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Original Research Paper

Rheumatology

A RARE CASE OF MACROPHAGE ACTIVATION SYNDROME

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ABSTRACT Macrophage Activation Syndrome is a rare and potentially fatal disease, to be considered in patients with rheumatic diseases who present with features of systemic inflammatory response syndrome and multiorgan dysfunction1-3.We report a patient of systemic lupus erythematosus who presented with atypical features of Macrophage activation syndrome

KEYWORDS :

Introduction

Macrophage Activation Syndrome is a rare and potentially fatal disorder of hyperactive histiocytes. There is an overwhelming activation of normal T cells and macrophages which can cause clinical and hematological alterations. The clinical presentation is similar to systemic inflammatory response syndrome. The mortality is high inspite of appropriate treatment and early diagnosis is essential for good outcome

Case History

A 31 years old lady a known patient of Systemic lupus erythematosus and Hypothyroidism presented with fever of 10 days duration, giddiness of 1 day duration which was not associated with postural changes.She also gave a history of photophobia , severe bitemporal headache and one episode of generalised tonic clonic seizure.There was no features or loclising symptoms of infection or new drug intake,

On examination,she was conscious, oriented and febrile.She was not cyanosed or icteric.Her pulse rate was 98 per minute and her arterial pressure was 110/80 mm of Hg.She had diffuse nonitchy salmon pink colored maculopapular rash over her whole body . Spleen was palpable 2 cm below midclavicular line.There was no focal neurological deficit.Heart sounds were normally heard and there was no murmur.Normal vesicular breath sounds heard over both lungs. Investigations revealed pancytopenia(platelets 71000 Hb 10.0g%,total leucocyte count 1800/microliter with absolute neutrophil count of 680 per microliter, platelets 71000 /microliter)raised LDH 756 IU/L), uric acid of 2.0 mg/dl,serum Triglycerides 401mg/dl and serum albumin of 2.4mg/dl/.Serum ferritin was 1818 ng/ml.Her ESR was 12mm in one hour and CRP was 0.5 mg/dl..All body fluid cultures were negative . MRI of brain and EEG did not reveal any abnormality.Further evaluation revealed raised antidsDNA titer,hypocomplementemia and raised anticardiolipin antibodies.She was diagnosed as case of Macrophage activation syndrome based on clinical and laboratory criteria and treated with pulse methylprednisolone and thereafter oral corticosteroids along with other supportive measures followed by Mycophenolate mofetil thereafter.She made rapid recovery in clinical and laboratory parameters with the above measures,

DISCUSSION

Macrophage Activation Syndrome (MAS) is a rare and potentially fatal disease of normal but overactive histiocytes occuring as a complication of systemic rheumatic diseases.An overwhelming activation of normal T cells and macrophages causes clinical and biochemical alterations. The clinical presentation is similar to systemic inflammation response syndrome (SIRS) and death is inevitable in the absence of treatment.

The clinical entity has to be suspected when patients with systemic rheumatic disease present with fever unresponsive to antibiotics, general fatigue, falling ESR, pancytopenia and multiple biochemical perturbations The diagnostic criteria is as follows.

(1) Fever- Seven or more days of a temperature as high as 38.5°C (101.3°F).
(2) Splenomegaly.
(3) Cytopenia - Counts below the specified range in atleast 2 of the following cell lineages:Absolute neutrophils less than 1000/mL;Platelets less than 100,000/mL; Hemoglobin less than 9.0 g/dL
(4) Hypofibrinogenemia or hypertriglyceridemia.
(5) Hemophagocytosis.
(6) Rash

At least five of them have to be there to have a definite diagnosis.

Macrophage Activation Syndrome has to be considered in patients with a SIRS like clinical presentation in systemic rheumatic diseases[4,5]. Hyperferritinaemia >10000 µg/l seems to be a good marker for defining patients with MAS. But low ferritin does not rule out the condition as it may possibly reflect ferritin measurements some time after the peak of macrophage activation[5].Similarly bone marrow examination or tissue biopsy to demonstrate hemophagocytosis is increasingly being considered as non essential for diagnosis as it may not be present in early stages.This may be the reason for the relatively less elevated ferritin in our patient.In the absence of prospective controlled trials, corticosteroids, cyclosporin A,intravenous immunoglobulins and rituximab and etoposide are administered with varied success[6].. This case is presented to enlighten physicians regarding the clinical entity of Macrophage Activation Syndrome to be suspected when patients presented with fever unresponsive to antibiotics, general fatigue, falling ESR, pancytopenia of unknown origin and liver dysfunction with elevated ferritin.

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