**ABSTRACT**

Hemophagocytic Lymphohistiocytosis (HLH) is a rare disorder of pathologic immune activation and uncontrolled cytokine release that causes life-threatening multi-system failure. The diagnosis of HLH is often missed or delayed due to its variable presentation and nonspecific symptoms. Here, we present a case of HLH secondary to B-cell lymphoma with initial presentation of acute hypoxic respiratory failure.

**RESULTS**

With patient remaining encephalopathic and becoming febrile. Further workup was done and revealed a ferritin level of >40,000 ng/mL (ref range 22 - 322 ng/mL) and interleukin-2 level of 34871.4 pg/mL (ref range 175.3 - 858.2 pg/mL). Other concerning labs were; hypertriglyceridemia, 452 and hypofibrinogenemia, 137.

A bone marrow biopsy (posterior iliac crest; aspirate and core biopsy) was immediately done and resulted as, “Neoplastic Large B-cell Proliferation (<5% of cellular marrow) accompanied by HLH”. Neoplastic cells express CD20, CD79a, PAX-5, BCL-2, BCL-6 and MUM-1.

**DISCUSSION**

1. Our patient met the diagnostic criteria based on the HLH-2004 and Modified 2019 guidelines, at least five out of the following nine diagnostic criteria for HLH:
   I. Fever
   II. Splenomegaly
   III. Cytopenias
   IV. Hypertriglyceridemia
   V. Hypofibrinogenemia
   VI. Elevated ferritin
   VII. Hemophagocytosis in bone marrow/spleen/lymph nodes
   VIII. Low/absent natural killer (NK)-cell activity
   IX. Elevated soluble CD25 (interleukin [IL]-2 receptor)

2. Our patient was treated with Dexamethasone, Etoposide, and Rituximab. Intrathecal Methotrexate put on hold due to persistent thrombocytopenia.

3. Due to the rarity of HLH, the true incidence is unknown and life-saving treatment is often delayed.

4. Our case highlights the importance of including HLH in the differential diagnosis of patients presenting with multi-organ involvement and elevated inflammatory markers.

**REFERENCES**


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