Extension of 2016 World Health Organization (WHO) classification into a new set of clinical, laboratory, molecular, and pathological criteria for the diagnosis of myeloproliferative neoplasms: from dameshek to vainchenker, green and kralovics

Improved Clinical, Laboratory, Molecular, and Pathological (CLMP) 2017 criteria for myeloproliferative neoplasms (MPN) define the JAK2(V617F) trilinear MPNs as a broad continuum of essential thrombocythaemia (ET), polycythaemia vera (PV), masked PV, and post-ET or post-PV myelofibrosis (MF). Normal versus increased erythrocyte counts (5.8 x 1012/L) on top of bone marrow histology separate JAK2(V617F) ET and prodromal PV from early and classical PV. Bone marrow histology of the JAK2(V617F) trilinear MPNs show variable degrees of normocellular megakaryocytic, erythrocytic megakaryocytic and erythrocytic megakaryocytic granulocytic (EMG) myeloproliferation, peripheral cytoses, and splenomegaly related to JAK2(V617F) allele burden. MPL515 thrombocythaemia displays predominantly normocellular megakaryocytic proliferation. CALR (Calreticulin) thrombocythaemia initially presents with megakaryocytic followed by dual granulocytic and megakaryocytic myeloproliferation without features of PV. The megakaryocytes are large, mature, and pleomorphic with hyperlobulated nuclei in JAK2(V617F) ET and prodromal, classical, and masked PV. The megakaryocytes are large to giant with hyperlobulated staghorn-like nuclei in MPL515 thrombocythaemia. The megakaryocytes are densely clustered, large, and immature dysmorphic with bulky (bulbous) hyperchromatic nuclei in CALR thrombocythaemia and MF.

Biography
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