Hemophagocytic Lymphohistiocytosis

Hemophagocytic lymphohistiocytosis is a life-threatening hematologic disorder involving severe inflammation due to increased activity of cytotoxic T cells and macrophages which leads to cytokine storm. It can be acquired or sporadic. Clinical features include fever and hepatosplenomegaly. Laboratory findings include increased serum ferritin and pancytopenia. Definitive management involves bone marrow transplantation.



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Characteristics

Excessive Immune System Activation
Excessive Immune-Moon
In HLH, there is excessive activation of CD8+ T-Cells. Most commonly, this occurs due to Epstein-Barr virus infection.

Cytotoxic T-cells and Macrophages
Side-toe-toxic-green Tennis-ball and Mac-man
CD8+ T-cells release IFN-? that in turn activates macrophages. This disrupts the immune homeostasis and phagocytosis of all blood cells takes place.

Cytokine Storm
Side-toe-kite in Storm
The cross-talk between various cells of the immune system forms an auto-amplification loop. This cross-talk is by pro-inflammatory cytokines primarily TNF-alpha, IL-1, and IFN-gamma. IL-6 and IL-12 are also implicated.

Acquired or Sporadic
Acquired-magnet and Sporadic-spear
Sporadic cases are associated with Chediak-Higashi syndrome. It can be acquired secondary to EBV infection or malignancy, usually lymphomas.

Presentation

Fever
Fever-beaver
Fever is typically seen in hemophagocytic lymphohistiocytosis. This is due to massive interleukin-1 release, which raises the temperature setpoint of the hypothalamus.

Hepatosplenomegaly
Liver-and-spleen-balloons
Hepatosplenomegaly can occur from extramedullary hematopoiesis because the medullary space is undergoing increased hemophagocytosis.

Labs

Increased Ferritin
Up-arrow Ferret-tin
Serum ferritin is an acute-phase reactant that is upregulated by TNF-alpha. It is characteristically elevated in patients with HLH.

Pancytopenia
Pan-side-toe-peanut
Pancytopenia is defined as a decrease in the number of all cell lines in the blood i.e erythrocytes, leukocytes, and platelets. It is due to phagocytosis.

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Management

Bone Marrow Transplantation

Bone Train-plant

Immunosuppressants are used to control cytokine storm. Hematopoietic stem cell transplantation is potentially curative and is the only definitive treatment.

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