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Hemophagocytic Lymphohistiocytosis, An Overlooked Culprit of Disseminated Intravascular Coagulation

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Introduction

Hemophagocytic lymphohistiocytosis (HLH) is an immune system disorder with aberrant activation of T cells and macrophages resulting in vast tissue destruction. There are two forms of HLH: inherited HLH (autosomal recessive) and secondary HLH, occurring after strong immunologic activation.

Primary HLH results from defective proteins involved in the cytolytic secretory pathway. These defective proteins prevent cytotoxic T lymphocytes and Natural Killer (NK) cells from maintaining immune homeostasis.

In contrast, secondary HLH results from upregulation of proinflammatory cytokines which results in overactivation of T lymphocytes and NK cells. However, the end result of both primary and secondary HLH is the same: cellular death and organ failure.

Case Presentation

A 56 y.o. female with a history of Essential thrombocytosis (ET) s/p hydroxyurea treatment presented to the ED with an episode of gross hematuria. She was hypertensive and febrile. Physical exam was significant for petechiae on all extremities, tachycardia, diffuse abdominal tenderness, and splenomegaly.

Considering her patient’s history of ET, which has a predisposition to progress to Acute Myeloid Leukemia, and long term exposure to hydroxyurea (a leukemogenic agent) suspicion for leukemia was very high.

However, a low fibrinogen level indicated DIC. It wasn’t clear as to whether the DIC resulted from bacteremia or acute promyelocytic leukemia (APL), a variant of AML known to precipitate DIC. As such, a bone marrow biopsy was conducted for further analysis.

A closer re-examination of bone marrow biopsy revealed histiophagocytic behavior (Figure D). Such morphological presentation raised the suspicion for HLH. As such, ferritin and triglyceride levels were ordered which revealed a level of >12,000 and 226, respectively.

A diagnosis of HLH was made considering negative blood cultures, biopsy findings, hyperferritinemia and hypertriglyceridemia. Pt was started on HLH-94 Protocol along with idarubicin and cytarabine for AML induction therapy.

Bone Marrow Biopsy

Figure A. The bone marrow biopsy is noted to be hypercellular. With process of age, bone marrows are gradually replaced with adipose. Considering patient’s age, the bone marrow biopsy report is consistent with HLH. A detailed analysis is suggestive of leukemia.

Figure B. Under magnification, Arrows point to myeloblasts. Suggestion of acute myeloblastic leukemia (AML).

Figure C. Anti-CD20 stain. The stain tests for CD20, a cell surface protein found on myeloblast.

Figure D. Arrows point to macrophages phagocytosing neutrophils; note the large vacuoles through red pathogenesis, such activity is suggestive of hemophagocytic lymphohistiocytosis.

Discussion & Follow-up

DISCUSSION:

This presentation illustrates a transformation of ET into AML with concurrent HLH: highlighting the importance of considering HLH in AML patients presenting with DIC, as early detection could be essential in managing both patients’ malignancy and hemodynamic status.

FOLLOW-UP:

- Patient was started on HLH-94 Protocol (etoposide and tapering steroids) along with idarubicin and cytarabine; subsequently platelets increased to 37,000, and both ferritin and fibrinogen normalized.
- Upon stabilization, pt was discharged and scheduled for follow up by Hematology Oncology at Penn State University for bone marrow transplant.

REFERENCES:

