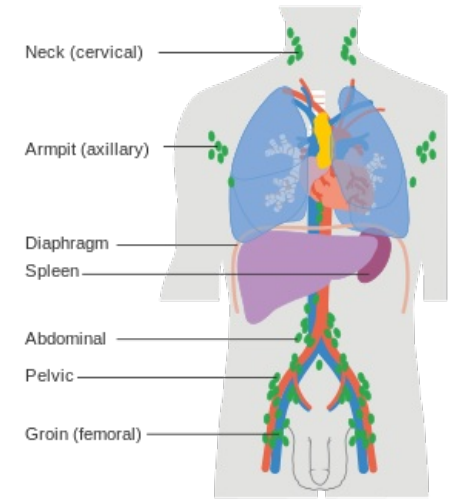
Malignant lymphoma

Malignant lymphoma = tumor of lymphatic tissue (lymphatic nodes + extranodulary lymphatic tissue).

- ***Hodgkin lymphoma***.
- ***Non-Hodgkin lymphoma*** (NHL).

1. B-lymphoma (80 %, usually more severe).
2. T-lymphoma.
3. NK-lymphoma.[1]

Units defined by their morphology + immunophenotypic features (CD Ag), cytogenetic traits, some even molecular-biological traits ([gene] translocations + protein production).[1]



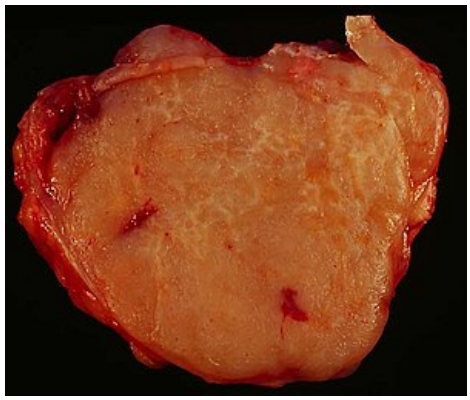
Lymphatic nodes with lymphoma.

Most important lymphomas

- diffuse large B-cell lymphoma (30 %),
- follicular lymphoma (22 %),
- MALT-lymphoma (8 %),
- chronic B-cell leukemia/lymphocytic lymphoma(7 %),
- Mantle cell lymphoma (6 %).

All malignant lymphomas can present so-called **B symptoms**: [1]

- weight loss (10% / half a year),
- subfebrile / febrile,
- night sweats.


Malignant lymphoma

MALIGNANT LYMPHOMAS IN CHILDREN

- 15% of malignant tumors in children and adolescents - the 3rd most common group of malignancies
- incidence increases with age:
  - NHL more common in children under 10 years of age
  - HL 2x more often than NHL in adolescents older than 15 years

Non-Hodgkin lymphomas

- a heterogeneous group of tumors of the lymphatic system
- in children, mainly tumors with a high degree of malignancy
- more often in boys

Etiology and pathogenesis

- the formation of lymphoma as a result of genetic aberrations affecting lymphocyte proliferation, differentiation and apoptosis
- higher incidence in children with primary and secondary immunodeficiencies
- Burkitt's lymphoma - associated with EBV infection, endemic in equatorial Africa
  - sporadic Burkitt's lymphoma - children in Europe and America, unrelated to EBV infection, has different clinical manifestations

Clinical picture

T-cell lymphoblastic lymphoma

- dyspnea due to mediastinal tumor
- taking the orthopneic position
- Superior vena cava obstruction - symptoms of superior vena cava syndrome
- the abdominal cavity is also often attacked - hepatosplenomegaly and kidney infiltration

Lymphoblastic lymphoma from B-precursors

- mostly localized disease on the skin, scalp, bone and peripheral lymph nodes

Sporadic Burkitt lymphoma

- involvement of the abdominal cavity
  - ileocecal intussusception in 1/4 of children
    - pain in P lower quadrant
    - dif.dg. appendicitis in these patients difficult
    - complete resection and end-to-end anastomosis
    - massive infiltration of the mesentery, retroperitoneum, peritoneum, kidneys, ovaries

Diff. large cell lymphoma

- affects peripheral lymph nodes, mediastinum, kidneys, pericardium and lungs
- manifestation of superior vena cava syndrome

Anaplastic large cell lymphoma

- fever, weight loss
- involvement of peripheral lymph nodes, mediastinum
- skin, soft tissue and bone involvement

Diagnostics

- cytomorfologie, histomorfologie, immunofenotypizace
- biopsy of nodes or extranodal areas, pleural or peritoneal effusion puncture, bone marrow aspiration
- respiratory patient with superior vena cava syndrome - !! very risky for an invasive diagnostic procedure - postponing the procedure, 24-48 hours of corticosteroid therapy, possibly in combination with cyclophosphamide
- determining the extent of the disease - CT of the chest and abdomen, PET, examination of the cerebrospinal fluid to rule out meningeal infiltration

Differential diagnosis

- Hodgkin's lymphoma
- post-transplant lymphoproliferative disease
- autoimmune lymphoproliferative disease

Therapy

- children with lymphoblastic lymphomas - application of ALL therapy protocols
- Burkitt's lymphoma, B-ALL, DLBCL - short and repeated blocks of intensive therapy (cyclophosphamide, MTX, vincristine)
- complications of therapy:
  - urate nephropathy - trp. rasburicase (recombinant urate oxidase)
  - acute infection

Hodgkin lymphoma

- the incidence is the same in both sexes
- most patients have high titers of EBV antibodies - activation of EBV infection may precede the development of HL
- division of HL:
  - classic - a tumor of lymphatic tissue from Hodgkin cells (mononuclear) and Reed-Sternberg multinuclear cells; 4 subtypes
  - HL with lymphocytic proliferation - from B-lymphocytes, with nodular proliferation of isolated large tumor cells

Clinical picture

- painless swelling of the supraclavicular and cervical nodes
- involvement of lymph nodes in the mediastinum
- systemic manifestations
  - fatigue, loss of appetite
  - B-symptoms: fever, night sweats, weight loss of more than 10% in the 6 months before diagnosis
- itchy skin (symptom of advanced disease)

Diagnostics

- lymph node biopsy + histology
- CT of the chest, CT/MR of the abdomen, PET

- staging: Ann Arbor classification

Therapy

- combined CHTR and RTR of affected areas

- cytostatics used: cyclophosphamide, vincristine, corticosteroids, anthracyclines

References

Related articles

- Hodgkin lymphoma
- Non-Hodgkin lymphoma
- CNS lymphoma
- Skin lymphoma

References

- DÍTĚ, P., et al. *Vnitřní lékařství.* 2. edition. Praha : Galén, 2007. ISBN 978-80-7262-496-6.

1. DÍTĚ, P.. *Vnitřní lékařství.* 2. edition. Praha : Galén, 2007. ISBN 978-80-7262-496-6.