HEMATOLOGY, TRANSFUSION AND CELL THERAPY

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Oral Presentations

Adult Hematology Abstract Categories, Chronic Myeloproliferative Diseases

OP 01

RETROSPECTIVE ANALYSIS OF PRIMARY MYELOFIBROSIS PATIENTS IN AZERBAIJAN

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Objective: Primary myelofibrosis (PMF) is a rare Ph chromosome-negative chronic myeloproliferative neoplasm characterized by the proliferation of atypical clonal megakaryocytes and fibrosis of the bone marrow. The activation of the JAK-STAT pathway plays a central role in the pathogenesis of the disease. The majority of patients with primary myelofibrosis have one of three main genetic mutations, including JAK2 V617F, CALR exon 9, or MPL W515. The clinical features of the disease are highly heterogeneous. Common symptoms and signs include fatigue, constitutional symptoms, itching, abdominal discomfort, bone pain, anemia, leukocytosis, thrombocytopenia, and splenomegaly. A number of clinical studies on the demographic and clinical features of myelofibrosis have been carried out in different countries. Detailed demographic and clinical characteristics of patients with BMF have not been thoroughly studied in Azerbaijan. The aim of our study was to characterize the demographic, clinical, and laboratory parameters of patients with primary myelofibrosis in Azerbaijan. All patients were registered at the Azerbaijan National Center for Hematology and Transfusion. **Methodology:** A retrospective analysis was conducted on the demographic, clinical, and laboratory data of 131 patients diagnosed with PMF between January 1, 2011, and December 1, 2023. The diagnosis of all patients was revised according to the WHO 2016 criteria for PMF. The fibrosis of the bone marrow was assessed histologically according to the Thiele grading system. Ultrasound examination was used to assess splenomegaly, with a craniocaudal size of >14 cm being considered as splenomegaly. All data were collected from clinical records. This was a retrospective, observational, single-center study. **Results:** A total of one hundred thirty-one (131) patients with primary myelofibrosis were analyzed. Of these, 65 (49.6%) were male. The median age of the patients was 57.5 years (range 19-80), with 9 (6.87%) patients being under 40 years of age. The median hemoglobin level was 10.7 g/dl (range 2.1-19.4), median white blood cell count was 12.86 × 10^12/l (range 0.45-121), median platelet count was 322 × 10^12/l (range 24-1940), and median LDH was 530 U/l (range 181-1586). Splenomegaly was detected in 96 patients, with an average spleen size (19.5 cm)reported. Fifty-one patients had Hgb < 10 g/dl. At the time of diagnosis, the pre-fibrotic stage was identified in the bone marrow examination of sixteen patients (17.8%). Splenomegaly was detected in 96 (91.4%) patients. Of the 66 patients who underwent genetic testing, 44 had a positive Jak2V617F mutation, 2 had a positive CALR mutation, and 1 had a positive MPL mutation. **Conclusion:** Thus, this study has investigated the demographic, clinical, and laboratory characteristics of patients with primary myelofibrosis in Azerbaijan.

https://doi.org/10.1016/j.htct.2024.04.003

OP 02

BIOCHEMICAL PROPERTIES OF RED BLOOD CELLS IN POLYCYTHEMIA VERA

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Objective: Polycythemia vera (PV) is a chronic myeloproliferative neoplasm characterized by an increase in red blood cell mass. Thrombotic complications are the main cause of morbidity and mortality in PV. Elevated hematocrit and increased blood viscosity are crucial risk factors for thrombus formation. The aim of our analysis is to evaluate the biochemical alterations in red blood cells (RBCs) and the hemoglobin structure in patients with PV that may be associated with thrombotic complications. **Methodology:** Blood samples were taken from 20 PV patients and 16 healthy individuals. The isolated RBCs were examined using Raman spectroscopy. **Results:** We found a larger contribution of ferrous heme iron, which is a molecular state typical for deoxyhemoglobin in PV samples compared to the control samples. Furthermore, a significant increase in the Fe II/Fe III ratio in PV samples was correlated with a higher hematocrit (Hct) to hemoglobin (Hgb) ratio. A positive trend between a higher Fe II/ Fe III ratio and a higher RDW-SD and RDW-CV was observed in PV samples. In RBCs collected from PV patients we observed a less stable hemoglobin structure. **Conclusion:** Higher values of RDW-SD and RDW-CV may reflect a higher Fe II/ Fe III and be a simple indicator of biochemical alterations in RBCs. A higher Hct/ Hgb ratio could indicate higher clonal myeloproliferative potential and be associated with shorter time to thrombosis in patients with PV. Our future analysis will focus on correlating the above observations with the prothrombotic activity to demonstrate a possible link between the biochemical alterations of RBCs and the thrombotic complications in PV.

