Plasma cell 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 deposits, 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.

Pathology

Macroscopy

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

Complications

- 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 - macroglossia.
- Myeloma kidney - clogging of tubules by protein chains - *'Bence Jones protein'*.

Life threatening conditions

- Failure of an organ affected by amyloidosis,
- bronchopneumonia - develops after vertebral fracture and spinal cord injury,
- bone marrow tumor generalization infection (same as 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.[1] It is almost non-existent in childhood and very rare in young people.

Clinical symptoms

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

The clinical picture is formed by a set of symptoms hidden under the acronym **CRAB**:

- **C**alcium – hypercalcemia, 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 osteoclasts activating factors, leading to bone osteolysis and pathological fractures that occur in up to 70% of patients.

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.

Examination methods

- X-ray - reveals pathological deposits (often the skull and spine are affected, but myeloma may be in any bone), as well as CT, MRI, PET,
- **detection of paraprotein in blood** – Plasma protein electrophoresis (see picture), serum paraprotein concentration is usually> 30 g/l,
- **detertion of free chain (FLC)** – the most sensitive method, it also detects non-secretory myeloma, when the determination of the paraprotein 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,

- detection of Bence Jones protein in the 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 beta $_2$ -microglobulin, hyperuricemia - their increased values have a negative prognostic significance.

Clinical stages and variants

Clinical stages

Classification according to Durie and Salmon[2]:

- **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** – araprotein IgG> 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),
  - **subclassification A** – serum creatinine ≤ 177 µmol/l,
  - **subclassification B** – serum creatinine > 177 µmol/l.

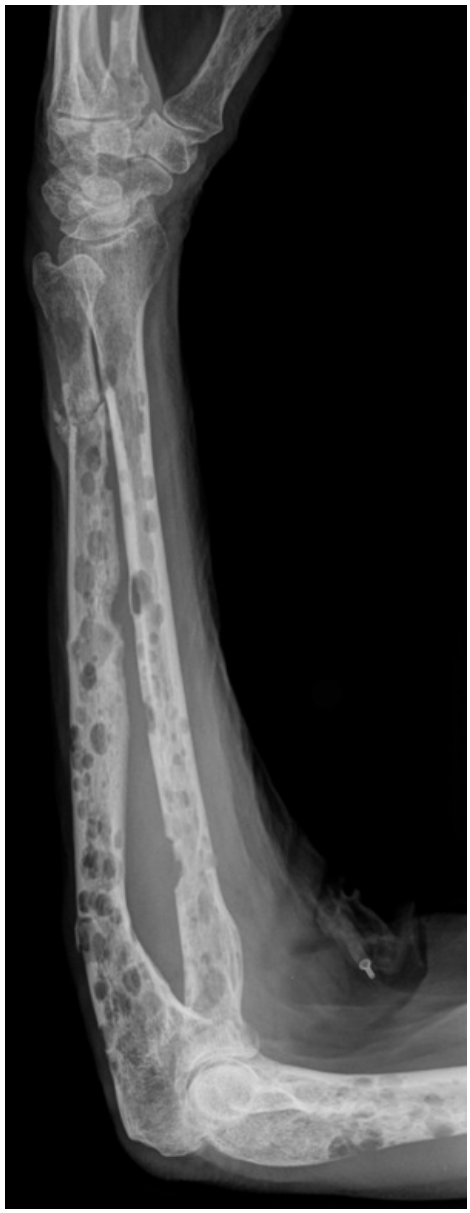
Clinical variants

Asymptomatic myeloma,

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

symptomatic myeloma,

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



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, since 2009 registered in the Czech Republic for primary therapy bortezomib [3]
- radiotherapy - for painful bone deposits,
- palliative treatment - in patients with severe comorbidities, melphalan and prednisone, or radiotherapy.

Links

Related articles

- Bence-Jones protein
- Plasma cells
- 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://ww

w.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/)

X-ray with multiple osteolytic lesions in the forearm.

References

1. ČEŠKA, Richard – ŠTULC, Tomáš. *Interna.* 2. edition. 2015. 909 pp. ISBN 978-80-7387-895-5.
2. ČEŠKA, Richard. *Interna.* 1. edition. Triton, 2010. 855 pp. pp. 710. ISBN 978-80-7387-423-0.
3. https://www.myeloma.cz/index.php?pg = multiple-myeloma-treatment-bortezomib-velcade

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