ABSTRACT

Polycythaemia vera (PV), Essential Thrombocythaemia (ET) and Primary Myelofibrosis (PMF) are myeloproliferative neoplasms (MPN) which are commonly absent of BCR-ABL 1 translocation. They are characterized by the activation of the Janus Associated Kinase 2 (JAK 2) showing variable frequency. Studies on them are lacking in Sri Lanka. This study was done on the clinical and laboratory aspects of these diseases with the involvement of 37 patients attending the Haematology clinic, Colombo South Teaching Hospital (CSTH), Sri Lanka. PV was found in 28 patients (77.5%) and 5 PMF (12.5%) and 4 ET (10%) patients were included. The mean age at diagnosis was 56.4 years, 64 years and 62 years for PV, ET and PMF respectively. Female predominance was seen among PV (53.6%) and ET and PMF were common among males at 75% and 60% respectively. The diagnosis of PV was incidental among 45.1% and 32.1% presented with thrombosis of which arterial thrombosis was 21.4%. All ET patients were asymptomatic at presentation and PMF patients had peripheral arterial occlusion, symptomatic anaemia and constitutional symptoms at 20%, 40% and 40% respectively. All PV patents were positive for JAK2V617F and it was found among 50% and 60% of ET and PMF patients respectively. Initial Blood counts of PV patients were characterized by 45%-60% of Haematocrit (HCT) in 75%, $>450x10^3$/L of platelet count in all patients. Platelet count $>1000x10^3$/L was seen among 75% of ET patients and Haemoglobin was <10g/dl in 80% of PMF patients. All ET patients and 66.4% of PV patients were belonging to the high-risk category. In the risk stratification of PMF, DIPSS intermediate 1, 2 and high-risk groups of patients were seen as 20% in each category. Hydroxycarbamide was given to 78.6% of PV patients. HCT of 95.8% of them became <45% after the initial 3 months. All ET patients had Hydroxycarbamide and platelet count was not normalized in all of them. Constitutional symptoms and splenomegaly were improved in PMF who were treated with Ruxolitinib. Anaemia was improved with Lenalidomide in the patient who had it. Response to recombinant Erythropoietin was 33.3% among them.