Primary myelofibrosis

Primary idiopathic myelofibrosis, also known as myeloid metaplasia, is a myeloproliferative disease with an incidence of 0.5-1.5 per 100,000 population. Approximately 10-20% of patients then develop AML. Younger patients (under 55 years of age) live after the diagnosis of myelofibrosis live for more than 10 years, older than about 5 years.

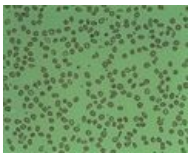
Etiology

The origin of this disease is not yet known. However, a primary disorder at the stem cell level is presumed. This theory is confirmed by the cure of some patients through bone marrow transplantation.

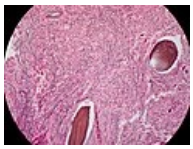
Pathogenesis and diagnostics

In primary myelofibrosis, there is a gradual attenuation of hematopoiesis in the bone marrow with its simultaneous fibrotization. **Collagen III** begins to accumulate in the tissue. This change is **reversible** and is likely to be caused by the production of excessive amounts of the growth factors **PDGF** and **bFGF** and the cytokine **TGF-β,** which is produced by megakaryocytes. **Fibroblasts** in the bone marrow are normal.

Simultaneously with this change, **extramedullary hematopoiesis** (formation of blood elements outside the bone marrow) is activated, which causes a small part of immature forms of blood elements (erythroblasts, myelocytes and promyelocytes) to be released into the blood. This is used in diagnostics. At the same time, we can find teardrops in erythrocytes in the blood. Extramedullary hematopoiesis leads to splenomegaly and hepatomegaly.



Poyclocytosis - erythrocyte in the shape of a drop

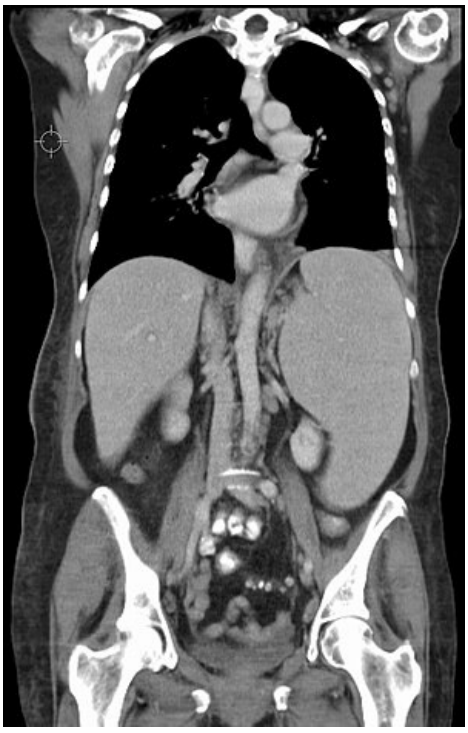


Myelofibrosis (reticular fiber staining)

Clinical picture

This disease consists of two phases. The first, so-called prefibrotic, is accompanied by only mild reticular fibrosis with hypercellular bone marrow. In the second, so-called fibrotic, phase, massive reticular and collagen fibrosis already occursApproximately 30% of patients do not have any symptoms and myelofibrosis is diagnosed at random. The most common symptoms include:

- B symptoms (weight loss, subfebrile to febrile, night sweats);
- fatigue;
- nausea.



Expressive splenomegaly in late stages

Therapy

Most methods are only palliative, the only curative option is **allogeneic transplantation.** Palliative approaches include cytoreductive therapy (interferon α, hydroxyurea), splenectomy, androgen and EPO or JAK kinase inhibitors.

Links

Related articles

- Myeloproliferative diseases
- Splenomegaly

Literature

- NEČAS, Emanuel. *Patologická fyziologie orgánových systémů.* - vydání. Karolinum, 2009. 379 s. ISBN

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- ČEŠKA, Richard. *Interna.* - vydání. Stanislav Juhaňák - Triton, 2015. ISBN 9788073878856.