

Idiopathic thrombocytopenic purpura

Idiopathic (autoimmune) thrombocytopenic purpura (ITP), is a condition in which the rapid breakdown of platelets occurs for an unknown reason with the participation of immune mechanisms. ^[1]This is the most common acquired bleeding disease in childhood.^[2]

Etiopathogenesis

Autoantibodies are formed in the body (B-lymphocytes with the help of CD4+ T-lymphocytes) against platelet surface antigens, no. against glycoprotein IIb-IIIa. The trigger is often an infection (upper respiratory tract infection, more rarely varicella, mumps, rubella, EBV infection, vaccination with a live vaccine). Platelets with bound antibodies are absorbed by macrophages and then disappear mainly in the spleen. Autoantibodies inhibit megakaryopoiesis, which results in reduced platelet production by bone marrow megakaryocytes, thrombopoietin levels are normal.^[2]

Clinical picture

Acute form

It is a childhood disease with a rapid course and often spontaneous resolution. Circulating immunocomplexes with an affinity for platelets are then quickly taken up by cells MMS. It usually follows a banal viral infection. Bleeding manifestations are present, **sudden onset** (within hours) - generalized purpura, haematomas, bleeding from mucous membranes, bleeding into organs (CNS) occur. Fortunately, the acute form is **rare**.

Chronic form

A drop in platelets below $150 \times 10^9/L$ lasting more than 6 months is typical;^[2]. A disease of **adult age** (more often women) with an insidious onset and chronic course, spontaneous remissions are rare. Principle: autoantibodies against platelet antigens → rapidly absorbed by MMS (spleen: production of antibodies + absorption and degradation of altered platelets); More common are "serious organ bleeding" (often fatal to the CNS).

Diagnostics

The diagnosis is often clinical - it is necessary to rule out thrombocytopenia of other etiology (in childhood marrow depression in acute leukemia, in adulthood MDS). We demonstrate antiplatelet antibodies. ITP can be a manifestation of SLE / B-lymphoproliferation.

Laboratory examination

- the number of thrombocytes variously reduced, in severe forms below $10 \times 10^9/l$;
- other parameters KO and hemostatic tests within the norm;
- in bone marrow proliferation of megakaryocytes.

Therapy

- the basis of immunosuppression - **prednisone** 0.5-1 mg/kg (after achieving remission, we continue with the maintenance dose), possibly cyclophosphamide, cyclosporine;
- **splenectomy** (in patients with increased destruction of platelets in the spleen);
- anti-CD20 monoclonal antibody **rituximab**;
- intravenous immunoglobulins in high doses (in the event of a deep drop in platelets + more pronounced bleeding symptoms with the risk of permanent consequences/life-threatening);
- in the period of more pronounced bleeding symptoms, non-specific haemostyptics at the same time, in extreme cases platelet transfusions (short-term effect).

Links

Related articles

- Thrombocytopenia
- Thrombocytopenia of the newborn



Purpura - petechiae

- Hematological tests
- Pre-transfusion examination
- Blood coagulation test
- Purpura

References

1. KLENER, P. *Vnitřní lékařství*. 3. edition. Galén, 2006. ISBN 80-7262-430-X.
2. LEBL, J - JANDA, J - POHUNEK, P. *Klinická pediatrie*. 1. edition. Galén, 2012. 698 pp. pp. 552-554. ISBN 978-80-7262-772-1.

Literature

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