Plasmacytoma

Plasmacytoma is a malignant tumor of **plasma cells**. It usually affects the bone marrow, where it causes bone damage such as pathological fractures. If there is a presence of more foci, we call it **multiple myeloma** (Kahler's disease). Myeloma cells produce **paraprotein**. Rarely, non-secretory myelomas may occur, in which, no paraprotein can be detected in serum.

Pathology

Macroscopy

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

Complications

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

Life-threatening issues

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

Clinical description

Epidemiology

The disease is more prevalent in men than in women (1.5: 1). The risk of developing it rises with with age (after 50) with the median age being 70 years. The incidence in the Czech Republic is 3-4: 100,000 inhabitants / year. The etiology of the disease is unknown. It occurs hardly ever in childhood and in young adults.[1]

Electrophoresis in monoclonal gammopathy

Clinical picture

The initial period may be asymptomatic (indolent lymphomas), but **pain** in the back and ribs is present in over 70% of patients. Pain affects physical activity.

The clinical picture consists of a set of symptoms, described by the mnemonic **CRAB**:

- **C**alcium – hypercalcemia, which is associated with bone degeneration. Hypercalcemia also causes depression, lethargy, and fatigue.
- **R**enal failure – renal failure due to tubulopathy, an uncommon symptom of 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 disorder - very common, tumor cells in the bone marrow produce osteoclasts activating growth factors, which leads to osteolysis of the bones and the formation of pathological fractures, which occur in up to 70% of patients.

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

Diagnostic methods

- X-RAY - reveals pathological lesions (with often affected skull and spine, but myeloma can basically occur in any bone), as well as CT, MRI, PET,
- **determination of paraprotein in blood** – electrophoresis of plasma proteins, concentration of paraprotein in serum is usually> 30 g / l,
- **The immunoglobulin free light chain (FLC)** – the most sensitive method, it also detects non-secretory myeloma, in which the paraprotein determination would be negative, the normal ratio of kappa-lambda light chains is 2:1, in the case of myeloma it could for example be, 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 bone marrow, as well as immunohistochemical and cytogenetic examination (occurrence of deletions and trisomies has a prognostic significance),
- cytogenetic examination - numeric and structural changes in chromosomes, important for prognosis,
- laboratory examination,
  - normocytic normochromic anemia, mild leukopenia and thrombocytopenia
  - increased sedimentation,
  - hypercalcemia,
  - renal functions - increased urea and creatinine,
  - examination of LD, thymidine kinase and beta$_2$-microglobulin, hyperuricemia - their increased values indicate a poor prognosis.

Clinical stages and variants

Clinical stages

The Salmon-Durie classification of MM [2]:

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

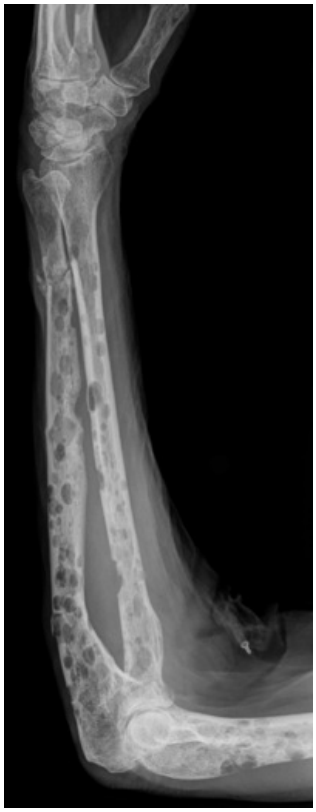
Clinical variants

Asymptomatic myeloma,

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

symptomatic myeloma,

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



Multiple osteolytic foci of myeloma

Therapy

- If myeloma happens to be asymptomatic, it does not have to be treated, only monitored
- if symptomatic, it is treated:
  - younger patients are indicated for autologous hematopoietic stem cell transplantation (aHSCT),
  - elderly patients are indicated for chemotherapy,
- chemotherapy – cyclophosphamide and dexamethasone (or thalidomide) are standardly used, bortezomib has been registered in the Czech Republic in 2009 for primary treatments,[3]
- radiotherapy – used for painful bone deposits,
- palliative care – in patients with severe comorbidities, melphalan and prednisone, or radiotherapy.

Links

Related articles

- Amyloidosis • Multiple myeloma

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/)

References

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

Sources

- STŘÍTESKÝ, Jan. *Patologie.* 1. edition. 2001. ISBN 80-86297-06-3.
- ČEŠKA, Richard, et al. *Interna.* 1. edition. Praha : Triton, 2010. 855 pp. pp. 708-711. ISBN 978-80-7387-423-0.