University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2018, Vol. 4(1)

A RARE CASE SCENARIO OF SYSTEMIC LUPUS ERYTHEMATOSUS

PREETHI SHAHILA
Department of General Medicine,
MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract :

Systemic lupus erythematosus (SLE) is the prototypic autoimmune disease characterized by the production of autoantibodies to components of the cell nucleus, in association with diverse clinical manifestations encompassing almost all organ systems. SLE is a complex disease with variable presentations, course and prognosis characterised by remissions and flares. Herewith we report a young female with SLE who presented to us with autoimmune hemolytic anemia and hepatitis. We present this case for its rarity.

Keyword : Systemic lupus erythematosus, autoantibodies, autoimmune disease, autoimmune hemolytic anemia, hepatitis

CASE REPORT:

A 28year old female was referred to us, as a case of transfusion induced hemolytic reactions .Her initial symptom was myalgia and fatigue which lasted for 3 months. She was treated outside as anemia and two units of blood transfusion was given ,following that she developed yellowish discolouration of sclera, history of high coloured urine and low grade intermittent fever. She gives history of symmetrical polyarthritis involving the knee, ankle and metacarpophalangeal joints for the past five years. She didn't give any history of jaundice, blood transfusions, herbal medications in the past.

On examination she was febrile, pallor and icteric. She had no pedal edema / generalised lymphadenopathy. She had no other signs of liver cell failure. VITALS: Pulse rate-102/mt, Blood pressure-100/60mmhg,Respiratory rate-28/mtTemperature-101 f, Jugular venous pulse was not elevated System examination-Abdomen was soft. Liver was palpable 3cm below the right costal margin, softand tender. Spleen was palpable 3cm below left costal margin. Cardiovascular system-hemic murmur was heard in the pulmonary area. Central nervous system and Respiratory system was unremarkable.

LABORATORY INVESTIGATION:

Routine urine analysis was normal. Blood hemoglobin- 4.5gms%,Total WBC count-5600cells/cubic mm,platelet-3,00,000/cubic mm.ESR-112mm,serum bilirubin total-12gms%,direct-7.4gms%,indirect-4.6gms%

.Liver enzymes-SGOT-134IU/L,SGPT- 78IU/L,ALP-45IU/L.Renal function test & serum proteins were normal. Serum IgM Leptospirosis, Malaria parasite smear, viral marker for hepatitis were negative. Rheumatoid factor was negative. VCTC&VDRL were non reactive. ECHO&X-Ray Chest PA view was normal. Ultrasound abdomen showed hepatosplenomegaly and features suggestive of hepatitis.

S.No	Investigation	Reports
1	Reticulocyte count	2.8
2	Peripheral smear	Microcytic hypochromic anemia, occasional spherocytes
3	Direct Coomb's Test	Positive
4	Osmotic fragility Test	Negative
5	Blood grouping & typing	AB positive (done outside), B positive (done in our hospital)
6	ANA	Positive

Table 1-shows investigation for hemolytic anemia.

S.No					
1	ds DNA	Positive	9	Scl-70	Negative
2	N RNP/Sm	Negative	10	Pm-Scl	Negative
3	Sm	Negative	11	Jo-I	Negative
4	SS –A	Negative	12	CENP B	Negative
5	Ro-	Negative	13	PCNA	Negative
6	SS-B	Negative	14	Nucleosomes	Positive
7	Histones	Negative	15	Ribosomal P protein	Positive
8	AMA-M2	Negative			

Table 2-shows ENA profile Anti-LKM-Negative,Anti-SM Antibody-Negative As she fulfills American college of rheumatology (ACR) classification criteria for SLE, she was diagnosed as a case of systemic lupus erythematosus with hepatic manifestation and autoimmune hemolytic anemia. She was treated with steroids. On follow up, she had no icterus clinically, her haemoglobin and liver function test became normal. She is on regular follow up.

Discussion

The term LUPUS means (Wolf in Latin)an erosive skin lesions of wolf's bite. Von Hebra coined the term lupus erythematosus(1816 -1880). kaposi recognised the systemic disease with visceral manifestation(1837-1902).Females are affected 9 times more than male. Sixty five percent havedisease onset between sixteen and fifty five years3.1 in 1000 females in India are at risk of developing SLE. Hepatic manifestation in SLE is uncommon.

The pathogenesis of Lupus was unclear. Genetics, environmental interactions results in abnormal immune responses that generate pathogenic autoantibodies and immune complexes that deposit in tissue, activate complement, causes inflammation which lead to irreversible organ damage1.

