

Practical application and clinical impact of the WHO histopathological criteria on bone marrow biopsy for the diagnosis of essential thrombocythemia versus prefibrotic primary myelofibrosis

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Aims: To evaluate the feasibility of the histopathological diagnosis of prefibrotic-early primary myelofibrosis (PM) as described in the World Health Organization (WHO) classification and to evaluate the clinical implications of prefibrotic-early PM in a series of patients previously diagnosed as having essential thrombocythemia (ET) according to the Polycythemia Vera Study Group criteria.

Methods and results: WHO criteria were applied to bone marrow biopsy specimens by two pathologists who then reclassified 127 cases as 102 ET (80.3%), 18 prefibrotic-early PM (14.2%) and seven fibrotic PM (5.5%). In 45 cases (35%), the final diagnosis was only reached by consensus. The megakaryocytic criteria that best discriminated between ET and prefibrotic-early PM were an increased nucleo-cytoplasmic ratio, presence of cloudlike nuclei, hyperchromatic-dysplastic nuclei, paratrabeular megakaryocytes and tight clusters. A histological score discriminated between ET (score ≤ 3) and PM (score ≥ 6), but 21 cases showed an intermediate ambiguous score. No significant differences were observed at diagnosis and at follow-up (median time 93 months) for thrombosis, major haemorrhage, laboratory data, transformation into overt myeloid metaplasia and survival.

Conclusions: The distinction between ET and prefibrotic-early PM is impaired by subjectivity in pathological practice and is of questionable clinical relevance, at least when considering individual patients.

Résumé en anglais

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