Title: The survival outcomes of patients with Myeloproliferative Neoplasms in Malaysia

Abstract

Background: The prognostication of myeloproliferative neoplasm (MPN) has always been challenging even in the advent of Janus kinase 2 (JAK2 V617F) molecular studies. The survival pattern of MPN in developing country such as Malaysia is still undetermined.

Materials and Methods: This was a retrospective national registry conducted from year 2009 to 2015 in Malaysia. Patients with the diagnosis of MPN are recruited and information included sociodemographic factors, MPN subtypes, presence of JAK2 V617F mutation, presence of complications such as thrombosis, haemorrhagic events. The survival outcome of these patients were then analysed.

Results: A total of 865 patients were included for survival analysis. One hundred and fifty four patients died during the follow up; 57.8% were males and 42.2% were females.The mean survival duration was estimated to be 23.2 years. The ethnic distribution among the deceased were 50% of Malay, 40.9% of Chinese, 7.1% of Indian and 1.9% of others. JAK2 V617F mutation was present in 61.1% of patients who died.. The causes of death were mainly non-haematological related (55.2%) while haematological related was only 23.4%. Essential thrombocythemia (ET) had the best survival outcome followed by polycythemia vera (PV), hypereosinophilia syndrome (HES), MPN-unclassifiable (MPN-U) and primary myelofibrosis (PMF). PMF had 1.2 times of risk of death when compared to the other subtypes , $p<0.001$. The JAK2 V617F mutation had no influence in the overall survival (OS). Haemoglobin <12g/dL, haematocrit <35%, platelet <150 $x10^9$/dL and white blood count <5 $x10^9$/dL were significantly associated with the inferior OS ($p<0.001$). The arterial or venous thrombosis had no effect on the OS while those with bleeding events had a marginally significant better outcome ($p=0.04$). The grade 2 and above bone marrow fibrosis had a worse OS compared to those with grade 1 and below bone marrow fibrosis. Patients presented with vasomotor symptoms were found to do better than those patients without ($p=0.003$).

Conclusion: Patients with ET had the best OS while PMF had the worst OS. This is in conjunction with low blood counts, worsening bone marrow fibrosis and without vasomotor symptoms. The survival outcome of the MPN registry is instrumental for future policy development of effective healthcare in Malaysia.