Myelodysplastic Syndromes

Myelodysplastic syndromes (MDS) are a group of hematological cancers in which malfunctioning pluripotent stem cells lead to hypercellularity and dysplasia of the bone marrow. MDS usually affects elderly patients or those who have had previous radiation or chemotherapy. Clinical features vary depending on the type of MDS and the affected cell lines but may include signs of anemia, recurrent infections, and petechial bleeding. Peripheral blood smear may show ringed sideroblasts, Howell-Jolly bodies, or a pseudo-Pelger-Huet anomaly, which are bilobed neutrophils. Management principles include supportive care and bone marrow transplantation. In 30% of cases, MDS may progress to acute myelogenous leukemia.



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Characteristics

Dysfunctional Bone Marrow
Bone Marilyn-Monroe Dysfunction
Myelodysplastic syndromes are characterized by dysfunctional bone marrow. Germline or acquired mutations in the genes responsible for proliferation can lead to hypercellularity and dysplasia of the bone marrow.

Elderly
Old-person
Myelodysplastic syndromes are more common in the elderly. As we age, we accumulate mutations in our pluripotent stem cells. With sufficient time and replication cycles, cancer will develop.

Radiation or Chemotherapy
Radiation-radio and Chemo-head-wrap
Radiation therapy is a medical treatment that employs ionizing radiation to kill abnormal cells or control their growth. Chemotherapeutic agents, also referred to as antineoplastic agents, are used to directly or indirectly inhibit the uncontrolled growth of cancer cells. However, these highly toxic agents may also lead to the development of certain neoplasms. Radiation and chemotherapy may accelerate the rate of mutations in cells.

Presentation

Anemia
Anemone
Anemia is characterized by a decrease in the number of circulating red blood cells (RBC), represented by a reduction in hemoglobin concentration (Hb), hematocrit (Hct), or RBC count. It can occur in myelodysplastic syndromes due to dysplasia of the bone marrow. Hyperproliferation of bone marrow may cause "crowding out" of erythrocyte precursors.

Increased Susceptibility to Infection
Up-arrow Infectious Virus and Bacteria
In patients with myelodysplastic syndromes, dysplasia of the bone marrow may lead to increased infection susceptibility because white blood cells are affected. A decreased innate immune response predisposes patients to infections.

Bleeding
Bleeding
Bleeding disorders are characterized by defects in hemostasis that lead to an increased susceptibility to bleeding. Patients with myelodysplastic syndromes may present with thrombocytopenia, predisposing them to bleeding.

Diagnosis

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Ringed Sideroblasts

Ringed Cinder-block

Ringed sideroblasts are a type of erythrocyte characterized by a perinuclear ring. This mitochondrial coloration is due to iron, so is best visualized with a Prussian Blue stain.

Howell-Jolly Bodies

Howling-Jolly Santa

Howell-Jolly bodies are seen in the blood smear of patients with myelodysplastic syndromes. They are a collection of basophilic remnants of DNA found in circulating immature RBCs that are normally removed by the spleen.

Pseudo-Pelger-Huet Anomaly

Sumo-player-duet with Animal

Pseudo-Pelger-Huet anomaly can be seen in patients with myelodysplastic syndromes. It is characterized by atypical granulocytes on peripheral blood smear, often with bilobed nuclei.

Bilobed Neutrophils

Nude-trojan with 2 Balloons

The pseudo-pelger-huet anomaly is characterized by neutrophils with bilobed or dumbbell-shaped nuclei on peripheral blood smear.

Management

Supportive Care

Supportive IV bags

Myelodysplastic syndromes are typically refractory to treatment. While bone marrow transplant may lead to remission, not all patients are candidates. This disease often affects elderly people, who are often poor candidates for such invasive treatment. As such, patients are typically managed supportively.

Bone Marrow Transplant

Bone Train-plant

Bone marrow transplant may be curative or achieve remission, but not all patients are candidates. Patients must first undergo bone marrow ablation via chemotherapeutic agents, followed by the transplantation of healthy bone marrow.

Considerations

Acute Myelogenous Leukemia (AML)

American Monkey Legion (AML)

Acute myelogenous leukemia (AML) is a malignant neoplastic disease that arises from myeloid cell lines. It is characterized by the proliferation of immature, nonfunctional cells in the bone marrow that are subsequently released into the bloodstream. It occurs with greater frequency in patients with myelodysplastic syndromes.

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