Weathering a cytokine storm: A Case of Hemophagocytic Lymphohistiocytosis

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Introduction

Hemophagocytic Lymphohistiocytosis (HLH) is a rare and life-threatening disease caused by excessive immune activation with histiocytic infiltration. It can mimic many diseases, some of which also trigger HLH including infection and malignancy. As such, the true incidence and prevalence of HLH in adults are unknown.

Case Report

A 29-year-old Hmong male presented with a six month history of abdominal pain, substantial weight loss and weakness. On presentation he was alert but severely cachectic, jaundiced with a blood pressure of 72/46. Physical examination revealed diffuse petechiae and splenomegaly.

Labs
- AST 504 U/L, ALT 247 U/L
- TBili 13.9 mg/dl
- INR 1.89
- Crt 1.67 mg/dl
- HIV, Quantiferon, sputum AFB smears neg
- Ferritin 13444 ng/ml
- Fibrinogen 104mg%
- Soluble IL-2 R 10210 pg/ml

Abdominal CT (figure 1) demonstrated splenomegaly, multiple liver and splenic lesions and lymph node mass.

His bone marrow biopsy was found to be hypocellular with histiocytic prominence, rare hemophagocytosis and many EBV positive cells.

Figure 1: Abdominal CT (figure 1) demonstrating splenomegaly, multiple liver and splenic lesions and lymph node mass.

Figure 2: Bone marrow smear Giemsa stain showing hemophagocytosis.

Figure 3: Bone marrow core biopsy EBER in situ hybridization.

Images provided by Mingyi Chen, MD, Ph.D, Department of Pathology, UC Davis Medical Center

Diagnostic Criteria of HLH

- Fever
- Splenomegaly
- Pancytopenia – Hgb<9 g/ml, Plt <100, Neutrophils <1 x 10^9 L/min
- Hypertriglyceridemia (≥265 mg/100 ml) and/or hypofibrinogenemia (≤ 150 mg/100 ml)
- Ferritin ≥ 500 ng/ml
- Hemophagocytosis in the BM, spleen or lymph nodes
- Low or absent NK cell activity
- Elevated soluble IL-2 receptor

The patient was started on IV dexamethasone and Rituximab, after 5 days of treatment he continued to decline and ultimately passed away.

Discussion

- HLH should be considered in patients who present with shock and multi organ failure, especially involving liver and bone marrow.
- To help identify HLH, a significantly elevated ferritin can be highly specific.
- Early diagnosis is imperative so treatment can be promptly initiated.

Resources: