The MPN-10 Score and Driver Mutations in Thai Myeloproliferative Neoplasm Patients

Naritsara Cherdchoo¹
Chantana Polprasert¹
Sunisa Kongkiatkamol¹
Ponlapa Rojnuckarin¹

¹Division of Hematology, Department of Medicine, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand

Background: Recent advances in molecular diagnostics found that the majority of classic myeloproliferative neoplasms (MPNs) harbor characteristic driver mutations in JAK2, CALR or MPL genes. The JAK2 protein transmits signals of various cytokine receptors, while CALR and MPL are involved only in the thrombopoietin receptor signaling. This may contribute to different symptoms of patients. The MPN Symptom Assessment Form total symptom score (MPN-SAF TSS) or MPN-10 score is a useful tool for the assessment of symptom burden in these patients. However, this tool has never been validated in Thai patients.

Objective: To determine the MPN-10 scores and correlation with clinical characteristics, genetic mutations, and outcomes of MPN patients.

Methods: A single center, cross-sectional study was conducted. MPN-10 scores and outcomes of MPN patients diagnosed according to WHO criteria at King Chulalongkorn Memorial Hospital, Thailand between 2014 and 2017 were reviewed.

Results: A total of 146 patients were enrolled. Forty-seven patients were diagnosed as polycythemia vera (PV), while 72 essential thrombocythemia (ET), and 27 primary myelofibrosis (PMF). One hundred and thirty-two patients (90.5%) had driver mutations. There were 107 JAK2V617F mutations: 46 PV (42.9%), 40 ET (37.3%), and 20 PMF (18.6%). Twenty-one patients harbored CALR mutation: 18 ET (85.7%) and 3 PMF (14.2%). One MPL mutated-ET and 14 triple negative (TN)-MPNs (10%): 11 ET (79%) and 3 PMF (21%), were also found. The mean MPN-10 score of PMF was 24.4 (8-47), which was significantly higher than those of PV, 11.2 (0-35) (p=0.001), and ET, 10.7 (0-36) (p=0.000). The mean MPN-10 scores of triple negative-ET vs JAK2V617F ET vs CALR-mutated ET were 8.3(0-19) vs 10.17 (0-25) (p=0.994) vs 12.5(0-36) (p=0.417). The MPN-10 is not associated with survival (p=0.643).

Conclusion: PMF has a higher symptom burden than PV and ET. Interestingly, mean MPN-10 scores in PV and ET of our patients are lower than those of Western studies. MPN-10 scores are not associated with driver mutations and could not predict survival.

Keywords: Myeloproliferative neoplasm, MPN-10 score, Polycythemia vera, Essential thrombocythemia, Primary myelofibrosis, Survival