Diseases caused by increased number of erythrocytes

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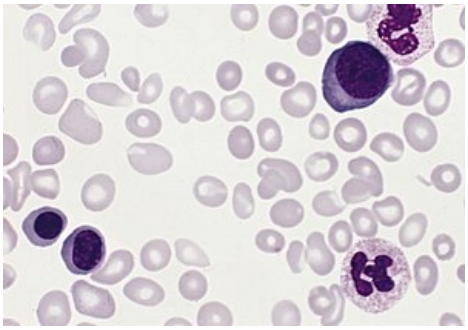
Polycythaemia vera

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

Secondary polyglobulia

Secondary polyglobulia is mostly compensatory in *chronic hypoxia* (leads to ↑ EPO) when staying at high altitude, chronic lung disease, right-to-left shunts, methemoglobinaemia.

Less often, the cause of ↑ EPO is unclear/a manifestation of a disease (kidney tumors, uterine myomatosis, polycystic kidney disease, etc.).

The clinical picture

- mainly symptoms of underlying lung/heart disease,
- plethoric appearance, cyanosis,
- headaches, dizziness, visual disturbances.

Laboratory Tests

- ↑ erythrocytes, HTK > 48%, oxygen saturation <92%, ↑ EPO (dif. dg. Polycytaemia vera).

Therapy

- Th. of underlying disease, venipuncture (↓ blood viscosity + reduces the risk of complications).[3]

Erythrocytosis

False polyglobulia is caused by a ***decrease in volume of plasma***, total erythrocyte volume is normal:

- ***Stress polycythemia*** in young people (+ hypertension, obesity = *Gaisböck's syndrome*)
- ***Relative polyglobulias*** in dehydration, extensive burns, etc.

Neonatal polycythemia

Neonatal polycythemia is defined as a central venous **hematocrit > 0,65**. However, even with a hematocrit> 0.70, only a small proportion of neonates have clinical signs of hyperviscosity. The **clinical picture** includes lethargy, hypotension, hyperbilirubinemia and hypoglycemia. Polycythemia may also be involved in the development of convulsions, stroke, renal venous thrombosis and necrotizing eneterocolitis.

Causes of neonatal polycythemia:

- Fetal growth restriction (FGR, IUGR);
- maternal hypertension;
- maternal diabetes;
- chromosomal defects: trisomy 21, 18, 13;
- twin-to-twin transfusion;
- delayed umbilical cord rupture;
- endocrine disorders: tyreotoxicosis, congenital adrenal hyperplasia (CAH).

Treatment is controversial and may not be necessary in neonates with mild symptoms (eg borderline glycaemia, impaired peripheral blood flow). Treatment is indicated in neonates with a hematocrit> 0.65 and symptoms with a potentially severe prognosis (eg refractory hypoglycaemia, neurological symptoms). A partial exchange transfusion is performed with a crystalloid solution (eg physiological solution) with a target hematocrit of 0.55.[4]

🔎 *For more information see Polycythemia (neonatology).*

Links

Related articles

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.
3. KLENER, P. *Vnitřní lékařství.* 3. edition. Galén, 2006. ISBN 80-7262-430-X.
4.

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