Conclusion: We report a rare case of ET with concomitant B-LPD. The patient is stable on Ruxolitinib and is on wait and watch approach for B-LPD.

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PP 13

Acute phase reactants in chronic inflammation leading to secondary myelofibrosis in polycythemia vera and essential thrombocytosis

E. Aladag[1], I. Haznedaroglu[1], N. Sayinalp[1], H. Demiroglu[1], H. Goker[1,*], S. Aksu[1], O. Ozcebe[1], A. Ayhan[2], Y. Buyukasik[1]

[1] *Hacettepe University Department of Hematology, Ankara, Turkey*
[2] *Hacettepe University Department of Pathology, Ankara, Turkey*

Objective: Polycythemia vera and essential thrombocytosis are chronic and progressive myeloproliferative neoplasms characterized by a clonal increase in hematopoietic stem cells in the bone marrow. Myelofibrosis in the bone marrow has been shown to be secondary to an inflammatory process.

Methodology: To investigate the association between the secondary myelofibrosis and acute phase reactants in patients with polycythemia vera and essential thrombocytosis. Fourty-six PV and 28 ET patients without myelofibrosis above Grade 1 were included in the present study. Bone marrow evaluations were performed retrospectively. C-reactive protein, ferritin, and albumin levels were measured.

Results: C-reactive protein (0.55 ng/L vs. 4.2 ng/L, $p < 0.001$) and ferritin (18.5 ng/mL vs. 118 ng/mL, $p = 0.001$) levels in patients with secondary myelofibrosis were found to be increased compared to baseline levels. Mean albumin levels in patients with secondary myelofibrosis, and CRP, ferritin, and albumin levels in patients without secondary myelofibrosis were similar at the diagnosis and at last visit. There were also similar the baseline levels of CRP, ferritin, and albumin between the patients with and without secondary myelofibrosis.

Conclusion: The increase in CRP and ferritin, which are indicators of chronic inflammation, may be used to show the inflammation and relevant secondary fibrosis in the bone marrow. Due to the similar CRP, ferritin, and albumin levels at the diagnosis, the prediction for the development of the secondary myelofibrosis is not possible in the present study.

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PP 14

Polycythemia vera: updates in diagnosis and treatment outcomes

L. Musteata[1,*], S. Pinzari[2], V. Musteata[1], N. Sghibneva-Bobeico[2], A. Dorogan[2]

[1] *State University of Medicine and Pharmacy, Chișinău, Republic of Moldova*
[2] *Institute of Oncology, Iași, Romania*

Objective: The objective of the study was to analyze the contemporary clinical and laboratory features of polycythemia vera (PV), as well as to evaluate the short- and long-term results of different treatment options.

Methodology: The clinico-hematological evolution features, complications, short- and long-term results of cytoreductive treatment were evaluated in a group of 114 PV patients, aged at 28–78 years old, who were followed up at the Institute of Oncology of Moldova between 1987–2019. The diagnosis was proved by the bone marrow biopsy and quantitative detection of JAK2 V617F mutation in pending cases. Physical and histopathologic examinations were associated with the repeated complete blood counts and abdominal ultrasound scan. The treatment included phlebotomies and cytoreductive chemotherapy with busulfan (56 patients) and hydroxycarbamide (58 patients) in standard doses. The life-table method was used for Kaplan–Meier Survival Analysis in order to evaluate the long-term results of treatment.

Results: The disease was commonly diagnosed in males – 66 (57.9%) patients. The females prevailed in the age groups of 40–49 years (31.3% versus 24.6% in males) and 60–69 years (25% versus 19.8% in males). The disease span from the onset of the initial clinical manifestations until the diagnosis lasted 4–9 months (median – 5.8 months) in the majority of patients (86.8%), that led to the development of thromboembolic complications in 28.1% of cases. The diagnosis was proved in stage IIA disease in 105 (92.1%) patients, IIB in 9 (7.9%) patients. The skin hiperemia was registered in 112 (98.3%) cases, scleral congestion – in 109 (95.6%), splenomegaly – in 77 (67.5%), erythromelalgia – in 71 (62.2%), aquagenic skin itching – in 68 (59.6%), hepatomegaly – in 61 (53.5%), vascular thrombosis – in 32 (28.1%). The complete blood count revealed the increase of hemoglobin (18.0–23.5 g/dL) and red cells (5.5–6.7 × 1,000,000 [MICRO]/L). The platelets range was 180–1690 × 1000 [MICRO]/L, leukocytes range – 5.1–21.3 × 1000 [MICRO]/L. Leukocytosis occurred in 69 (60.5%) patients, thrombcytosis – in 61 (53.5%). The bone marrow biopsy detected a hyperplasia due to the proliferation of erythroid, granulocyte and megakaryocyte cell lines. The study of short-term results asserted the complete remissions in all cases under chemotherapy combined with phlebotomies. The overall one-, 5-, 10- and 15 year was 100%, 98.6%, 85.9% and 67.1%, respectively. 73 (64.04%) patients remain in stage II disease after the treatment during 5–26 years of follow-up. The survival median was not reached.

