■ Article

Optimizing the Management of Patients With Myelofibrosis

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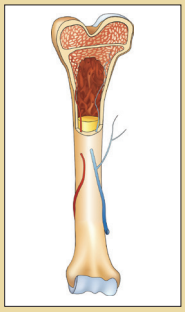

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Myelofibrosis (MF) is a rare myeloproliferative neoplasm of the bone marrow associated with shortened survival. The disease is characterized by splenomegaly, cytopenias, and multiple disease-related symptoms that reduce quality of life. The clinical management of MF can be challenging because of its heterogeneous presentation and disease course. Therefore, knowledge of the underlying pathology and clinical manifestations of MF is needed. Ruxolitinib, a Janus kinase (JAK) 1 and 2 inhibitor, is the first therapy to be approved by the U.S. Food and Drug Administration for intermediate- or high-risk MF. Ruxolitinib therapy offers advantages over the previous palliative treatments and has shown durable reductions in splenomegaly and disease symptoms as well as improvements in quality of life. Two-year follow-up of the phase III trials also has shown that ruxolitinib treatment was associated with a survival advantage relative to control groups. Dose-dependent thrombocytopenia and anemia are expected but manageable adverse effects caused by the targeted JAK inhibition of ruxolitinib. This review provides an overview of MF and assessment of the primary clinical disease manifestations, with a focus on ruxolitinib from the oncology nurse perspective.

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Myelofibrosis (MF) is a rare myeloproliferative neoplasm that is predominantly diagnosed in older individuals (median age 65), although it also can affect younger people (Cervantes et al., 2009). No known risk factors exist for MF (Girodon et al., 2009), which occurs with an annual incidence of anywhere from four to six people per 100,000 in the United States (Mehta, Wang, Iqbal, & Mesa, 2014). It can develop as primary MF (PMF) or secondarily from polycythemia vera (PV) or essential thrombocythemia (ET) (Mesa et al., 2011) and is characterized by fibrosis of the bone marrow, ineffective hematopoiesis, increased risk of transformation to acute myelogenous leukemia, and shortened survival (Cervantes et al., 2009; Gangat et al., 2011; Passamonti et al., 2010). Because of the heterogeneous nature of MF, diagnosis is complex and is established based on criteria from the World Health Organization (Vardiman et al., 2009). The clinical course and prognosis can vary widely, but most patients will develop typical manifestations such as progressive splenomegaly, cytopenias, and debilitating symptoms, which substantially diminish quality of life (QOL) (Gregory, Mesa, Hoffman, & Shammo, 2011).

Allogeneic stem cell transplantation is the sole treatment for a potential cure, but often is associated with significant morbidity and mortality, and few patients with MF are eligible because of advanced age and comorbidities (Mesa, 2013). Palliative cytotoxic therapies were, until recently, the only available options for the alleviation of splenomegaly (Rambaldi, Barbui, & Barosi, 2008). However, none of those therapies had long-lasting efficacy, and only provided limited, transient benefit (Vannucchi, 2011).

The Janus kinase (JAK)/signal transducer and activator of transcription (STAT) pathway plays a pivotal role in hematopoiesis. Researchers have discovered that dysregulation of the JAK/STAT pathway, resulting from somatic mutations in JAK2