

# Thrombotic thrombocytopenic purpura

It is a rarer disease compared to ITP , but has a serious prognosis.

- Fevers,
- Hemolytic anemia (microangiopathic, schistocytes in blood smear),
- Thrombocytopenia with bleeding manifestations,
- Neurological symptomatology,
- Renal impairment.

## Pathogenesis

### Idiopathic form

- In the plasma, **von Willebrand factor of high molecular weight** (as a result of the deficiency of a specific plasma metalloproteinase) with the ability to induce platelet microthrombotization.

### Hereditary form

- Mutation of the metalloproteinase gene from the ADAMTS group on chromosome 9,
- In the acquired forms, antibodies against metalloproteinases have been demonstrated,
- In order to cause a TTP attack, the participation of other agents (infection, etc.) is necessary.
- Histologically: multiple hyaline microthrombi in arterioles (central CNS + kidneys).

### Secondary thrombotic thrombocytopenic purpura

- In patients after allogeneic transplantation , in systemic diseases, generalized malignancies, after administration of certain drugs (quinine, ticlopidine), complications of intestinal infections.

## Clinical course

- severe, violent,
- Patient at risk of ischemia + CNS hemorrhage + renal failure,
- There may be extensive bleeding into the skin ,
- Acute forms with relapses of thrombocytopenia / chronic forms.

## Therapy

- **Recurrent forms** – immunosuppressants (prednisone, cyclophosphamide), rituximab,
- **Idiopathic forms** – based on administration of fresh frozen plasma + plasmapheresis ,
- **Secondary forms** – therapy of the underlying disease, discontinuation of suspected drugs,
- **Hemolytic uremic syndrome** (HUS) = organ limited form of TTP,
  - **Epidemic form** – childhood disease, preceded by enterocolitis ( E. coli ); hemolytic anemia, thrombocytopenia, renal impairment; th.: anti-infective + symptomatic th., or temporary hemodialysis,
  - **Sporadická forma** – intestinal symptomatology is absent, symptoms are more varied; th.: plasmapheresis.

## Links

### Related articles

- Thrombocytopenia
- Thrombocytopenia of the new born
- Hematological examination
- Pre-transfusion examination
- Examination of blood coagulation
- Purpura

### References

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