

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

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.

Dacrococytes (Teardrop Cells)

Dracula-cry

Peripheral blood smear of patients with primary myelofibrosis can show dacrococytes, 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.