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The epidemiology and presenting clinical characteristics of myeloproliferative neoplasms in Malaysia

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Background: The evolution of molecular studies in Myeloproliferative Neoplasms (MPN) has enlightened us the understanding of this complex disease consisting of Polycythemia Vera (PV), Essential Thrombocythemia (ET) and Primary Myelofibrosis (PMF). The epidemiology is well described in the western world but not in Asian countries like Malaysia.

Method: This retrospective national registry of MPN was conducted from year 2009 to 2015 in Malaysia.

Result: A total of 1010 patients were registered over a period of 5 years. The mean age was 54 years with male predominance. The ethnic distribution revealed that Chinese had a relatively high weighted incidence proportion (43.2%), followed by Indian (23.8%), Malay (15.8%) and other ethnic groups (17.2%). The types of MPN reported were 40.4% of ET (n=408), 38.1% of PV (n=385), 9.2% of PMF (n=93), 3.1% of Hypereosinophilic Syndrome (HES) (n=31) and 7.9% of unclassifiable MPN (MPN-U) (n=80). Splenomegaly was only palpable clinically in 32.2% of patients. The positive JAK2 V617F mutation was present in 644 patients with 46.6% in PV, 36.0% in ET, 9.0% in PMF and 7.4% in MPN-U and had significantly lower hemoglobin (p<0.001), hematocrit (p<0.001) and White Blood Cells (WBC; p<0.001) than those with negative mutation. Significant differences in platelet and WBC count were detected in ethnic groups and MPN sub-types. There were more arterial thrombosis events seen in those with JAK2 V617F mutation as compared to venous thrombosis events (23.1% vs. 4.4%). The bleeding rate was only 6.6%. Among the risk factors, previous thrombosis, old age (≥60 years) and hypertension were significantly correlated to positive JAK2 V617F mutation. The arterial thrombosis event is associated with higher presenting HB, HCT and PLT while the bleeding event is associated with lower presenting HB, HCT but higher PLT. The presence of JAK2 V617F mutation is associated with higher risk of arterial thrombosis.

Conclusion: Chinese ethnicity is associated with higher rates of MPN. The history of thrombosis, age \geq 60 years and hypertension are risk factors that can be correlated to JAK2 V617F mutation. This study is instrumental for policy makers to ensure preventive strategies that can be implemented in future.

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