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# A RARE CASE OF SLE PRESENTING AS HLH

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## Abstract

Hemophagocytic Lymphohistiocytosis (HLH) presentation in systemic lupus erythematosus (SLE) is a very rare entity. The current incidence is 1 person per million population per annum. A 24-year-old male presented with fever for one month along with loss of appetite and severe weight loss. Vital signs were within normal limits except for body temperature of 102 °F. The patient is also presented with pancytopenia, hepatosplenomegaly, liver function abnormalities, coagulopathy, and hyperferritinemia. All other tests for infectious diseases were negative. So, rheumatological examination was done and DCT was found be strongly positive, ICT- negative , c3- <40 ( 88-165), c4- 28.2 mg/dl( 14-44 mg/dl), ANA immunoblot- smith antibody- +++, SSA- +, dsDNA- negative. So SLE was confirmed and he is treated with inj methylpred for 3 days f/b tab wysolone 1 mg/ kg po.patient improved dramatically.

## Introduction

Hemophagocytic Lymphohistiocytosis (HLH) is varocious and fatal condition caused by hyperactivation of immune system. Due to this enormous amount of inflammatory cytokines are released by uninhibited production of activated CD8 T cells, lymphocytes, and macrophages (1). It is a rare disease generally seen in children and young adults (2). The general presentation of the disease is chronic fever, pancytopenia, enlargement of liver and spleen, defects in liver function, coagulation disorders and hyperferritinemia (3). HLH is primary due to genetic defects and secondary HLH associated with SLE is a rare phenomenon and its projected prevalence is around 0.9% - 4.6% (4, 5). Here, we present a rare case of young male with SLE presented with HLH.

**Case presentation**

A 24 male patient came with complains of fever since one month ,loss of appetite ,weight loss around 6 kg in one month, loose stools since 5 days. He didn't have any known comorbidities. His vitals were normal (PR 96/ min, BP 90/70, RR 25/min, SPO2- 99% in room air) with 102 °F fever with the presence of pallor.

Systemic examination revealed normal CVS -S1S2 heard, Bilateral FINE CREPTS heard during respiratory system examination, Per abdomen - soft, splenomegaly of 4 cm was noticed from left costal margin, CNS - NFND.

Laboratory investigations showed Hb - 7.7, TLC-2290, Platelet -2.17 lakh, albumin 2.7, total protein- 7.2, RFT , ELECTROLYTES , Urine Routine -normal , ESR - 80 mm/hr, CRP - 62.8 mg/dl , SERUM LDH - 1023, serum ferritin - >1000, serum Fibrinogen - 158 mg/dl, serum TRIGLYCERIDES- 283.

Ultrasonography of abdomen revealed hepatosplenomegaly.

Microcytic hypochromic and Normocytic normochromic RBCs, leucocytopenia , adequate platelets ,reticulocyte count - 0.2 %,PT- 11.0 were noted in peripheral smear.

Coagulation tests such as INR- 0.92, APTT- 44.8 were observed. Electrolytes were serum calcium - 6.6 , vitamin D- 11.4, serum Magnesium- 1.9, serum phosphorus- 2.9, TFT – normal and elevated pancreatic enzyme (Serum amylase- 650, serum lipase – 3139)

Serological tests ( HIV, HBsAG, HCV) were negative. Infectious tests such as dengue ( NS1, IGG, IGM,) - negative, leptospirosis IGM- negative, Scrub typhus- negative, MALARIA mp/mf - negative, Blood culture – negative. So rheumatological tests were done and DCT - positive(++), ICT- negative , c3- <40 ( 88-165), c4- 28.2 mg/dl( 14-44 mg/dl). ANA immunoblot- smith antibody- +++, SSA- +, dsDNA- negative.

High resolution CT thorax showed paraseptal and centriacinar emphysematous changes in the bilateral lung fields with basal fibrotic strands –ILD,

CT abdomen- Hepato-splenomegaly, pancreatitis. So patient is diagnosed to have SLE .TREATED WITH INJ METHYLPRED FOR 3 DAYS f/b TAB WYSOLONE 1 mg/ kg po. Patient improved dramatically.

**Discussion**

Here, we discuss a young male adult case of SLE presented with HLH with prolonged fever, appetite loss, weight loss around 6 kg in one month. The initial differential diagnosis was wide such as infections, malignancy or autoimmune diseases. According to the HLH-2008 diagnostic criteria, the diagnosis of HLH is made by five out of eight diagnostic criteria of HLH such as fever, pancytopenia, splenomegaly, bone marrow hemophagocytosis, and hyperferritinemia (6, 7).

Hyperactivation of immune system in HLH causes the release of massive amount of inflammatory cytokines by uninhibited production of activated CD8 T cells, lymphocytes, and macrophages. Fever is produced by interleukins and tumor necrosis factor alpha (TNF-α).

 

Activated macrophages secretes ferritin, which in turn causes increased levels of plasminogen activator causing hyperfibrinolysis (8).

In a retrospective study, it was found that unusual presentation of SLE presented with initial symptoms of HLH (9). We have also found the similar findings in our case.

Management of secondary HLH is targeted at managing the primary condition. In HLH due to SLE, high dose of steroids and immunosuppressive agents are found to be effective (10). Similarly, our patient was treated with inj methylpred for 3 days f/b tab wysolone 1 mg/ kg po and he improved dramatically.

In HLH, mortality rate is high which is around 20% - 88% caused by secondary infections and advancement of the underlying secondary causes (11). So the diagnosis and treatment should be done at the earliest to prevent mortality.

## Conclusion

Adult males with HLH presenting with SLE is very rare. Both have similar clinical features which possess great diagnostic challenge. So testing of SLE should be done in patient presenting with HLH. Since the mortality is very high, prompt diagnosis and appropriate treatment should be done.

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