Acute immune thrombocytopenic purpura

This article has been translated from WikiSkripta; the **formatting** needs to be checked.

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

Pathogenesis

Causes

- IgG or IgM antibodies (they bind to the surface of thrombocytes).

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

The spleen removes sensitized platelets from the circulation and is a source of antibody production. Antibody production can occur for no apparent reason, but can be induced by a common viral disease. The rapid destruction of platelets in RES is also induced by the interaction of circulating immunocomplexes (antigen-viral antibody) with platelets. These immunocomplexes also damage the vessel wall. Platelets bind to a damaged vessel wall easily, which exacerbates thrombocytopenia.

The body compensates for the loss of platelets by their increased production in bone marrow, therefore an elevated megakaryocyte count is present, immature forms can be found among them.

Clinical picture

thumb | 250 px | Petechia even purpura

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

In the anamnesis we can find out about the viral disease in the interval of 2-3 weeks.

The disease usually begins in full health:

- petechiae,
- hematoma,
- mucosal bleeding (bleeding is spontaneous or after minimal trauma).

On the other hand petechia may be absent in patients with thrombocytes < 25 000/µl, most likely because thrombocytes are young, have higher endurance and more functionally capable. Můžeme se setkat i s hematurií a krvácením do GIT.

Splenomegaly a lymphadenomegaly is rare, also increased temperatures, nechutenství, bolesti končetin. Nález splenomegalie a lymfadenomegalie vyžaduje vyloučení jiných příčin trombocytopenie (m. Gaucher, leukemie, SLE).

The overall condition of the child is usually very good and without alterations (approximately 1% of patients may bleed into the CNS, which is also the most common cause of the otherwise very low mortality).

Clinical stages of ITP

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

Laboratory findings

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

If in doubt about the diagnosis of ITP, 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.

It is generally recommended to treat children with platelet counts < 20 000/µl, when there is a risk of intracranial haemorrhage, and children with significant bleeding symptoms. Therapy is unlikely to have a significant effect on the duration of the disease as such, but it may accelerate the rise of platelets to "safe" levels.

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

Alternatively, with a milder course, boluses of methylprednisolone may be omitted at the outset.

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

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

Course of the disease and prognosis

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

Reference

Related articles

- Thrombocytopenia of newborns
- Blood count/Haematological examinations
- Pre-transfusion examination
- Examination of blood clotting
- Purpura
- ITP

Source

- Havránek J: Acute immune thrombocytopenic purpura