Polycythemia Vera

Polycythemia vera (PV) is a chronic blood cancer caused by an acquired gene mutation of the blood cells in the bone marrow, which creates an overproduction of blood cells, particularly red blood cells. White blood cells and platelets are often also increased. An excess amount of red blood cells can increase the blood viscosity (thickness), slowing blood flow. As blood flow is slowed, the risk blood clotting increases. Excess cells can grow in the spleen, causing it to become enlarged. PV is one of a group of rare blood cancers called myeloproliferative neoplasms (MPNs) that develop due to an acquired mutation in the DNA of the stem cells in the bone marrow. Conversion to myelofibrosis or acute leukemia can occur.

Statistics

- In every 100,000 people, there are approximately 0.4 to 2.8 people diagnosed with polycythemia vera annually.
- A majority of polycythemia vera cases are diagnosed in people over the age of 60.

Risk Factors

The exact causes of the acquired genetic mutation in polycythemia vera are not yet known. A family history of PV is rarely present; however, sometimes multiple family members will have the disease. Risk factors may include the following:

- **Mutations**: Almost all people with PV have a JAK2 gene mutation (Janus kinase). Of these, 95 percent have a mutation of JAK-2-V617F (Exon 14) with most of the remainder having mutations in Exon 12.
- **Gender**: Men have a slightly higher risk than women to develop polycythemia vera.
- **Age**: People over age 60 are more at risk.
- **Environment**: Exposure to radiation may increase risk.
- High white blood count increases the risk of blood clotting.

Symptoms

Many patients with PV do not have symptoms when they are diagnosed. Diagnosis often occurs during a routine exam or blood test. However, people may experience persistence of any of the following symptoms.

- Headaches
- Sweating and night sweats
- Ringing in the ears
- Blurred vision or blind spots
- Dizziness or vertigo
- Skin with red or purple hue
- Reddened face
- Unexpected weight loss
- Bleeding or clotting excessively
- Feeling full quickly on eating
- Abnormal discomfort on left side below the ribs
- Itching, especially after a shower or bath
- Burning and redness of the hands or feet
- Fatigue
- Bone pain
- Gout attacks
- Shortness of breath
- Weakness
- Bruising excessively
- Numbness or tingling in feet

Prevention

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

Treatment

PV is a chronic disease, but can be managed well. PV causes differing reactions in each patient. In some cases, symptoms may not be present, in which case treatment may not be required. However, treatment options usually include phlebotomy (removal of blood), and low-dose aspirin. Drug therapy such as hydroxyurea, anagrelide, ruxolitinib, and interferon are also options. Antihistamines and avoidance of hot showers can help with itching. Clinical trials can also be an important treatment option.

Sources: Leukemia and Lymphoma Society, MPN Research Foundation, and National Cancer Institute

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