FEATURE ARTICLE

Polycythemia Vera: A Review

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Polycythemia vera (PV) is a clonal stem cell disorder characterized by increased erythrocyte production. Its etiology is not fully understood, and hemorrhage, thrombosis, and hyperviscosity may occur at any time during the course of this disorder. Treatment depends on the most affected cell type, duration and severity of the condition, and patient age. PV treatment can involve phlebotomy, administration of myelosuppressive agents, and biologic therapy. The inconsonant nature of the disease and complications related to its various treatments present nursing care challenges. Nurses must work with patients with PV to ensure compliance with treatment, teach awareness of disease- and treatment-related complications, and provide needed support.

Key Words: myeloproliferative disorders, polycythemia vera, phlebotomy

Polycythemia constitutes a variety of disorders characterized by excessive red blood cell production. These disorders also are referred to as erythrocytosis because leukocytes and platelets are present in the blood, but in far smaller proportions. Polycythemia is caused by increased proliferation or decreased apoptosis of erythroid progenitors, or by delayed erythroid differentiation with an increased number of progenitor cell divisions. Primary polycythemias result from abnormalities expressed in hematopoietic progenitors. In contrast, circulating factors, such as erythropoietin-stimulating erythropoiesis, cause secondary polycythemia. Acquired and congenital abnormalities can cause primary and secondary polycythemia (Prchal, 2001a, 2001b; Solberg, 2001).

Polycythemia vera (PV) is the only known type of primary acquired polycythemia (Pearson, 2001). This chronic myeloproliferative disorder is characterized by the insidious onset of erythroid proliferation (erythrocytosis) and secondary platelet proliferation. PV can progress from a proliferative stage to a metastatic phase and develop into a malignant phase (Gilbert, 2003). If the excessive production of erythrocytes or platelets is controlled, patients can live for prolonged periods of time with this chronic disorder. However, the clinical course of PV can be complicated by a variety of events, such as bleeding, thrombosis, weakness, weight loss, and neurologic impairment. Life-threatening consequences of disease progression, including myelofibrosis or acute leukemia, also may occur (Hoffman, 2002).

PV is a rare disorder with an incidence of 2.3 per 100,000 and occurs more frequently in men (Tefferi, 2001). Although rarely seen in individuals younger than age 40, the disorder can occur in children and young adults (Hoffman & Boswell, 1995). The disease develops slowly, usually after age 50–60. Risk factors are unknown, but the incidence is highest among people of eastern European Jewish ancestry (Lynch, 2000). Unfortunately, PV prevention is unknown.

Signs and Symptoms

The evolution of PV begins with an asymptomatic phase that can include the clinical findings of splenomegaly, erthrocytosis, and thrombocytosis (Bilgrami & Greenberg, 1995). Patients begin to exhibit symptoms during the erythrocytotic phase secondary to the excessive proliferation of red blood cells and platelets (Hoffman & Boswell, 1995). Symptoms may include pruritus (especially after a hot bath), headache, weakness, dyspnea, visual disturbances, parasthesias, and epigastric complaints (Knoop, 1996). In a study conducted by Merup et al. (2002), the most frequently reported pretreatment symptoms were fatigue, headache, and muscle pain. Figure 1 lists the clinical and pathologic criteria for diagnosing PV.

PV can progress to an inactive phase in which patients may not require phlebotomy or chemotherapy for a period of time. Postpolycythemic myeloid metaplasia (PPMM) follows the inactive phase, which is characterized by splenomegaly, anemia, thrombocytopenia or thrombocytosis, and systemic symptoms such as fever and weight loss (Rosenthal & Murphy, 1995). Treatment of patients in the PPMM phase can include steroids, myelosuppressive agents, or splenectomy. Supportive measures also may require transfusion of packed red blood cells and platelets (Berlin, 2002).

Myelofibrosis associated with the PPMM phase and myelosuppressive therapy may cause patients to develop acute myeloid leukemia or acute promyelocytic leukemia (Kajiguchi, Simokawa, Saito, & Takeyama, 2000). However, the risk of leukemic transformation is considered low (i.e., less than 5%) (Fenaux et al., 1990).

Treatment

Treatment of PV must be individualized according to age, gender, clinical status, disease manifestations, and hematologic findings (Spivak, 2002). Phlebotomy is integral to

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