

Leukaemia Section

Short Communication

Essential thrombocythemia

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Clinics and pathology

Disease

Chronic myeloproliferative syndrome.

Phenotype / cell stem origin

Pluripotent stem cell is involved.

Epidemiology

Annual incidence is less than $1/10^6$; sex ratio 1M/; median age 50-60 years.

Clinics

Often revealed by haemorrhages or thrombosis; splenomegaly is found in 50% of cases; blood data: the disease is defined by a thrombocytosis $> 600 \times 10^9/L$; the platelet count is actually often $> 1000 \times 10^9/L$.

Prognosis

Evolution: chronic disease; can evolve towards polycythemia vera or myelofibrosis, seldom towards ANLL; prognosis: often fair, is variable according to age and depends on haemorrhages, thromboses, and embolisms, which are the major causes of death in this disease.

Cytogenetics

Cytogenetics, morphological

A normal karyotype is found in 95% of cases; +9 is the only anomaly having been described in as far as 4 cases!

Genes involved and Proteins

Note: genes involved are unknown.

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