INTRODUCTION:

Hemophagocytic lymphohistiocytosis(HLH) is a life threatening and an aggressive condition of excessive immune activation.It was first described as “familial hemophagocytic lymphohistiocytosis”(FLH) in 1952.HLH is of two types,primary HLH and secondary HLH.Primary HLH occurs with mutation of FLH genes or immune deficiency syndrome related genetic disorders.Secondary HLH is a sporadic or acquired disorder.Infection is a common trigger in both the types.Among infections,viral pathogens being the most common,bacterial,parasitic and fungal infections may also trigger this condition.

It is a fatal syndrome with high mortality,so delaying treatment may be life threatening to the patient.Diagnostic criteria requires five of the eight criterias as per HLH 2004 trial.These criterias are 1)fever>38.5c 2)splenomegaly 3)bicytopenia 4)hypertriglyceridemia or hypofibrinogenemia 5)hemophagocytosis in bone marrow,spleen,lymph node or liver 6)low or absent NKcell activity 7)Ferritin>500ng/ml 8) Elevated soluble C025 (soluble IL-2 receptor alpha) two standard deviations above age-adjusted laboratory-specific norms (> 2400 U l/ml).Other genetic and immunological profile testing can be performed.Our report describes a case of HLH triggered by Plasmodium vivax infection treated successfully with steroid and antimalarial agents.

CASE REPORT:

A 22yrs old female primi in 1st trimester of her pregnancy presented to us with history of high grade fever with chill and rigor since last 10days,associated respiratory discomfort since 2 days.She was conscious,obeying hemodynamically stable.She was admitted in ICU examined and investigated.On initial investigation she was found to have Hb 7.5gm/dl TC 1800 per cumm,ESR 100,platelet count 20000,Plasmodium vivax rapid test positive,PBS for MP-many dividing schizonts and few ring trophozoites of plasmodium vivax seen,S.bilirubin 3.10,Direct 1.90,SGOT 70,SGPT 34, INR 1.3,S.creatinine0.6 Chest x ray showed bilateral opacities.Patient was initially kept on NIV but later on intubated in view of persistent hypoxia and respiratory distress.Patient was ventilated in pressure control mode as per ARDS protocol.Patient was started on antimalarial drug i.v artesunate,antibiotic and other supportive medications.I.v fluid was used judiciously.2D ECHO done which was normal.Urine,blood and tracheal cultures were negative for any pathogen.Patient was transfused 4 units of platelet(RDP) as she was having mucosal bleeding.Repeat

platelet count 30000 TLC 3500,Hb 6.7.USG abdomen showed hepatosplenomegaly,GB sludge,with single live intrauterine16wks pregnancy in variable presentation.She was given two units of blood transfusion.Patient became hemodynamically unstable for which noradrenaline was started. Patient had spontaneous abortion.Repeat cultures from suspected sites were negative.Blood sample for D dimer,serum ferritin,serum triglyceride,serum fibrogen level were sent suspecting it to be secondary HLH.D dimer level 980ng/ml(high),fibrinogen179.40ng/100ml(low),ferritin>1000 (high),triglyceride 244mg/dl(high).Bone marrow examination was planned but attendant refused.Patient was started on intravenous hydrocortisone 100mg TDS.Patient's condition started improving from 2nd day of starting steroid.Her platelet count went up to 78,000,TC 4,400.She was off vasopressor and progressively weaned and extubated the next day.Patient was later on shifted out of ICU within 2 days with steroid tapered and changed to oral prednisolone.Patient later on had no parasitemia in the blood with serum ferrin in decreasing trend.

DISCUSSION:

In our case 1)fever,2)splenomegaly,3)bicytopenia,4)hypertryglyceridemia with hypofibrinogenaemia and5)hyperferritinemia were present meeting 5 out of 8 criterias of HLH 2004 trial.Further confirmation could not be done as bone marrow examination was not allowed by the attendant of the patient.But meeting 5 of the criterias with high suspicion of secondary HLH associated with malaria,patient was started on steroid.Patient showed remarkable response to steroid with significant improvement within 3 days.

HLH is a life threatening condition and so early clinical suspicion in a rapidly detoriating patient even in the absence of typical HLH 2004 trial ,will result in timely referral to a hematologist and may change the outcome of this highly morbid and fatal syndrome.

References

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