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Case Report

Macrophage Activation Syndrome - an unusual Complication of SLE

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Abstract

Macrophage Activation Syndrome (MAS) is a rare and potentially fatal condition that can complicate many autoimmune diseases. It is a type of Hemophagocytic Lymphohistiocytosis (HLH). HLH is a potentially fatal disease of normal but overactive histiocytes and lymphocytes commonly occurring in infancy but can occur in all age groups. There are two types of HLH: Familial and acquired. Familial HLH is an autosomal recessive disorder and accounts for 25% of the cases. Acquired type is secondary to drugs, malignancies, rheumatological disorders. There are five subtypes of inherited HLH: familial HLH and types 1-5. HLH may result from an inappropriate immune response to a viral illness like EBV. It may be due to X-linked Lymphoproliferative Disease (XLP) caused by a mutation in SH2D1A or XIAP gene.

Usually, MAS term is used for HLH secondary to rheumatological disorders that occur mostly in individuals with Systemic Juvenile Idiopathic Arthritis (SJIA) and those with adult-onset Still disease. The condition is thought to be caused due to excessive activation and expansion of T lymphocytes and macrophage histiocytes that show hemophagocytic activity. The expansion of these cells leads to a massive systemic inflammatory response associated with three cardinal features: Cytopenias, liver dysfunction, and coagulopathy, resembling Disseminated Intravascular Coagulation (DIC). MAS incidence in SLE patients is estimated to be 0.9% - 4.6%, and it is widely under recognized and underreported. In this report, we enumerate one such case with their clinical presentation, evaluation, and management.

Case

A 07-year-old girl presented with a history of intermittent fever with evening rise of temperature for the last 20 days; fever was associated with an erythematous rash over her face, arms, and thighs, reduced appetite, generalized weakness, and loss of 4kg weight over the last month. There was no significant past medical history or family history. On physical examination, she was conscious, active, and oriented to time, place, and person. She was afebrile with a pulse of 114/min, BP 114/80 mm of Hg, respiratory rate of 26/min; she was dehydrated with significant pallor, and bilateral neck lymphadenopathy. Facial puffiness was present. Mild splenomegaly was present. Lab reports showed severe microcytic anemia (4,7 g/dL, MCV 48.4, MCH 16.5, MCHC 34.1), thrombocytopenia (1,58,000 cells/µL), normal lymphocyte count (4300 cells/µL), ESR 110, CRP 6.3, hypofibrinogenemia (570mg/dl), increased Lactate Dehydrogenase 835 IU/L. Her PTT was 28 sec, and her INR was 1. The blood levels of albumin, BUN, serum creatinine, AST, ALT, and total bilirubin were normal. Urinalysis showed proteinuria (+++). Epithelial cells 2-3cells/HPF, pus cells 8-10/HPF, hyaline casts, Urine Protein creatinine ratio 10.54. Serological tests for HBV, HCV, HIV were normal. Blood and urine cultures were negative. The patient was positive for ANA, anti-dsDNA (200IU/ml); her serum C3 was low (23mg/dl). Her USG abdomen showed moderate splenomegaly with mild free fluid in the abdomen. Bone marrow aspirate showed erythroid hyperplasia, hypercellular forage with evidence of hemophagocytosis. The patient was diagnosed as a case of SLE with MAS. She was managed with transfusion of 300 ml of packed RBC over three days along with IVIG 22.5 gm over 24 hrs, intravenous methylprednisolone (30mg/kg/day) for three days, followed by prednisolone (1mg/kg) and cyclophosphamide (500mg/month) for six months. The hospital course was complicated due to the development of two episodes of seizures on day 4 of management. The patient was managed with IV Lorazepam and IV Phenytoin. The patient showed significant improvement since day five and was discharged on day 14. The patient improved with this management and remained well after a two-year follow-up [1-5].

Discussion

Macrophage activation syndrome is an inflammatory state caused by the activation of macrophages and T cells. These cells produce excess amounts of inflammatory cytokines like IFNγ, TNF, IL-1, IL-6, IL-10. MAS usually presents with fever, pancytopenia, hepatomegaly, splenomegaly, CNS symptoms. The patient may develop a rash, epistaxis, hemoptysis, or GI bleeding. Lab findings include elevated liver enzymes, hypofibrinogenemia, elevated triglyceride, and ferritin levels with an abnormal coagulation profile. Bone marrow aspirate shows evidence of haemophagocytic cells [6-10].

HLH diagnostic criteria by International Histiocyte society:

The diagnosis of HLH is established when one of the following two criteria are met:

- A molecular diagnosis consistent with HLH (mutations found in either PRF1 or MUNC13-4)

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- Diagnostic criteria of HLH is fulfilled (at least five of the eight criteria listed below are present)

✓ Persistent fever
✓ Splenomegaly
✓ Pancytopenia

  - Hemoglobin <90g/L
  - Platelets <100 x 10^9/L
  - Neutrophils < 1.0 x 10^9/L

- High triglyceride and/or low fibrinogen
- Hemophagocytosis in bone marrow, spleen, lymph node without any evidence of malignancy
- Serum ferritin ≥500 µg/L
- Low or absent NK cell activity
- High serum sIL2Rα

Our patient met five criteria, genetic testing, NK cell activity, and serum sIL2Rα were not available at our center.

Conclusion

MAS in SLE is an under recognized and underreported but potentially life-threatening complication that can be found at any age group and, as in our case, can be the first symptom of SLE. Early diagnosis and prompt therapy are essential in such cases to prevent adverse patient outcomes.

Conflict of Interest

The author declares that there are no conflicts of interest.

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