Acute myeloid leukemia

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Acute myeloid leukemia (AML) is a disease caused by malignant transformation of hematopoietic stem cells that differentiate into myeloid or myelomonocytic precursor cells (blasts), more rarely erythroid or megakaryocyte blasts. The autoregulatory processes fail, the cells do not differentiate, but have preserved their ability to proliferate.

Epidemiology

The risk of AML manifestation increases with higher age (it is most common in people over 80 years) with an incidence rate of 3,4 per 100 000.

Etiologiy

The cause of the disease is unknown, some of the risk factors are known to include exposure to radiation or to certain viruses. Patients with severe congenital abnormalities (Down syndrome, Klineferter syndrome etc.) are at higher risk for developing AML. The mutation of the gene coding for the receptor tyrosine kinase is present in many cases and results in the transformation of *normal* hematopoiesis to *leukemic* (or ***leukemogenesis***). The resulting overgrowth of pathological leukocytes gradually completely suppresses healthy blood elements in the peripheral blood.

upright=1.4|náhled|video in English: definition, pathogenesis, symptoms and complications, diagnosis, treatment, AML starts at 6:00:00

Microscopic image

- **bone marrow** is notably **hypercellular** with massive infiltration of leukemic cells (precursor cells - blasts - more than 30 % of cells (normally less than 5 %), some contain rod-shaped formations of azurophilic granule material - **Auer rods**), mature granulocytes are missing in the infiltrates
- after therapy comes remission caused by the disappearance of blasts, the bone marrow is then hypocellular
- a condition in which leukemic infiltration with suppression of non-tumour hematopoiesis is present in the marrow (but the clinical picture of leukemia does not develop) is referred to as **smouldering leukemia** (also pre-leukemia)
- extramedullary **infiltrates** are in the **liver** (mainly periportally), **spleen** and **lymph nodes** (infiltration in these organs is less pronounced than in chronic leukemias, therefore hepatosplenomegaly is not so pronounced), then in the **meninges**, **kidneys**, etc.
- some infiltrates (especially retroorbital and epidural) have a greenish color on incision, which quickly disappears in the air - such a deposit is referred to as granulocytic sarcoma (also myeloid sarcoma, **chloroma**). Its green color is attributed to the accumulation of the enzyme *myeloperoxidase (MPO)* in the blasts

Clinical manifestations

- resulting from anemia - paleness, fatigue, exhaustion and more,
- resulting from thrombocytopenia - bleeding (petechiae, ecchymosis, epistaxis),
- resulting from neutropenia - infections (oral cavit, upper respiratory tract and others),
- skin infiltrates and neurological symptoms of CNS damage

Findings in peripheral blood

- typical **hiatus leucaemicus** - the presence of immature blast cells and mature cells without intermediate stages of granulopoiesis (promyelocytes, myelocytes, metamyelocytes)
- more than 5 % of blasts in peripheral blood (less then 5 % and the presence all stages of granulopoesis is characteristic for CML)
- the number of leukocyte elements in the blood is usually higher than 100.10^9/l (norma je 4–10.10^9/l).

Classification

- *WHO classification* based on immunophenotypization (different CD antigens in the membrane of blasts):

1. AML with genetic abnormalities,
2. AML with with multilineage dysplasia (involvement of more than one abnormal myeloid cell type)
3. AML related to previous chemotherapy or radiation

4. Unspecified AML including those that do not fall into one of the above groups - **FAB classification** (French-American-British):

    - M0 - acute myeloid undifferentiated - no signs of myeloid differentiation
    - M1 - acute myeloid with minimal maturation - undifferentiated myeloblasts
    - M2 - acute myeloid with maturation - differentiated myeloblasts
    - M3 - acute promyelocytic - large promyelocytes with abundant granules and Auer rods
    - M4 - acute myelomonocytic - proliferation of myeloid and monocytic precursors
    - M5 - acute monocytic - presence of monoblasts and monocytes
    - M6 - acute erythroid - presence of myeloblasts and erythroblasts
    - M7 - acute megakaryoblastic - presence of megakaryoblasts and megakaryocytes

    - acute basophilic leukemia
    - acute myelofibrosis
    - acute panmyelosis with fibrosis

- the most common types of leukemias are **M1, M2 and M4**,
- type **M3** has a special clinical and biological behaviour when compared to the other forms of AML - often associated with rapid onset of DIC (bleeding due to the consumption of clotting factors) that can cause intracranial haemorrhage
- types **M4 and M5** typically infiltrates into organs (liver, spleen, skin, CNS)

Therapy

Patients that develop **leukostasis** (also symptomatic hyperleukocytosis) are treated with a series of leukodepletic procedures in order to **reduce the number of leukocytes** to less than $50{,}000 \times 10^9$/l, which is shown to reduce the risk of developing tumor lysis syndrome.

Curative treatment aims to achieve complete remision (CR). First is induction chemotherapy that combines antracykline a cytarabine. After achieving remission (normal blood count in peripheral blood and number of blasts in bone marrow is under <5 %), **consolidation therapy** is initiated and its composition varies according to the patients' prognosis. High-risk patients are directed to allogeneic hematopoietic stem cell transplantation after the consolidation treatment is ended.

Paliative care is initiated in patients whose general condition, age, comorbidities or personal preferences do not allow them to choose curative treatment. The patient is administered low doses of cytarabine, cytoreductive treatment with hydroxyurea and complex supportive care.

Treatment of relapse requires high-dose chemoterapy and allogeneic transplantation. However, not everyone can tolerate such intensive therapy, so the risks must be considered individually.[1]

Links

Related articles

- Leukemia
- Acute lymphoblastic leukemia
- Chronic myeloid leukemia
- Chronic lymphatic leukemia
- Hairy cell leukemia

References

1.

Sources

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Template:Navbox - onemocnění krve

Kategorie:Patologie Kategorie:Hematologie Kategorie:Vnitřní lékařství Kategorie:Onkologie