48. PRIMARY MYELOFIBROSIS: CLINICAL PICTURE, DIAGNOSIS AND TREATMENT

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Introduction. Primary myelofibrosis is a rare chronic myeloproliferative disorder with an annual incidence of 0.5 cases per 100.000 individuals and is characterized by bone marrow fibrosis, extramedullary haematopoiesis, splenomegaly and leukoerythroblastic blood picture. Primary myelofibrosis typically occurs in people older than 50 years, the mean age at diagnosis being 65 years.

Aim of study. The study of clinical manifestations, diagnostic methods and treatment principles in patients with primary myelofibrosis.

Methods and materials. A study was conducted based on a group of 52 patients who were registered at the Hematology and Diagnostic Consultative Center of the PMSI Institute of Oncology of Moldova. The group of individuals enrolled in the study consisted of 30 (58%) women and 22 (42%) men. The age of patients ranged from 38 to 83 years and the median was 64 years. The diagnosis of primary myelofibrosis in all cases was morphologically confirmed.

Results. The disease has developed mainly in people aged between 60 and 69 years (46.15%). The main complaints were discomfort and/or pain in the left hypochondriac region in 30 (58%) cases and with asthenic syndrome in 22 (42%) cases. During the objective examination, splenomegaly was determined in 42 (80%) patients and hepatomegaly in 26(50%). Autoimmune haemolytic anemia was determined in 4 (8%) patients and metaplastic anemia in 8 (15%). In 48 (92%) patients, hyperthrombocytosis was found with values from 450-2450 x 109 /L, and in 4 (8%) autoimmune thrombocytopenia. The treatment was performed with Hydroxycarbamide in the regimen and standard doses with achievement of partial remission. In cases with immune complications, corticosteroid has been administered with efficacy in 7(63.6%) patients. Four patients were followed up until death. The overall survival rate over 1 year, 3 and 5 years was 96.1% accordingly.

Conclusion. Primary myelofibrosis is more common in people aged 60-69 years, mainly in women (58%). The primary objective sign was splenomegaly in 42 (80%) cases. Partial remission was achieved in all patients on a treatment background. The overall survival rate over 1 year, 3 and 5 years was 96.1%.