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Myelofibrosis management updates

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yeloproliferative neoplasms (MPN) are myeloid hematologic neoplasms that are characterized by expansion of terminal myeloid cells (white blood cells, platelet, red blood cells) and their precursors in the bone marrow (BM) and peripheral blood. MPN include primary myelofibrosis, essential thrombocythemia, polycythemia vera, chronic myeloid leukemia and a few other less common neoplasm. The term myelofibrosis (MF) combines primary myelofibrosis and myelofibrotic transformation of other myeloproliferative neoplasms, namely essential thrombocythemia and polycythemia vera, due to the similar disease characteristics and clinical outcomes. MF is manifested by increased interstitial collagen deposits in the BM, extramedullary hematopoiesis which can result in massive enlargement of the spleen, abnormal blood counts, and debilitating symptoms (e.g., fatigue, weakness, abdominal pain, cachexia, weight loss, pruritus, night sweats, and bone pain) which are thought to be caused by the combined effects of massive splenomegaly and elevated levels of pro-inflammatory cytokines. Survival of patients with MF is compromised and ranges from 2 to 11 years depending on defined prognostic factors. The majority of patients suffer a heavy burden of symptoms due to their disease and/or their low blood counts. Traditional therapeutic options are only palliative and include splenectomy, chemotherapy, immunomodulatory drugs and targeted therapies such as Jak-2 inhibitors. Only allogeneic hematopoietic stem cell transplantation can favorably impact the natural disease course of MF but, unfortunately, few patients are eligible for this treatment. The scarcity of therapeutic options is partially attributable to our limited understanding of the key processes driving this malignancy. There have been several advances in the understanding of the disease biology and the available therapeutic interventions.

Biography

Abdulraheem Yacoub is a young physician-scientist and an Assistant Professor of Medicine in the division of Hematology and Oncology at the University of Kansas. He is nationally recognized for his efforts in improving medical care and patient education in the MPN field. He is a recipient of the MPN Heroes Award and a member of the Myeloproliferative Diseases Research Consortium.

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