SURVIVAL AND DISEASE PROGRESSION IN ESSENTIAL THROMBOCYTHEMIA ARE SIGNIFICANTLY INFLUENCED BY ACCURATE MORPHOLOGIC DIAGNOSIS: AN INTERNATIONAL STUDY.


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SUMMARY:

This international study examined the role of bone marrow morphology in distinguishing essential thrombocythemia (ET) from early/prefibrotic primary myelofibrosis (PMF) that clinically presents like ET. Bone marrow specimens of 1104 ET, obtained within one year of diagnosis, underwent a central re-review. Diagnosis was confirmed as ET in 891 patients (81%) and revised to early/prefibrotic PMF in 180 (16%); 33 cases were not evaluable. Ten/15-year survival (76%/59% vs. 89%/80%), leukemic transformation (5.8%/11.7% vs. 0.7%/2.1%) and fibrotic progression (12.3%/16.9% vs. 0.8%/9.3%) rates were significantly worse in early/prefibrotic PMF vs. ET; the respective death, leukemia and myelofibrosis incidence rates per 100 patient-years were 2.7% vs. 1.3% (RR=2.1, p=0.0002), 0.6% vs. 0.1% (RR=5.2, p=0.001) and 1% vs. 0.5% (RR=2.0, p=0.04). Multivariable analysis confirmed these findings and also identified age >60 years (HR=6.7, p<0.0001), leukocyte count >11x10^9/L (HR=2.01, p=0.0002), anemia (HR=2.95, p=0.0001), and thrombosis history (HR=2.81, p<0.0001) as additional risk factors for survival.