

什麼是骨髓增生腫瘤？臨床表現與鑑別診斷

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Myeloproliferative neoplasm (MPN) was previously called as myeloproliferative disorder (MPD), first recognized by William Dameshek in 1951. Initially, MPD included chronic myeloid leukemia (CML), polycythemia vera (PV), essential thrombocythemia (ET), primary myelofibrosis (PMF) and erythroleukemia. In 2001, WHO assigned MPD as chronic myeloproliferative diseases (CMPD), including CML, chronic neutrophilic leukemia, PV, ET, PMF, chronic eosinophilic leukemia, and MPN, unclassifiable. Little was known about the molecular pathophysiology of CMPD until recent years. Recognition of bcr-abl gene in CML and development of tyrosine kinase inhibitors have CML an unique disease in CMPD. Nowadays more genetic abnormalities have been identified for patients w/ MPN. Examples include PDGFRB, PDGFRA, and FGFR1 in eosinophilic disorders and JAK2 V617F mutation in BCR-ABL negative MPN. 2008 WHO classification for MPN is now available.

The manifestations of MPN are heterogeneous, mainly depending on which clone is highly proliferative. Diagnostic criteria are also available for each types of MPN. Reactive bone marrow, inflammation, and even metastatic malignancies should be the differential diagnoses of MPN. Genetic diagnoses are playing a more and more important role in the diagnoses.