Acute autoimmune thrombocytopenic purpura

Idiopathic thrombocytopenic purpura (ITP) is the most common cause of thrombocytopenia in childhood.

Pathogenesis

Causes

- autoantibodies IgG or IgM (binds to the surface of platelets).

Plateletss coated with bound antibodies are then taken up and destroyed by RES cells, mainly in the spleen.

The spleen not only removes sensitized platelets from the circulation, but also is a source of antibody production. The formation of antibodies can occur for no apparent reason, but can be induced by a common viral illness. Rapid destruction of platelets in the RES is also caused by the interaction of circulating immune complexes (antigen-virus antibody) with platelets. These immune complexes also damage the vascular wall. Platelets are collected on the vascular wall damaged in this way, which worsens the thrombocytopenia even more.

The body compensates for the loss of platelets by increasing their production in the bone marrow, which is why we usually find an increased number of megakaryocytes, among which the younger forms predominate.

Clinical picture

- The disease most often affects children aged 2-4 years;
- both sexes are affected equally often;
  - children older than 10 years are more likely to fall ill with a chronic form;
  - children < 1 year also more often in a chronic form + often in connection with another autoimmune disease.

In the anamnesis, we can find out information about a viral disease at an interval of 2-3 weeks.

The disease usually begins from full health with the appearance of:

- petechia,
- hematomas,
- mucosal bleeding (bleeding occurs spontaneously or after minimal trauma).

On the other hand, petechiae may also be absent in patients with platelets < 25 000/µl, most likely because the platelets are young and thus more functionally competent. We can also encounter hematuria and bleeding into the GIT.

Splenomegaly and lymphadenomegaly are rare, as well as temperature, loss of appetite, pain in the limbs. The finding of splenomegaly and lymphadenomegaly requires the exclusion of other causes of thrombocytopenia (m. Gaucher, leukemia, SLE).

At the same time, the overall condition of the child is usually very good and without alteration (in approx. 1% of patients bleeding into the CNS can occur, which is also the most common cause of otherwise very low mortality).

Clinical stages of ITP

1. no bleeding, platelets > 20 000/µl;
2. petechiae, hematomas, platelets < 20 000/µl;
3. obvious bleeding, platelets < 20 000/µl.

Laboratory findings

- In the blood count we find an isolated decrease in platelets, the exception being a drop below 20 000/µl,
- sometimes we can find eosinophilia, sometimes mild anemia or leukocytosis,
- thrombocytes have relatively large dimensions (young platelets),
- if we perform an examination of the bone marrow, we find an increased number of megakaryocytes with a predominance of immature forms.

If there is any doubt about the diagnosis of ITP, a bone marrow aspiration should be performed!

Therapy

There is still no consensus among hematologists on the question of whether or not to treat children with acute ITP.

In general, it is recommended to treat children with platelet levels < 20 000/µl, when there is a risk of intracranial bleeding, and children with significant bleeding manifestations. Therapy probably does not have a significant effect on the duration of the disease as such, but it can accelerate the rise of platelets to "safe" values.

- Regime measures: restriction of physical activity → bed rest.
- corticoids: methylprednisolone 10–30 mg/kg/24 h i.v. during a 30-minute infusion for 3 days, then continue with prednisone 1-2 mg/kg/day for 10-20 days.

Alternatively, with a milder course, the methylprednisolone boluses can be omitted in the beginning.

- HDIVIG: 7S preparations (e.g. Endobulin®, Flebogamma®, Venimmun®) in a dose of 800 mg/kg/day single i.v., alternatively in a dose of 400 mg/kg/day i.v. for 5 days (Šašinka et al.), possibly repeat the next day if there is insufficient response,
  - HDIVIGs should only be reserved for the inpatient treatment of patients with extremely low platelet counts or for patients who are already bleeding into vital organs, especially the CNS,
  - 80% of children will have an immediate rise in platelets above 100 000.

Platelet concentrate, plasmapheresis - are reserved for patients with life-threatening bleeding.

Course and Prognosis

- The course of typical ITP is usually mild and most children recover within 6 months even without treatment,
- relapses are rare, but can occur several years after the initial episode, especially in connection with intercurrent infection,
- children with a sudden onset and significant thrombocytopenia have the best hope for a quick recovery,
- if thrombocytopenia persists for more than 6 months, it is already a chronic form of ITP.

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- Havránek J: Acute autoimmune thrombocytopenic purpura