According to American college of rheumatology, there are 11 classification criteria for SLE.More than 4 of these criteria, SLE likely to be the diagnosis1. Malar rash, discoid rash, photosensitivity, oral and nasopharngeal ulcer, non erosive arthritis, serositis (pleuritis/pericarditis), renal disorder (proteinuria or cellular cast), neurologic disorder (psychosis/seizures) hematogical disorder (hemolytic anemia or leukopenia (<4000/micro.l),lymphopenia (1500/micro.l) or thrombocytopenia (<1,00,000/micro.l). ANA positive by immunofluorescence, Anti ds DNA, Anti-Sm/Anti-phospholipid +Ve. Visceral manifestation is a life threatening manifestation in SLE like Lupus nephritis, neuro pshychiatic syndromes, cardiac involvement, pleuro-pulmonary manifestation, hematological
manifestation, ophthalmic,G.I.T and hepatic involvement. Clinically hepatic disease is unusual in SLE.

Fatty liver and abnormal liver enzymes is a common finding. Both Autoimmne hepatitis and hepatitis in lupus are similar clinically and serologically. ANA is positive in both disorders. Absence of Anti -Sm and anti-mitochondrial antibodies, Presence of anti-ribosomal P protein antibodies suggest lupus hepatitis3.

Anti-ribosomal P protein antibodies are believed to be correlated with nephritis (class v), hepatitis (chronic active hepatitis), neuropsychiatric manifestation (depression and pshycosis) and dermatitis in SLE9. Anti ribosomal P protein antibodies was detected in 40-50% in asian patients with SLE.
Transfusion induced hemolytic reaction occurs due to preformed antibodies that lyse donar erythrocytes1.Acute reaction results in hemodynamic instability and renal failure. Delayed reaction occurs 1-2 weeks later, due to prior sensitisation to RBC alloantigen. Warm autoimmune haemolytic anemia occurs in SLE. Blood group compatability test was performed to detect the autoantibodies. Specialised procedure like adsorption test can be used8 Mismatched blood transfusion occurs in SLE because of change in blood groups, due to the high titre and multiple isoagglutinins formed against red cell antigens7.Red blood cell transfusion should be avoided, unless patient presenting with life threatening anemia8.A transient blood group change was reported in a female with SLE, from A to AB due to agglutinin to anti-B antibodies5.

In life threatening complication, monthly pulses of cyclophosphamide combined with pulse corticosteroids for six to twelve months followed by quarterly pulses of cyclophosphamide for atleast one year beyond remission or azathioprine as maintanence therapy, if no response rituximab or mycophenolate mofetil can be used3.

Lupus hepatitis was reported in 4 (3%) patients out of 131 SLE patients with the presence of anti ribosomal P antibodies4,6.A total 242 patients on follow up for 1 year, lupus hepatitis was reported in 14 cases(5.8%).They are respond to high dose steroids10. In this study 305 patients followed up for 7 years retrospectively 30 patients with SLE identified ,of whom 20 (6.6%)had severe haemolytic anemia2.
Patients with SLE need not fit into the classification criteria of ACR initially, but may satisfy it as the disease progresses. Any young female presenting with autoimmune haemolytic anemia or hepatic manifestation, SLE should be suspected at the earliest.

REFERNCES

1.Harrisons principles of internal medicine,18th edition.volume 2;Part 15:chapter-319.
2.Sultan SM,Begum S,Isenberg DA,Prevalance,pattern of disease and outcome in patients with systemic lupus erythematosus with severe haematological problems. Rheumatology Oxford 2003 feb;42(2):230-4
3.Kelley's textbook of rheumatology,8th edition. Volume 2;Part 11:chapter- 74,75.
4.Arnett FC,Reichlin M .Lupus hepatitis: an under-recognised disease feature associated with autoantibodies to ribosomal P,Am J Med 1995 Nov;99 (5):465-472.
5.Nakamura N,Fujita T,Nakamura M, et al. systemic lupus erythematosus&blood type Lancet 2006 Sep 16;368 (9450):1022.
6 Ebert,Ellen C.MD,Haqspiel,Klaus D.MD,gastrointestinal and hepatic manifestation of systemic lupus erythematosus,journal of clinical gastroenterology may-june 2011;volume45 issue 5p: 436-441.
7.Daniel Jeffrey Wallace,Bevra Hahn,Edmund L.Dubois.Haematological&lymphoid abnormalities in systemic lupus erythematosus chapter 41 pg 811
8.Petz LD.A physician's guide to transfusion in autoimmune haemolytic anemia.British journal of haematology 2004 Mar;volume 124 issue 6:712-6.
9.Ben-Ami SD,Blank M,Altman A. The clinical importance of anti-ribosomal-P antibodies, Harefuah 2010 dec;149 (12):794-7,810.
10.Piga M,Vacca A.Porru G,Cauli A,Mathieu A. Liver involvement in systemic lupus erytematosus: incidence, clinical course and outcome of lupus hepatitis clin Exp Rheumatol. 2010 Jul- Aug;28(4):504-10.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities