BEYOND RICHTER'S: A CASE OF HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS FROM ANAPLASMA IN A PATIENT WITH CHRONIC LYMPHOID LEUKEMIA

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Introduction

• Hemophagocytic Lymphohistiocytosis (HLH) is a life-threatening hyperinflammatory syndrome triggered by excessive immune activation.
• HLH is characterized by excessive activation of macrophages and T-cells, leading to widespread phagocytosis of blood cells. This can result in cytopenias, hepatosplenomegaly, hyperferritinemia, and elevated soluble IL-2 receptor levels.
• Infections, malignancies, or autoimmune diseases can induce secondary HLH. Tick-borne infections, such as Anaplasma phagocytophilum, although rare, can precipitate HLH.
• Ferritin >10,000 ng/mL should raise suspicion for HLH.

Case Description

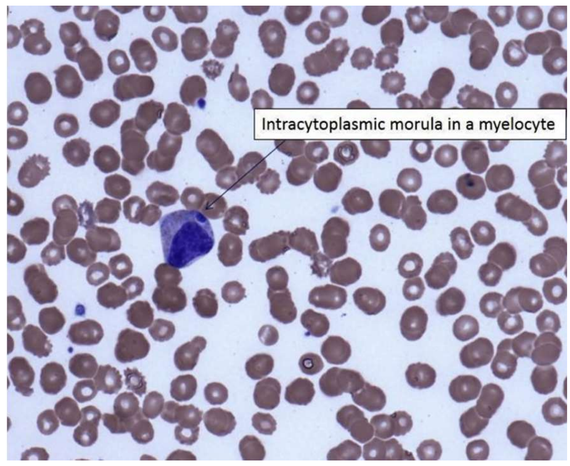
Patient Profile: 69-year-old female PMHx of CLL presented with confusion, fever, abdominal discomfort, hepatosplenomegaly, and mixed acid-base disorder
Diagnostic Workup:
• CBC: Bicytopenia
• Ferritin: 18,648
• triglycerides >372 mg/dL
• PCR: Positive for Anaplasma phagocytophilum.
• Peripheral smear at MGB with intracellular morulae
• Bone marrow biopsy at MGB: No hemophagocytosis.

HLH Diagnostic Criteria Fulfilled (6/8):
• Fever
• Splenomegaly
• Cytopenias
• Hypertriglyceridemia
• Elevated ferritin
• Elevated Il-2

Treatment:
• Initiated targeted doxycycline therapy for Anaplasma.
• Supportive care, including transfusions and monitoring.
• No immunosuppressive therapy required due to rapid clinical improvement.

Outcome:
• Significant recovery within 72 hours of targeted therapy.
• Complete resolution of symptoms and normalization of blood markers within 14 days.

Image



Peripheral blood smear showing intracellular morulae of *Anaplasma phagocytophilum*, the infectious trigger of HLH in this case

Discussion

•HLH may present with non-specific symptoms and can mimic sepsis or malignancy
• Ferritin >10,000 is a key red flag in identifying HLH.
• History is key to identifying triggers such as infections.
• Early intervention may avoid the need for immunosuppression.
•Maintain a high index of suspicion for HLH in cases of unexplained systemic inflammation.
• In endemic areas, consider tick-borne pathogens as potential HLH triggers.
• The HLH syndrome overall mortality rate remains high at 41%
•Delayed diagnosis may result in severe complications, including acute respiratory distress syndrome (ARDS) and multi-organ failure.
•Immunocompromised patients—including those with malignancies, organ transplants, or autoimmune conditions—are particularly vulnerable to severe forms of HGA and are at higher risk for developing HLH.

References

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•Janka GE. **Hemophagocytic Syndromes in Children and Adults.** *Hematology Am Soc Hematol Educ Program.* 2005.