediately after treating her with a high dose of steroid.

KEY WORDS: Autoimmune disease, Environmental factor, Malar rash, Multisystem disease

INTRODUCTION

Systemic lupus erythematosus (SLE) is disease of young females, predominantly affecting females than males most commonly seen between the age of 20 and 30 years. The prevalence of SLE ranges from about 0.03% in Pale-skinned people to 0.2% in Afro-Caribbeans.[1] In India, SLE patients seem to have a high familial aggregation of autoimmune rheumatic diseases which is more pronounced in the background of parental consanguinity.[2] The most common cause is unknown but environmental factors, viral infections, hormones ultraviolet (UV)-rays, chemicals, physical, or emotional stress could flare up SLE.

CASE REPORT

A 21-year-old female with known case of SLE on treatment presented with sudden onset of fever for the past 1 day which was high grade, continuous not relieved by paracetamol. She also complains of knee pain in both the legs for the past 1 day. The patient also complaints of papular rashes with over the cheeks after 2 days. The patient was febrile.

Examination

On examination, minimal restriction of movements was present in both the knee joint. No other specific abnormalities detected.

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Investigations

Investigations such as complete blood count, IgG, IgM, blood smear examination, and RA factor were carried out and there were no significant findings.

During the patient's stay in the hospital, the patient was found to have an unknown insect bite. ANA profile was done and the test results showed high dsDNA [Table 1], anti-nRNP/Sm [Table 2], and presence of anti-SS-A antibody [Table 2], anti-nucleosome antibody [Table 2]. Thyroid function tests, liver function test, and creatinine level were normal. There were no other significant abnormalities in other parameters.

Differential Diagnosis

Hence, other diseases such as rheumatoid arthritis, and infectious diseases including scrub typhus, dengue, chikungunya, and malaria were excluded from the study.

Management

The patient was confirmed to have an acute flare-up of SLE. The patient was started on methylprednisolone 30 mg IV for 2 days. The patient improved symptomatically. There were no signs of fever and joint pain was also reduced. The patient was stable. Hence, tapering of steroids was done and maintained on tablet hydroxychloroquine, tablet osteomin, trolamine salicylate (topical medication), and other supportive medications were given.

The mortality was reduced as the patient was immediately treated with a high dose of steroid.

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DISCUSSION

SLE involves multiple organs with the presence of autoantibodies. Other than environmental factors, TREX-1 gene defects, even chromosomal abnormalities like Klinefelter syndrome can also cause SLE. The symptoms of SLE include fever, fatigue, mouth ulcers, skin lesion, myalgia, joint pain, hair loss, headaches, and confusion.

The confirmation of SLE is based on Systemic Lupus International Collaborating Clinics classification criteria which have two major criteria[3]

1. The clinical criteria include the following:
- Acute cutaneous lupus erythematosus including "malar rash" [Figure 1]
- Chronic cutaneous lupus erythematosus (example: "Discoid rash")
- Oral ulcers
- Non-scarring alopecia
- Synovitis (≥2 joints) or tenderness on palpation (≥2 joints) and morning stiffness (≥30 min) (non-erosive arthritis) but never associated with deformity
- Serositis (pleurisy)
- Renal involvement (24-h urine protein, >0.5 g)
- Neurological involvement (neuropsychosis/ neuropathy)
- Hemolytic anemia (Hb <10g/dl)
- Leukopenia or lymphopenia
- Thrombocytopenia (<100,000/μL).

2. Immunological criteria include the following:
- High level of antinuclear antibodies (ANA)
- Presence of anti-dsDNA antibodies
- Anti-Sm antibodies presence

Test Name	Patient Value	Ref Range
HAEMATOLOGY		
Haemoglobin	11.7	M: 13.5 - 17.0 gm/dl; F: 12.0 - 15.5 gm/dl
RBC Count	3.90	Male: 4.6 - 6.0 Million/cu.mm, Female: 4.2 - 5.4 Million/cu.mm
PCV	35.1	Male: 40 - 52 % ; Female: 38 - 45 %
MCV	83.0	76 - 96 fl.
MCH	26.7	27 - 31 pg
MCHC	32.0	32 - 36 %
RDWCV	10.00	11.5 - 14.5 %
RDWSD	56.99	35.0 - 56.0 fl
Total WBC Count	8,500	4000 - 11000 cells/cu.mm
Neutrophils	65	40 - 65 %
Lymphocytes	29	30 - 50 %
Eosinophils	06	2 - 8 %
Platelet Count	1.98	1.5 - 4.5 lakhs/cu.mm
MPV	9.55	7.0 - 11.0 fl
PDW	12.42	7.0 - 17.0 fl
PCT	0.18	0.10 - 0.28 %
ESR 1 hour	49	Male: 5 - 20mm, Female: 5 - 15mm
BIOCHEMISTRY		
SGOT	20.5	5 - 40 IU/L
SGPT	20.8	5 - 41 IU/L
Sr.Creatinine	0.85	Male:0.9 - 1.3 mg/dl, Female: 0.6 - 1.1 mg/dl
Calculated GFR	80.3	
IMMUNOLOGY		
dsDNA	166.9	25 U/ml NEGATIVE > 25 U/ml POSITIVE
C3	141.8	90 - 180 mg/dl
C4	28.0	10 - 40 mg/dl

