Plasmocellular myeloma

Plasmocellular myeloma, or **plasmacytoma**, is a malignant tumor of the plasma cells. It is usually found in the bone marrow, where it destroys bone and can cause pathological fractures. If there are more foci, we call it **multiple myeloma** (Kahler's disease). Tumor cells produce protein chains - paraprotein. However, there are rarely non-secretory myelomas in which no paraprotein can be detected in serum.

Patology

Macroskopy

Myeloma takes the form of dark red osteolytic deposits in the bone marrow (calva, vertebrae, pelvis, humeral shaft, femur).

Complication

- Pathological fractures (there is a risk of spinal cord lesions in the case of vertebrae).
- AL amyloidosis
  - Myocardium - rhythm disorders, heart failure.
  - Large intestine - malabsorption.
  - Kidneys - renal failure (proteinuria, periorbital edema, uremia).
  - Liver, blood vessels, nodes...
  - Tongue - macroglossis.
- Myeloma kidney - clogging of tubules by protein chains - Bence Jones protein.

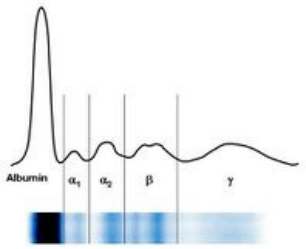
It threatens the patient's life

- Failure of an organ affected by amyloidosis,
- bronchopneumonia - develops after vertebral fracture and spinal cord injury,
- ifunction in bone marrow tumor generalization (same as in leukemia).

Clinical part

Epidemiology

The disease affects men more often than women (1.5: 1). The incidence is rising from the age of 50, the median age is 70 years. The incidence in the Czech Republic is 3-4: 100,000 inhabitants / year. The etiology of the disease is unknown. It is almost non-existent in childhood and very rare in young people.



Normal electrophoresis of the plasma proteins

Clinical picture

The initial period may be completely asymptomatic (in indolent lymphomas), but pain in the back and ribs predominates in> 70% of patients. Pain is tied to physical activity.

The clinical picture consists of a set of symptoms hidden under the abbreviation **CRAB**:

- **C**alcium - hypercalcaemia, which is associated with bone loss. Hypercalcemia also causes depression, lethargy and weakness.
- **R**enal failure – renal failure due to tubulopathy, uncommon myeloma,
- **A**nemia – normocytic normochromic anemia is the most common symptom, it is associated with weight loss, fatigue, shortness of breath and paleness
- **B**one – bone involvement - very common, tumor cells in the bone marrow produce osteoclast-activating factors, leading to bone osteolysis and pathological fractures that occur in up to 70% of patients.


Electrophoresis with monoclonal gamapathy

Susceptibility to infections, manifestations of hyperviscous syndrome (headache, vision disorders) and palpable infiltrates over osteolytic deposits (especially on the skull) also contribute to the clinical picture.

Investigation methods

- X-ray - reveals pathological lesions (often affected skull, spine, but myeloma can be in virtually any bone), as well as CT, MRI, PET,
- **determination of paraprotein in blood** - electrophoresis of plasma proteins (see picture), serum paraprotein concentration is usually> 30 g / l,
- **free chain determination** (FLC) - the most sensitive method, it also detects non-secretory myeloma, when the paraprotein determination would be negative, the normal ratio of kappa: lambda light chains is 2: 1, in the case of myeloma it is, for example, 350: 1,

- determination of Bence Jones protein in urine,
- bone marrow examination - the number of plasma cells usually exceeds 10% of all nuclear cells in the marrow, as well as immunohistochemical and cytogenetic examination (occurrence of deletions, trisomies that are of prognostic significance),
- cytogenetic examination - numerical and structural changes of chromosomes, important for prognosis
- laboratory examination,
  - normocytic normochromic anemia, mild leukopenia and thrombocytopenia
  - often increased sedimentation,
  - hypercalcemia,
  - renal function - increased urea and creatinine,
  - examination of LD, thymidine kinase and beta2-microglobulin, hyperuricemia - their increased values have a negative prognostic significance.

Clinical stages and variants

Clinical stages

Classification according to Durie and Salmon

- **clinical stage I** - paraprotein concentration IgG <50 g / l, or IgA <30 g / l, proteinuria <4 g / 24 hours, no osteolytic changes without hypercalcemia,
- **clinical stage II** - values are between stages I and III
- **clinical stage III** - IgG paraprotein concentration> 70 g / l, or IgA> 50 g / l, proteinuria> 12 g / 24 hours, multiple bone deposits, hypercalcemia> 2.75 mmol / l, hemoglobin concentration <85 g / l ,
- subclassification A, B according to the value of serum creatinine (renal impairment),
  - **subclasification A** – serum creatinine ≤ 177 µmol/l,
  - **subclasification B** – serum creatinine > 177 µmol/l.

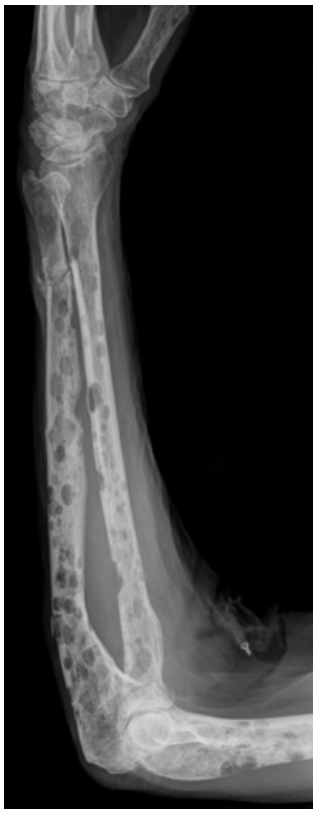
Clinical variants

Asymptomatic myeloma,

- smoldering myeloma,
- indolent myeloma,
- stage I multiple myeloma,

Symptomatic myeloma,

- stage II and III multiple myeloma,
- solitary plasmacytoma.



Multiple osteolytic lesions of the myelom

Therapy

- If myeloma is asymptomatic, it is not treated, it is only monitored,
- if symptomatic, it is treated:
  - younger patients are indicated for autologous hematopoietic stem cell transplantation,
  - elderly patients are indicated for chemotherapy,

- chemotherapy - cyclophosphamide and dexamethasone (or thalidomide) are used as standard, bortezomib has been registered in the Czech Republic since 2009 for primary treatment
- radiotherapy - for painful bone deposits,
- palliative treatment - in patients with severe comorbidities, melphalan and prednisone, or radiotherapy.

Links

Related articles

- Plasma cell myeloma
- Amyloidosis

External links

- MUDr. Zbyněk Mlčoch: Mnohočetný myelom – příznaky, léčba, komplikace, definice (http://www.zbynekmlcoch.cz/informace/medicina/nemoci-lecba/mnohocetny-myelom-priznaky-lecba-komplikace-definice)
- Pořad ČT: Medicína pro 21. století – Mnohočetný myelom (https://www.ceskatelevize.cz/porady/10175805663-medicina-pro-21-stoleti/209572231040005-mnohocetny-myelom/)
- Pořad ČT: Na pomoc životu – Mnohočetný myelom (https://www.ceskatelevize.cz/porady/10110975060-na-pomoc-zivotu-mnohocetny-myelom/20738254069/)

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