https://doi.org/10.1016/j.htct.2024.04.004

OP 03

SECONDARY SOLID CANCER FREQUENCY AND RISK FACTORS IN PHILADELPHIA- NEGATIVE CHRONIC MYELOPROLIFERATIVE NEOPLASMS

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Objective: Philadelphia chromosome-negative myeloproliferative neoplasms (Ph- MPNs) are characterized by clonal myeloproliferation and somatic mutations. Major complications of Ph-MPNs are thrombosis, bleeding, transformation to myelofibrosis and leukemia. One important concern in the course of Ph-MPNs is risk of development of secondary solid cancers (SSC). In a large cohort of Turkish Ph-MPN patients, we aimed to determine the types and frequencies of SSC, to identify risk factors for SSC including role of cytoreductive therapies and to study impact of SSC on survival in Ph-MPNs. **Methodology:** 1013 patients diagnosed with Ph-MPN from 1995 and 2022 under follow up at adult hematology sections of Istanbul Bakırköy Dr Sadi Konuk Hospital and Istanbul University Medical Faculty were included in this retrospective study. **Results:** Of the 1013 Ph- MPN patients enrolled in our study, 65, 46 and 37 patients were diagnosed with essential thrombocythemia (ET), polycythemia vera (PV) and primary myelofibrosis (PMF), respectively. Patient clinical and laboratory characteristics are summarized in Table 1. Sixty-seven patients (6.6%) developed SSC, predominantly carcinoma (64.2%), non-melanoma skin cancer (23.9%), sarcoma (4.5%), and melanoma (3%). Median time to SSC diagnosis was 80.03 ± 60.5 months with no significant difference among Ph-MPN subtypes. Compared to patients with no diagnosis of SSC, patients with SSC were older at time of Ph- MPN diagnosis (63 vs. 54 years; $p<0.001$) and included a higher proportion of males ($p=0.025$). Ph- MPN patients with SSC and without SSC showed no significant difference for complete blood count parameters, spleen size, Ph-MPN diagnosis groups, driver mutation frequencies and follow-up time. Arterial thrombosis frequency was higher in patients with SSC (37.3% vs. 25.3%; $p=0.030$). SSC rates were 5.7% in patients not exposed to cytoreductive treatment and 5.3%, 4% and 2.1% with exposure to ruxolitinib, anagrelide, and interferon (IFN), respectively. A trend toward lower SSC rates was noted with IFN therapy (3% vs. 97%; $p=0.066$). SSC incidence was significantly higher in patients exposed to hydroxyurea (HU) as first-line monotherapy compared to other treatment groups (7.8% vs. 4.6%; $p=0.046$). Median OS in patients with SSC and patients with no diagnosis of SSC group were 273 months and 195 months, respectively. PV patients, who developed SSC, had significantly worse median OS compared to PV patients without SSC (Figure-1). **Conclusion:** The strengths of our study are that it enrolls a larger patient population, includes PV, ET and PMF subgroups, separately examines development of SSC after MPN, has a long follow-up period and has multicenter design. In MPN patients, malignancy screening gains more importance for those aged ≥65 and males. Our study evaluated with data from previous studies suggest that increased risk of developing SSC in MPN patients may be associated with cytoreductive therapy. Further studies with more pateints are needed