PB2052 CLINICOPATHOLOGIC VARIABLES AFFECTING DISEASE OUTCOME IN PRIMARY MYELOFIBROSIS. A SINGLE CENTER EXPERIENCE.

**Topic:** 16. Myeloproliferative neoplasms - Clinical

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**Background:**

Primary myelofibrosis (PMF) is a clonal myeloproliferative neoplasm. The prognosis of PMF is variable and clearly worsens with evolution of specific factors which accelerate disease progression.

This is a single center retrospective study carried out in National Center for Cancer Care and Research, Hamad Medical Corporation over the period 2008-2021.

In this study, we included all patients given a diagnosis of World Health Organization-defined PMF (prefibrotic and overt fibrotic). We collected demographic data, pathologic bone marrow, driver mutations, clinical and hematologic characteristics of the study patients and correlated with disease progression. Dynamic International Prognostic Scoring System – Plus (DIPSS Plus) was used as well but was further grouped into two (low and high risk groups) instead of four groups.

**Aims:** Here we aim to study different clinicopathologic and genetic parameters that affect the disease course in PMF patients along with their treatment history and disease outcome, in order to find out the most significant factor/s.

**Methods:**

Our Cohort included 61 patients diagnosed with PMF. Variables studied were diagnosis (prefibrotic and overt fibrotic), age, anemia, DIPSS scoring as modified into 2 groups (low/intermediate-1 vs intermediate-2/high), cytogenetic (favorable and unfavorable karyotype), JAK2V617F mutation, need to treatment, complications mainly thromboembolic complications.

Time to progression, was considered as the time to progression, was defined as the time to acquisition of ≥1 of the following: Hemoglobin < 10 g/dl, Leukocyte count >25 x10⁹/L, Platelet count < 100 x10⁹/L, Circulating blast cells ≥1%, transfusion dependency. Overall survival (OS) and progression free survival (PFS) were assessed using time-dependent, competitive risk analysis.

Kaplan-Meier method was used for survival analysis. Results were considered statistically significant when P values were less than 0.05

**Results:** The most significant factors adversely affected overall survival are refractory anemia (p. value 0.006), fibrotic stage of the disease (p. 0.030), higher risk group according to DIPSS scoring system (p.0.03), unfavorable karyotype (p.0.02) and eventually AML transformation (P.0.00).

For factors that had a significant impact on disease progression, again included refractory anemia (p.0.00), patients with higher DIPSS score (p.0.00), patient with PMF secondary to other myeloproliferative neoplasms had worse disease outcome compared to primary PMF (p.0.026), patients who needed treatment had shown disease progression compared to those who did not need treatment (p. value 0.001). Disease progression was also significantly associated with patients who received Ruxolitinib compared to those who did not (p.value 0.000), and patients who needed...
Regarding DIPPS-Plus, 39 (63.9%) patients were encountered in low to Intermediate-1, while 22 (36.1%) in Intermediate-2 to high risk groups. For disease stage, 32 (52.4%) were prefibrotic, but 29 (47.5%) were fibrotic. 44 (72.1%) patients needed treatment, while 17 (27.86%) did not.

On the other hand, disease progression and the overall survival were not affected by age, molecular genetics findings, hematologic counts, driver mutations or thromboembolic complications.

**Summary/Conclusion:**

Among all factors studied we found that refractory anemia, unfavourable karyotype and higher risk group according to DIPSS scoring are the most important factors affecting the disease progression and overall survival with statistically significant difference.