Review article

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CONTEMPORARY APPROACH IN THE DIAGNOSIS AND MANAGEMENT OF PRIMARY MYELOFIBROSIS

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Primary myelofibrosis (PMF) is an infrequent chronic myeloproliferative neoplasm. PMF is a result of clonal expansion of myeloid cells and is distinguished by the variable presence of mutations, morphologically by increased proliferation of megakaryocytes, progressive bone marrow fibrosis, hepatosplenomegaly, anemia, leukoerythroblastosis, with constitutional symptoms and shortened survival. World Health Organization defined the current diagnostic criteria for PMF in 2016, which involve a combined assessment of clinical, histological, mutational and laboratory features of diseases. Recently, a several new PMF prognostic scoring systems have started being used in the clinical practice, which are based solely on genetic markers or include clinical variables in addition to mutations and karyotype. In the treatment of myelofibrosis, risk adapted therapy has been applied, which implies the selection of the type of therapy according to the risk category obtained by calculating the valid prognostic scores. Allogenic stem cell transplant remained the only potentially curative therapy for PMF treatment but is suitable only for a small number of high risk patients who have a matching donor. In the last decade, the development and approval of ruxolitinib for the treatment of PMF has been of the greatest importance in the treatment of this disease, although it is a palliative therapy. Ruxolitinib is a potent JAK1/JAK2 inhibitor that leads to decreases in splenomegaly and symptoms and has prolonged overall survival in patients with this disease.

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