

Hemophagocytic lymphohistiocytosis in a patient with worsening weakness

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Abstract

Hemophagocytic lymphohistiocytosis is an under-recognized clinical entity that can present in adults. It is important to consider in the differential diagnosis of patients presenting with non-specific multi-organ dysfunction and inflammation.

Keywords:

Hemophagocytic lymphohistiocytosis, HLH, macrophage, inflammation, ferritin

Key Clinical Message:

Hemophagocytic histiocytosis is important to consider in the setting of multi-organ dysfunction, especially with an elevated ferritin and evidence of HLH on bone marrow biopsy.

Manuscript:

A 59-year-old man presented to the hospital for progressive weakness. Medical history was notable for acute inflammatory demyelinating polyneuropathy for which he was receiving intravenous immune globulin. Physical exam demonstrated 2/5 strength in bilateral lower extremities and absent ankle and knee jerk reflexes. He was admitted to the hospital and rapidly developed fevers, acute renal failure, thrombocytopenia and coagulopathy. Serum ferritin was elevated at 32400 ng/mL (normal range, <388). Hemophagocytic lymphohistiocytosis (HLH) was suspected and a bone marrow biopsy was performed. It showed prominent macrophage hemophagocytosis (figures 1 and 2), supporting the diagnosis. HLH is an aggressive macrophage activation syndrome. In both children and adult patients primary and secondary forms are known, the latter being more common and often associated with infections and malignancy [1]. Five out of eight diagnostic criteria are needed to be classified as HLH, including a fever greater than or equal to 38.5 degrees Celsius, splenomegaly, peripheral blood cytopenias, hypertriglyceridemia, low or absent natural killer cell activity, elevated soluble CD25 two standard deviations above age-adjusted laboratory-specific norms, bone marrow hemophagocytosis and serum ferritin > 500 ng/mL [2]. The patient died prior to initiation of therapy.

Author Contributions:

Edward Nabrinsky: writing and editing of manuscript, selection of figures

Ronald Sirota: editing of manuscript, selection of figures

Jason Macklis: editing of manuscript

References

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