Myelofibrosis

Red blood cells, white blood cells, and platelets are produced within the bone marrow. When there is an injury to any part of the body, the connective tissue thickens and causes fibrosis or scarring. If the connective tissue thickens throughout the bone marrow, it compresses the healthy cells present there. This leads to decreased blood cell production resulting in anemia, a lack of white blood cells, and insufficient platelet production.

Myelofibrosis typically occurs in adults over the age of seventy. The exact cause of this disease is still unknown. Approximately 65% of patients are known to have a JAK-2 gene mutation. In myelofibrosis, there is insufficient space in the bone marrow to produce blood cells, so the body starts producing blood in the spleen. Consequently, the spleen gradually enlarges and, in advanced stages, may fill the entire abdomen. Symptoms experienced by these patients may include fever, night sweats, fatigue, weight loss, and itching. Some patients may also experience bone pain. If the platelet count is too low, bleeding may occur. In some patients, the disease can progress to AML, a highly dangerous blood cancer.

The presence of myelofibrosis is suspected when a blood sample is examined under a microscope (peripheral smear). In such cases, the doctor may collect a bone marrow sample and biopsy and send them for examination. The presence of fibrosis in these samples confirms the occurrence of myelofibrosis. Furthermore, studying genes such as JAK-2 helps in confirming the diagnosis of the disease.

The International Prognostic Scoring System (IPSS) calculates the average life expectancy of a patient by taking into account factors such as age, white blood cell count, hemoglobin level, number of blast cells in the blood, and weight loss. This calculation categorizes patients into low-risk, intermediate-risk, and high-risk groups. If the risk is low and the patient is asymptomatic, no treatment is necessary. The patient should undergo regular examinations every three to six months to assess the need for treatment. For low-risk patients experiencing symptoms, drugs such as ruxolitinib, interferon, and hydroxyurea are prescribed.

High-risk patients and low risk patients who do not respond to drug therapy require individualized treatment plans based on their overall health status. For patients in excellent health, a bone marrow transplant is the optimal treatment option. However, for patients with poorer health conditions, symptoms can be managed using medications. Drugs such as erythropoietin injection, danazol, thalidomide, and prednisolone can be used to address anemia. The drugs which improve hemoglobin level are continued for for prolonged duration. For high-risk patients with multiple complications, such as weight loss, ruxolitinib tablets may be prescribed. Ruxolitinib inhibits the JAK-2 gene and has shown positive results in

patients without the JAK-2 mutation as well. It significantly reduces fatigue, fever, weight loss, and spleen enlargement. Although the medication is costly and does not cure the disease, it effectively alleviates the symptoms. Patients taking this medication are at risk of reactivating hepatitis B and developing tuberculosis. If there is no improvement after three months of treatment, the medication is usually discontinued.

There is currently no cure for myelofibrosis other than a bone marrow transplant. However, approximately 50% of patients undergoing bone marrow transplantation die as a result of treatment side effects. Therefore, many patients manage the disease through medication.

If none of the drugs prove effective, blood and platelet transfusions become the only viable option. Removing an enlarged spleen can do more harm than good. After the surgery, there is a high risk of blood clot formation in the blood vessels, bleeding, and systemic infection. These complications can be life-threatening. Patients receiving treatment for myelofibrosis should consult a doctor every two to three months to optimize treatment based on its effects and side effects.

People with myelofibrosis do not need to isolate themselves as the disease does not spread from person to person. This disease is not a hereditary disease.

Ongoing research aims to develop new drugs for the treatment of myelofibrosis. These drugs are tested in clinical trials with patient participation, which not only contributes to the development of better medications but also provides patients with access to potentially beneficial treatments at no cost.

For more information about myelofibrosis, consult the team of doctors responsible for your treatment.

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