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Myelofibrosis

Myelofibrosis (MF) is a chronic blood malignancy caused by abnormal blood cells built up in the bone marrow. Increased production of abnormal blood stem cells creates overproduction of megakaryocytes, which make platelets and fibroblast growth factor. Ultimately this causes the formation of fibrous tissue in the bone marrow. The fibrous tissue decreases the bone marrow's ability to make normal blood cells. Often, blood cell production then moves to other areas of the body such as the spleen and liver. Most patients exhibit symptoms of chronic inflammation due to "cytokines" released from the abnormal cells. MF is one of a group of rare bone marrow cancers called myeloproliferative neoplasms (MPNs) that develop due to a stem cell mutation in the bone marrow.

Statistics

- A majority of MF cases are diagnosed in people **over the age of 50**.
- About **50% to 60% of MF patients** have a mutation in the **Janus kinase 2 (JAK2) gene V617F**.
- About **20% to 35% of patients** have a Calreticulin (CALR) mutation.
- An MPL gene mutation is present in **5% to 10%** of patients with MF.

Risk Factors

The causes of the acquired genetic mutation seen in MF are not yet known.

- MF can develop after other myeloproliferative diseases such as polycythemia vera (PV) or essential thrombocythemia (ET) and is then designated as "secondary MF," which may also be referred to as "post-PV MF" or "post-ET MF."
- Researchers are exploring the link between myelofibrosis and various gene mutations, such as JAK2, CALR, and MPL.
- Exposure to petrochemicals and ionizing radiation may increase risk of developing myelofibrosis.
- Primary MF carries a worse prognosis than secondary MF.

Symptoms

Some patients with MF do not have symptoms when they are diagnosed, especially when the disease is in its early stages. Diagnosis in an early phase often occurs during a routine exam or blood test. Many MF patients will have the following symptoms.

- Fatigue, weakness, or shortness of breath
- Anemia
- Unexplained weight loss
- Night sweats
- Bruising or easy bleeding
- Enlarged liver
- Frequent infections
- Discomfort in upper left side of abdomen and an abnormally quick feeling of fullness at meals, caused by an enlarged spleen
- Bone pain, joint pain, or gout
- Fever and itching
- Portal hypertension, which may cause enlarged veins in the stomach and esophagus and bleeding from the bowel
- Growth of blood-forming cells outside the bone marrow, which can be painful
- Teardrop red cells in the blood
- Early blood cell precursors (nucleated red blood cells and mature white blood cells, normally only in the bone marrow) found in the blood

Prevention

MF cannot be prevented. Research is underway to learn more about how the disease develops.

Treatment

MF patients who do not experience symptoms typically do not receive treatment and may remain in stable health for years. However, if symptoms are present, treatment options include blood transfusions, hydroxyurea, ruxolitinib, and other similar medications such as interferon alfa, and less often, radiation, splenectomy, and stem cell transplants. Other treatment options may be available through clinical trials.

About Texas Oncology

With more than 530 physicians and 280 locations, Texas Oncology is an independent private practice, a member of The US Oncology Network, that sees more than 71,000 new cancer patients each year. Founded in 1986, Texas Oncology provides comprehensive, multidisciplinary care, and includes Texas Center for Proton Therapy, Texas Breast Specialists, Texas Colon & Rectal Specialists, Texas Oncology Surgical Specialists, Texas Urology Specialists and Texas Infusion and Imaging Center. Texas Oncology's robust community-based clinical trials and research program has contributed to the development of more than 100 FDA-approved cancer therapies. Learn more at TexasOncology.com.

Sources: Leukemia and Lymphoma Society, MPN Research Foundation, National Cancer Institute, National Library of Medicine, and National Organization for Rare Disorders

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