ANALYSIS OF CLINICAL AND HEMATOLOGICAL CHARACTERISTICS OF PRIMARY MYELOFIBROSIS IN THE YOUNG PATIENTS FROM THE REPUBLIC OF MOLDOVA: A SINGLE-INSTITUTION EXPERIENCE

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Background

For a long time it has been believed that PM is a disease of the elderly. In the last 10 years, many articles have been published on the detection of PM in young people and even children. Despite an enormous amount of research, etiology is still unknown. The absence of long-term observations and conclusions about the course of the disease puts specialists in a delicate position when they meet young patients.

Objective

Determination of clinical hallmarks of primary myelofibrosis (PM) in patients under 40 years of age.

Materials & Methods

Design: We performed this retrospective, clinico-analytical study and evaluated the data of 46 patients with PM, registered at the Department of Hematology of the Institute of Oncology in the Republic of Moldova (RM). In our country, all patients with the diagnosis of hematological malignancies are followed up by doctors from the Department of Hematology of the Institute of Oncology. These patients are managed at the Consultative Diagnostic Center, outpatient, and inpatient wards . At the moment there are 1110 patients with MPN and 450 patients with PM, including 46 young persons. These are the newest data that enroll all patients and newly -diagnosed young patients with PM. For convenience, we divided them into three groups according to age: 18-20, 21-30, and 31-40 years.

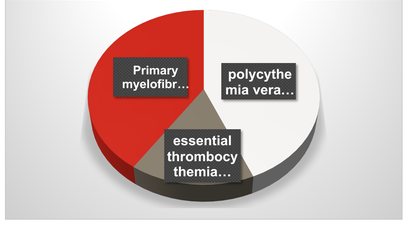


Fig.1 Patients with MPN

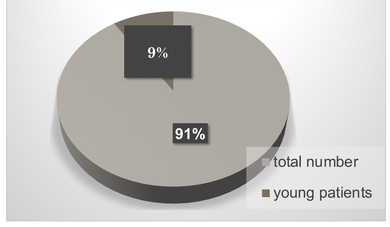


Fig.2 Patients with PM

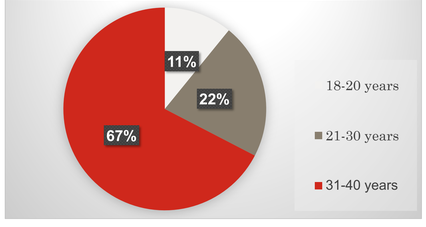


Fig.3 Age distributions

Results

There were 11 male and 35 female patients. The major features of the first group were massive splenomegaly dominating in all 10.9% of patients and mostly normal CBC values. In the third group, we observed elevated hemoglobin level in 13.0% and frequent thrombocytosis in 52.1% of cases. Massive splenomegaly was detected in 19.6% of cases and normal spleen size - in 6.5% of cases. Two patients underwent splenectomy. Histological investigations revealed fibrosis MF1-2 and hypercellular bone marrow in almost all cases. Thrombocytosis and splenomegaly of various sizes were found in the entire group of patients aged 21–-30 years.

	1st group	2nd group	3-rd group
Low hb	2,1%	6,5%	17,4%
Hemolitic anemia	4,3%	2,1%	4,3%
Normal range of Hb	4,3%	13,0%	32,6%
Elevated Hb	-	-	13,0%

Tab.1 Hemoglobin level

	1st group	2nd group	3rd group
Normal size	4,3%	17,4%	30,4%
Moderate hepatomegaly	-	2,1%	23,9%
Severe hepatomegaly	6,5%	2,1%	13,0%

Tab. 3 Liver size

Conclusions

This analysis did not reveal a correlation between proliferative syndrome and the degree of leukocytosis and thrombocytosis. The disease may start with initial changes in the CBC, which should alert any specialist. All these patients remain under observation in our department.

	1st group	2nd group	3rd group
Low level of PLT	-	2,1%	4,3%
Normal range 150-400 x10^9/l	8,7%	6,5%	10,9%
PLT > 450 x10^9/l	2,1%	13,0%	52,1%

Tab.2 Platelet level

	1st group	2nd group	3rd group
Normal size	-	4,3%	6,5%
Mild splenomegaly	-	2,1%	21,7%
Moderate splenomegaly	-	6,5%	15,2%
Severe splenomegaly	10,9%	6,5%	19,6%
Splenectomy	-	2,1%	4,3%

Tab. 4 Spleen size