Table 1: Hematology, biochemistry, immunological report of the patient

- Antiphospholipid antibodies such as anticardiolipin and anti-β 2-glycoprotein I such as IgA-, IgG- or IgM-antibodies; false-positive Venereal Disease Research Laboratory test)
- Low complements levels
- Positive for direct Coombs test in the absence of hemolytic anemia.[3]

Confirmation of the Diagnosis

At least one clinical and one immunological criteria have to be fulfilled or deposits of anti-dsDNA on glomerular basement membrane (lupus nephritis) confirms the diagnosis of the disease.[3]

On characteristic clinical findings of the skin, joints, and renal and the central nervous system SLE is diagnosed, as well as on serological parameters such as ANA, in particular, antibodies to dsDNA SLE is confirmed. Most of the clinical symptoms do not always occur simultaneously and can develop at any stage of the disease.[3]

In this case, the patient presented with symptoms similar to scrub typhus, dengue, malaria, and other fever causing infectious diseases but the laboratory investigation confirmed that it is acute flare-up of

Test Name	Patient Value	Ref Range
ANA PROFILE		
Anti nRNP/Sm Antibody	61+++	< 10 NEGATIVE
Anti Sm Antibody	NEGATIVE	< 10 NEGATIVE
Anti SS - A Antibody	21+	< 10 NEGATIVE
Anti Ro - 52 Antibody	NEGATIVE	< 10 NEGATIVE
Anti SS - B Antibody	NEGATIVE	< 10 NEGATIVE
Anti Scl -70 Antibody	NEGATIVE	< 10 NEGATIVE
Anti PM Scl Antibody	NEGATIVE	< 10 NEGATIVE
Anti Jo - 1 Antibody	NEGATIVE	< 10 NEGATIVE
Anti CENP B Antibody	NEGATIVE	< 10 NEGATIVE
Anti PCNA Anitibody	NEGATIVE	< 10 NEGATIVE
Anti dsDNA Antibody	NEGATIVE	< 10 NEGATIVE
Anti Nucleosome Antibody	7+	< 10 NEGATIVE
Anti Histone Antibody	NEGATIVE	< 10 NEGATIVE
Anti Rib . p - Prot Antibody	NEGATIVE	< 10 NEGATIVE
Anti AMA - M2 Antibody	8+	< 10 NEGATIVE

Table 2: Antinuclear antibody profile of the patient

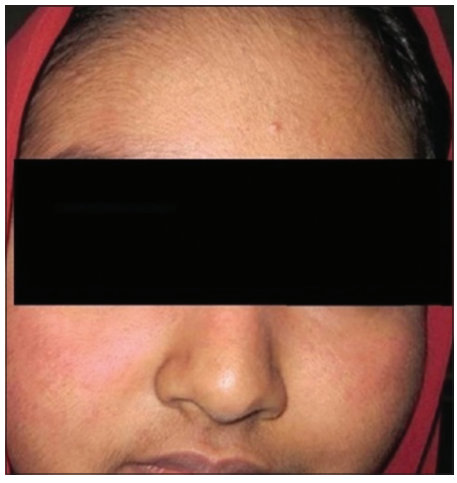

Figure 1: Typical clinical presentation of malar rash

SLE. Here, the triggering factor was unknown insect bite that caused acute flare-up of SLE. The patient presented with malar rashes with butterfly distribution and a papule over the cheeks after the bite of unknown insect, which is a typical sign of dermoepidermal lupus erythematosus.

In genetically predisposed patients, there are certain viruses such as cytomegalovirus, Epstein–Barr virus, and parvovirus B19 – have also been associated with the pathogenesis of the disease, which triggers autoimmunity through molecular mimicry, activation of T lymphocytes by superantigens, and bystander activation.[4] Vitamin D deficiency is known to trigger flare-up of SLE.[4] Furthermore, psychological factors such as depression, anxiety, alexithymia, negative emotions, stress, and sex hormones such as estrogens, progesterone, and prolactin androgens can also give rise to flare-up of SLE. Inherited mutation in complement components C1q, C2, and C4, in the immunoglobulin receptor FcyRIIIb or in the DNA exonuclease TREX1 plays a major role in SLE. UV-radiation can cause defects in apoptosis or in the clearance of apoptotic cells that leading to inappropriate exposure of intracellular antigens on the cell surface, which makes polyclonal B-and T-cell activation and autoantibody production can cause SLE.[1] Environmental factors support this fact leading to flare-up of SLE. The important mechanism of tissue damage in SLE in activation is immune complex formation which can even lead to organ damage and vasculitis.[1]

CONCLUSION

SLE flaring up after unknown insect bite is uncommon. Confirmation of flare-up should be done after excluding other infectious disease causing similar symptoms after a laboratory test. Acute mortality in SLE occurs due to acute flare-up of the disease. In this patient, since immediate treatment was given on time, mortality was reduced occurred due to its flare-up, and patient is asked to continue the given medications to reduce further flare-up of the disease. Although acute flare-up of SLE cannot be minimized, mortality occurring due to acute flare-up can be prevented by high-dose steroid treatment and regular follow-up.

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