

# Hemophagocytic lymphohistiocytosis: macrophages engulfing red cells and neutrophils

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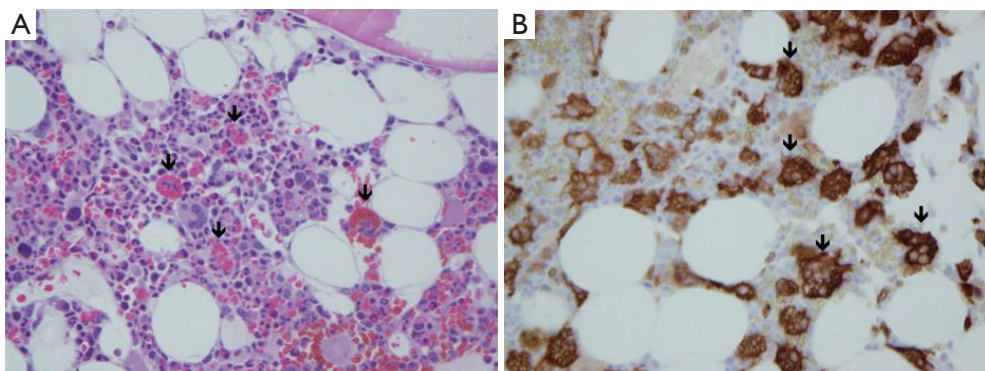
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## Case presentation

A 47-year-old female presented with a 3-day history of high grade fever, myalgia, abdominal discomfort, vomiting, shortness of breath, headache, and altered mental status. She was hypotensive with acute respiratory distress syndrome and multi-organ failure. Laboratory workup was significant for pancytopenia, elevated liver enzymes, acute kidney injury, elevated creatine kinase levels and ferritin level of 10,000 mcg/L. She was also noted to have hypertriglyceridemia, elevated D-dimer and low fibrinogen levels. All cultures of blood, urine, bronchoalveolar lavage fluid and cerebrospinal fluid were negative. Imaging did not reveal any hepatosplenomegaly,

lymphadenopathy or neurologic involvement. All rheumatologic workup, hepatitis B, C and HIV were negative. A bone marrow examination was undertaken to rule out hemophagocytic lymphohistiocytosis (HLH). Bone marrow was mildly hypercellular. There were many areas in the bone marrow with appearance of clusters of RBCs mimicking foci of fresh hemorrhage (arrows in panel A, H&E staining). Bone marrow flow cytometry was unremarkable. Immunohistochemical staining for CD163 (hemoglobin-haptoglobin scavenger receptor) was done to evaluate the possibility of HLH. This stain highlighted many CD163<sup>+</sup> macrophages (arrows in panel B) engulfing hematopoietic cells (mostly red cells and occasional



neutrophils), in the bone marrow, which were mimicking as foci of hemorrhage with H&E staining. This case exemplifies the importance of CD163 staining for cases with high clinical suspicion of HLH. Without CD163 immunostaining, this diagnosis can be missed, especially in subtle hemophagocytosis. The patient received 6 sessions of plasmapheresis and was started on HLH-2004 protocol with etoposide, high dose dexamethasone and cyclosporine with good clinical response and gradual improvement of all her organ functions in addition to the dramatic

lowering of serum ferritin levels.

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### **Footnote**

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

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