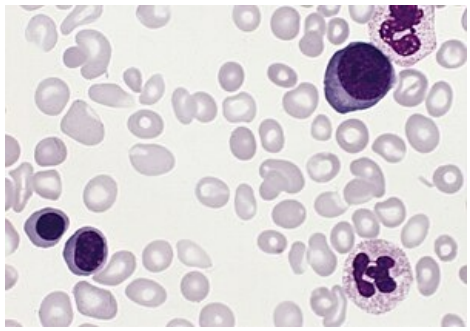
Polycythaemia vera

Polycythaemia vera (PV, primary polycythaemia, Vaquez's disease) is a disease with a high number of erythrocytes + high concentration of Hb → increases blood viscosity + arterial thrombotisation (cause of death is thrombosis of coronary + cerebral arteries).



Polycythaemia vera blood count

Pathogenesis

- Clonal proliferation of a pluripotent hematopoietic stem cell, kt. differentiates mainly into the erythrocyte line;
- increased sensitivity of BFU-E progenitors to the effects of erythropoietin;
- the possibility of differentiation into erythroid precursors even without erythropoietin;
- there is also a slight increase in bb. other rows.

Clinical picture

- Headache , dizziness , etc.,
- digestive problems, frequent Gastroduodenal ulcer disease ,
- pruritus,
- frequent bleeding / conversely arterial and venous thrombotic events, incl. coronary artery obliteration,
- Arthritis uratica (gout),
- brownishness, cyanosis ,
- splenomegaly (in advanced stages up to the pelvis).

Diagnostics a diff. dg.

- Increased value of Hb + HTK (50-70%),

  can be masked if plasma is also increased;

- there may be leukocytosis + a slight shift to the left,
- often also thrombocytosis,
- blood oxygen saturation < 92% can be the cause of secondary polyglobulia ,
- low ferritin values more common in primary polycythemia than in secondary polyglobulia; concentration of vitamin B12 + its binding capacity increased, conc. folate reduced,
- in PV, the concentration of endogenous erythropoetin is reduced,
- the histological height is essential. bone marrow (PV image different from reactive polyglobuli) + molecular-biological examination,
- dg. PV rests hl. in the detection of general signs of myeloproliferative disease, in the case of embarrassment to exclude a secondary cause (difficult),
- frequent thrombotic/bleeding events (in patients with secondary polyglobulia few).

Therapy

- Medical venipuncture , erythrocytapheresis (more expensive, exceptionally),
- IFN-α ,
- hydroxyurea,
- anagrelide + therapeutic venipuncture in patients resistant to IFN + hydroxyurea,
- treatment of pruritus + hyperuricemia .

Prognosis

- Average survival 15 years,
- cause of death: thrombosis, acute leukemia , other cancer, bleeding , etc.[1][2]

Summary video

Links

Reference

1. NEČAS, Emanuel. *Patologická fyziologie orgánových systémů : Část I.* 2. edition. V Praze : Karolinum, 2009. 379 pp. ISBN 978-80-246-1711-4.
2. DÍTĚ, P.. *Vnitřní lékařství.* 2. edition. Praha : Galén, 2007. ISBN 978-80-7262-496-6.

Related articles

- Myeloproliferative disease
- Secondary polyglobulia
- Erythrocytosis