STREPTOCOCCUS MITIS INFECTION PRESENTING AS HLH: Case Report

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ABSTRACT

Hemophagocytic lymphohistiocytosis (HLH) is a rare but aggressive and life-threatening condition characterized by excessive immune activation. The syndrome of HLH was first described in 1939¹. It can be associated with various infections. HLH-94 or HLH-2004 protocol are employed for the treatment of HLH.

Keywords: HLH, IAHLH

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INTRODUCTION

Hemophagocytic lymphohistiocytosis (HLH) is a rare but aggressive and life-threatening condition characterized by excessive immune activation. This can be seen in cases of all ages though frequently seen in paediatric age. HLH can be inherited or acquired. The syndrome of HLH was first described in 1939^{1,2}.

Infection Associated Haemophagocytic Syndrome (IAHS) can be associated with viruses, fungi, mycobacteria, and parasites. A number of bacteria can be associated with HLH (IAHS).^{4,6}

CASE REPORT

A 16-year-old female was admitted to hospital with 2 weeks history of sore throat, fever, right knee, and elbow and shoulder joint pain, general weakness. The examination revealed a temperature of 102° F, Pulse 102, RR 26/m and right knee tenderness and swelling (Arthritis), right elbow/shoulder tenderness with no swelling. On investigation HB 8.9gm/dl, Leucocytosis (16.7), CRP: positive, ASO: 4100, USGe/o mild pleural effusion and minimal pericardial effusion. Echo mild MR and mild TR.

The patient was labeled as Rheumatic fever and managed with methyl prednisolone along with antibiotics and supportive care. The patient improved and was discharged. But in view of persistent symptoms, the patient was again admitted to the same hospital. Investigation revealed: ANA: Positive, Antacids DNA: equivocal, Antismith: Negative. The patient was labeled as SLE and was started on Hydroxychloroquine (HCQ), oral steroids, IV antibiotics, and supportive treatment and referred to SKIMS Rheumatology for further management. Examination revealed: Pallor: *Present*, Icterus: *Present*, Right knee & Elbow tenderness.

Investigations revealed Hb: 9.1, TLC: 3.2, Platelets 84000, Bilirubin: 2.6, AST: 795, LDH: 4916, Tg: 370, Serum Ferritin: 2000.

In view of the patient's clinical details and investigations, three possibilities were made:

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1. SLE with autoimmune hepatitis
2. Systemic JIA
3. HLH

The patient was started on IV Antibiotics (Piperacillin-tazobactam and amikacin) and Oral Steroids (1mg per kg body weight).

On further evaluation, Blood Culture: *Positive for Streptococcus Mites*. Brucella and Widal serology: *Negative*, Indirect and Direct Comb's Tests: *Negative*, Urine Culture: *Sterile*. Echocardiography: *Mild MR, Mild PAH, Mild TAR*, Bone marrow: Hemophagocytosis, NK Cell Activity: 0-79% (3-22%)

On the basis of clinical presentation, investigations, and response to treatment, the patient was diagnosed as a case of secondary HLH (Post-Streptococcal mitis)⁵ by fulfilling the diagnostic criteria for HLH³.

DISCUSSION

A variety of infecting organisms can be responsible for HLH. Viruses, bacteria, fungi, and parasites could lead to the development of HLH. Viruses account for a majority of Infection associated IAHS. The bacteria responsible for HLH reported are Brucella, gram-negative bacteria and tuberculosis. Streptococcus Mitis associated HS is not reported as an etiology in IAHS till date. Streptococcus mitis is alpha-hemolytic variable or non-groupable viridans streptococcus, part of the normal flora of the mouth and sometimes responsible for the development of dental caries⁷. HLH -94 or HLH-2004 protocol are employed for

the treatment of HLH. HLH-2004 is a modification of HLH-94 in which cyclosporin used in the induction phase and hydrocortisone is added to the intrathecal methotrexate.

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