Extracorpuscular hemolytic anemia

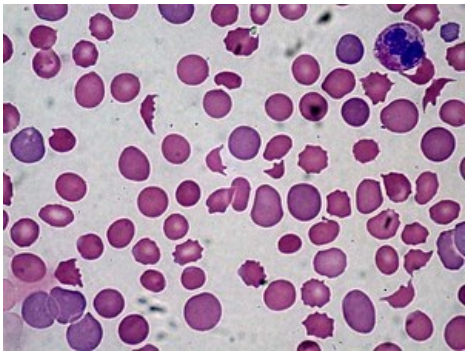
It is anemia when the **cause** of hemolysis lies **outside the erythrocyte**. They are mostly **acquired**. We divide them into:

- **immune** - an antibody binds to the erythrocyte, which activates complement or marks the cell for elimination in the monocyte-macrophage system;
- **non-immune** - the erythrocyte is damaged mechanically or by toxins, its lysis occurs or it is removed by the monocyte-macrophage system due to its changed shape.

Anemia from mechanical damage to erythrocytes

Characteristic **presence of deformed erythrocytes** – **fragmentocytes** (schistocytes):

- **In the presence of thrombi in the microcirculation** (microangiopathic hemolytic anemia) – DIC, TTP, hemolytic-uremic syndrome, Kassabach-Merrit syndrome (cavernous hemangioma + consumptive coagulopathy + anemia).
- During implantation of **artificial valves.**
- In case of **tumor angiopathy** (growth of a tumor into a blood vessel - e.g. liver cancer or Grawitz's tumor).
- In malignant **hypertension**.



Disseminated intravascular coagulation (DIC) with microangiopathic hemolytic anemia

Anemia from damage to erythrocytes by toxins and parasites

Bacterial exotoxins (phospholipases, hemolysins - Clostridium welchii, pneumococci, streptococci...), **snake and spider venoms**.

Malaria – breakdown of erythrocytes during multiplication of plasmodia.

Anemia from erythrocyte damage by antibodies and complement

Binding of immunoglobulins (autoantibodies) or **complement components** to erythrocytes, which are thus opsonized and captured by splenic macrophages (a more important role of macrophages than MAC-mediated hemolysis).

Primary forms occur without a predisposing disease, **secondary forms** are accompanied by some autoimmune diseases (systemic lupus erythematosus), tumors (lymphomas, carcinomas), viral infections, use of certain drugs.

The diagnosis is carried out by the **direct Coombs test** – antibodies (from rabbit serum) against immunoglobulins are bound to immunoglobulins attached to the surface of erythrocytes and this leads to their agglutination, with the indirect Coombs test free antibodies are determined.

In paroxysmal cold hemoglobinuria, hemolysin-type antibodies are present, which are activated by cold..

Fetal erythroblastosis (morbus haemolyticus neonati)

It occurs **when the mother and fetus are incompatibility in the Rh system** (mother Rh–, fetus Rh+). The mother creates antibodies against the D- antigen on the surface of the erythrocytes of the fetus after previous immunization (previous pregnancy (including miscarriage), when maternal and fetal blood are mixed during childbirth, or transfusion of Rh+ blood to the mother). During the following pregnancy, the antibodies cross the placental barrier and **cause hemolysis in the blood of the fetus**. This can lead to:

- **Jaundice** (icterus neonati gravis) – unconjugated bilirubin passes through the immature blood-brain barrier and leads to a greenish-yellow coloration of the basal ganglia, nuclei of the thalamus, cerebellum and olive – core icterus (Kernikterus) -damage to the CNS.
- **Flushing of immature erythrocytes** (erythroblasts) - hence erythroblastosis.
- **Extramedullary erythropoiesis** – hepatosplenomegaly.

The overall condition of the fetus can be different, from postpartum anemia with jaundice to CNS damage, the most severe form (**hydrops foetus congenitus**) leads to intrauterine death of the fetus and its maceration (foetus maceratus).

Other extracorpuscular causes of hemolysis

Copper - in hemodialysis patients and patients with Wilson's disease.

Extensive burns – thermal damage to erythrocytes.

Hypersplenism – combination of splenomegaly with anemia, or with leukopenia a thrombocytopenia.

Links

related articles

- Anemia
- Hemolytic anemia corpuscular
- Rh system

Source

- PASTOR, Jan. *Langenbeck's medical web page* [online]. [cit. 12.4.2010]. <http://langenbeck.webs.com>.