Primary Myelofibrosis

Primary myelofibrosis (PMF) is a type of cancer of bone marrow and is characterized by bone marrow fibrosis, extramedullary hematopoiesis, and JAK2 tyrosine kinase mutations. It can present with fatigue, weight loss, and splenomegaly. Pancytopenia, dacrocytes, and dry tap on bone marrow aspiration are diagnostic clues. Primary myelofibrosis treatment options include stem cell transplantation, transfusion, and ruxolitinib.



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Characteristics

Bone Marrow Fibrosis
Bone Arrow Fibrous-sack
Bone marrow fibrosis is characterized by the formation of fibrous/scarring tissue in bone marrow leading to abnormalities in the formation of blood cells, including red blood cells, white blood cells, and platelets. Abnormalities in the DNA of a hematopoietic stem cell can lead to overproduction of abnormal cells. The overproduction of abnormal cells will result in a shortage of normal blood cells.

Extramedullary Hematopoiesis
Ex-Medusa He-man-top-hat
Extramedullary hematopoiesis occurs over the course of primary myelofibrosis. Due to bone marrow failure, accessory organs participate in hematopoiesis. These organs are primarily the spleen, liver, and lymph nodes.

JAK2 Kinase Mutation
Jack in (2) Tutu Kite-ace Mutant
JAK2 kinase mutations are the most common mutations in patients with PMF. The JAK2 gene is involved in the JAK/STAT pathway that works by promoting cell division and growth. These mutations result in the overproduction of abnormal cells squeezes out normal blood cells. Other gene mutations include CALR and MPL.

Symptoms

Fatigue
Sleepy-guy
Fatigue is the most common symptom. It occurs due to anemia and cytokines. Other symptoms include bone pain, fever, weight loss, and night sweats.

Weight Loss
Skinny-with-baggy-pants
Weight loss is multifactorial in etiology. These etiologies include abdominal pain, increased proinflammatory cytokines, decreased oral intake, and abdominal organ enlargement, which compresses the stomach and leads to early satiety.

Splenomegaly
Spleen-balloon
The spleen works as the primary site of extramedullary hematopoiesis. This results in the enlargement of the spleen (splenomegaly). This enlargement can lead to several other symptoms, such as left upper abdominal pain and early satiety/fullness of the stomach due to compression. Liver and lymph node enlargement can also occur due to the extramedullary hematopoiesis. Additionally, patients with splenomegaly are at increased risk of rupture.

DIagnosis

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Pancytopenia

Pan-side-toe-peanut

Fibrous bone marrow formation interrupts normal blood cell production, resulting in anemia, leukopenia, and thrombocytopenia i.e. pancytopenia. The anemia is normocytic and normochromic. It is often severe with a hemoglobin value of less than 10 g/dL. Leukopenia and thrombocytopenia lead to increase risk of infection and bleeding, respectively.

Dacrocytes (Teardrop Cells)

Dracula-cry

Peripheral blood smear of patients with primary myelofibrosis can show dacrocytes, which results from red blood cells squeezing out from fibrotic tissue on bone marrow. Another morphology that can be present in a blood smear is nucleated RBCs and immature granulocytes. This morphology results from leukoerythroblastosis in the bone marrow.

Dry Tap on Bone Marrow Aspiration

Dry Tap from a Bone Marrow Aspirating-ass

Dry tap on bone marrow aspiration happens from fibrosis. When the physician attempts to draw bone marrow, there is little to be aspirated.

Treatment

Stem Cell Transplantation

Steam-cell-train-plant

Allogeneic stem cell transplantation is a potential treatment for primary myelofibrosis. It is beneficial in patients with intermediate or high-risk disease based on the prognostic scoring system from the International Working Group for Myelofibrosis Research and Treatment.

Transfusion

Transfusion-IV

Blood transfusions with packed red cells are indicated in patients with severe anemia. Additionally, symptomatic deficiencies in other cell lines may be treated in kind. Bleeding disorders due to low platelets can be treated with blood transfusions, and infections due to low leukocyte counts can be treated with leukocyte stimulating medications.

Ruxolitinib (JAK2 Inhibitor)

Rocks-light

Ruxolitinib works by inhibiting JAK1 and JAK2 (Janus Associated Kinases), which are involved in the signaling of cytokines and growth factors in hematopoiesis and immune functions. It is used to treat intermediate and high-risk myelofibrosis patients. The use of ruxolitinib has been shown to reduce splenomegaly and relieve symptoms on patients.

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