Conclusion: The reluctant evolution, progressive growth of hemoglobin and red cell count, gradual increase of blood hyperviscosity and the lack of hemato-oncological vigilance of primary care physicians may lead to the development of

thrombotic and vascular complications in some PV cases. Chemotherapy improves significantly the patient's quality of life, reduces the rate of thromboembolic events and extends the life-span, comparable with that of total population of Moldova.

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PP 15

Disease and clinical characteristics of patients with chronic myeloproliferative neoplasms: 11-year single center experience

P. Akyol, A. Yıldız, H. Afacan Öztürk, S. Maral, M. Reis Aras*, F. Yılmaz, B. Saglam, M. Tıglıoglu, M. Albayrak

Diskapi Yildirim Beyazit Training and Research Hospital, Ankara, Turkey

Objective: BCR/ABL-negative myeloproliferative neoplasms are characterized by over-production myeloid lineages in the bone marrow. Polycythemia vera, essential thrombocythemia and primary myelofibrosis are the most common myeloproliferative neoplasms. Diagnosis is made according to the WHO diagnostic criteria from clinical data, hematological and biochemical analysis and BM histology. The aim of this study was to analyse patient demographic characteristics, clinical features, laboratory findings, mutational status together with complications, clinical course and survival.

Methodology: This study was conducted on patients diagnosed with myeloproliferative neoplasms between 2008 and 2019. Hemogram and biochemical parameters, demographic information, mutation analysis, management, complications and follow-up periods were recorded for all patients. Survival rates were calculated and the effect of the parameters on overall survival was analyzed.

Results: Evaluation was made of 247 patients, comprising 105 polycythemia vera, 126 essential thrombocythemia and 16 primary myelofibrosis patients. The overall frequency of driver mutations was 96.1% for PV, 71.4% for ET and 75% for PMF. Hydroxyurea was the most commonly used first-line treatment agent and the most common indication for switching to second-line treatment in all disease subgroups was the development of side-effects. During follow-up, 11 polycythemia vera, 14 essential thrombocythemia and 2 primary myelofibrosis patients developed thromboembolic complications. Median overall survival could not be reached in polycythemia vera and essential thrombocythemia patient and determined as 70.3 months in primary myelofibrosis patients. Age, LDH, ferritin and platelet/lymphocyte ratio at the time of diagnosis and thromboembolic complications were determined to have a statistically significant effect on survival in all patients.

Conclusion: Lower survival rates were seen in the primary myelofibrosis patients although thromboembolic complications were observed at similar rates in all 3 disease subgroups. In addition to known risk factors such as age and thromboembolic complications, parameters such as LDH, ferritin and PLR, which may be considered to indicate disease activity and inflammation, can also be used as prognostic markers.

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PP 16

The frequency of calreticulin and mpl gene mutations in bcr-abl and jak2 unmutated chronic myeloproliferative neoplasms and its effect on the outcome

T. Tiryaki[1], A. DağLar Aday[2], M. Güzel Mastanzade[1], M. ÖZbalak[1], D. Özlük[1], F.Y. Onal Hindilerden[1], M. Yenerel[1], A. Yavuz[1], S. Kalayoğlu Beşışık[1,*], M. Nalçacı[1]

[1] *İstanbul University, İstanbul Medical Faculty, Department of Internal Medicine, Division of Hematology, İstanbul, Turkey*
[2] *İstanbul University, İstanbul Medical Faculty, Department of Medical Genetics, İstanbul, Turkey*

Objective: The World Health Organization (WHO) embedded calreticulin receptor (CalR) and myeloproliferative leukemia virus (MPL) gene mutation in diagnostic criteria for primary myelofibrosis (PMF) and essential thrombocythemia (ET), since 2016). We aimed to identify the frequency of CalR and MPL gene mutations and their effects on clinical outcomes in bcr-Abl and Jak2 unmutated chronic myeloproliferative neoplasms (MPNs).

Methodology: We screened bcr-abl negative and Jak2 unmutated MPNs diagnosed and treated between March 2004 and January 2013 at İstanbul Medical Faculty. We revised the MPN diagnosis according to the latest WHO classification. The association of CalR and MPL mutation with thrombotic complications, leukemic transformation, and survival was was defined.

Results: A total of 46 ET ($n = 34$) and PMF ($n = 12$) patients enrolled in the study. The demographic characteristics were similar between the two disease groups. Patients' mean age was 53.5 years (range 23–93 years) and gender distribution as 18 male to 28 female. A total of 18 patients (39.1%) had CalR mutation, and 4 (8.69%) patients MPL mutation. None of the ET patients had MPL mutations. CalR positive PMF patients' mean age was lower compared to patients without the mutation (p: 0.028). During the follow-up period, 8.3% of PMF and 5.9% of ET patients experienced leukemic transformation. None of the leukemic transformed patients had gene mutations. Among thrombosis complications, six patients developed thrombosis. All of them were ET patients, and 3 of them had CalR mutation two as CalR type 1 and one as CalR type 2. The mortality ratio was higher in patients in PMF, regardless of mutational status (p: 0.006).

Conclusion: Our study cohort is small to make a definite conclusion. Apart from the diagnostic guide, CALR mutations seem to have a prognostic effect is different in PMF and ET. This prognostic significance of CALR could be different among the MPN categories.

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