Leukaemia Section
Short Communication

Myelofibrosis with Myeloid Metaplasia (MMM)
Idiopathic myelofibrosis
Agnogenic myeloid metaplasia

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Clinics and pathology

Disease
Chronic myeloproliferative syndrome.

Phenotype / cell stem origin
Pluripotent stem cell is involved.

Epidemiology
Annual incidence: 2/10^6; slightly more frequent in males; age is usually over 50 yrs.

Clinics
Asymptomatic for a long time, revealed by symptoms related to the splenomegaly, or by anaemia/asthenia; splenomegaly is the major sign; hepatomegaly in 50%; blood data: anisocytosis, poikilocytosis (as tears drops, are characteristic); anaemia is frequent; hyperleucocytosis in 60%; thrombocytosis may be present; erythromyelemia.

Cytology
Bone marrow: fibrosis is major (fibrosis is a secondary event in this disease), while there is extramedullary hematopoiesis (myeloid metaplasia) in the spleen, the liver, and anywhere (e.g. skin).

Prognosis
Evolution: this is a chronic disease, with a proliferative stage followed by a pancytopenic stage; pancytopenia and portal hypertension are the major causes of death in this disease; evolution towards ANLL is found in 15-20% of cases; prognosis: is highly variable; survival is frequently over 10-15 yrs, but death occurs within a year in some cases; cases with pancytopenia directly at diagnosis bear a worse prognosis; probable prognostic factors are: the presence of an abnormal karyotype and a low haemoglobin level, possibly a low platelets count and a high WBC, a higher age, and hepatomegaly.

Cytogenetics

Cytogenetics, morphological
An abnormal karyotype is found in 40% of cases at diagnosis: der(1), in particular partial trisomy 1, del(5q) or -5, -7, +8, +9, del(13q), del(20q) are seen solely or simultaneously in 5-10% of cases with chromosome anomalies, and other (various) anomalies in 40%.

Genes involved and Proteins
Note: genes involved are unknown.

To be noted
'Acute myelofibrosis' is a megakaryoblastic leukaemia (M7 ANLL) with prominent fibrosis.

References


This article should be referenced as such: