Cutaneous T-lymphomas

Primary cutaneous T-lymphomas include those that affect only the skin at the time of diagnosis

Symptomatology

We can observe non-specific and specific changes . Non-specific changes include eczema and pruritus. Specific changes occur in about 17% of patients with malignant lymphoma. They are formed on the basis of infiltrations by tumor cells. The most common manifestations are infiltrated deposits anywhere on the body or solitary red-purple bumps in the forehead.

Primary cutaneous T-lymphoma with non-aggressive clinical behavior

Mycosis fungoides

thumb|right|Mycosis fungoides kolene thumb|right|Mycosis fungoides thumb|right| Mycosis fungoides – červen 1902 thumb|right|Mycosis fungoides – říjen 1902 **Mycosis fungoides** is a cutaneous epidermoid T-lymphoma characterized by the proliferation of small or medium-sized lymphocytes. It primarily affects the skin, but may gradually develop into squamous deposits with gradual tumor formation, affecting the lymph nodes and internal organs.

Symptomatology

Development takes years (without treatment 10-15 years). Gradual development in stages. .

1. **Premycotic stage**: in the form of so-called *parapsoriasis en plaques* ((formation of peeling surfaces on the trunk) or in the *poikilodermic* form (telangiectasia and depigmentation).

1. **Infiltration stage**: :formation of tumors with disintegrating ulcers.
2. **Tumor stage**:formed tumors , some with a sunken hat-shaped surface (hence the name). Internal organs are also affected at this stage.

Manifestations are asymptomatic, but when the nerves are affected, **itching occurs that does not respond to antihistamines.**

Clinical variants

- Folliculotropic MF

- Pagetoid reticulosis
- Granulomatous sagging skin
- Sézaryho syndrom

Template:Heslo Sézaryho syndrom|náhled **Sezary's syndrome** is a leukemic form of Mycosis fungoides with concomitant erythroderma, generalized lymphadenopathy and hepatosplenomegaly. Clinically, there is red-purple discoloration of the skin, peeling of the entire skin, loss of nails, alopecia, and persistent itching

Histopathological diagnosis

Epidermotropysm and Pautrera microabscesses in the epidermis.

Therapy

Photochemotherapy (PUVA) for treatment of skin only.. Application of carmustine, corticoids and retinoids. Interferon α is used in case of major symptoms.

Course of disease and prognosis

Chronic course of disease for decades. Prognosis based on the progression of disease .

Primary cutaneous anaplastic large cell lymphoma

Tumor from cells expressing the CD30 antigen.

Symptomatology

Occurs as individual bumps on the limbs or torso in adults and children these are rare. About 10% of patients have affected lymphatic nodes , which, however, does not worsen the prognosis of the disease.

Histopathological diagnosis

CD30 + tumor cells predominate. More than 75% of CD30 + cells are required to confirm the disease.

Therapy

Radiotherapy, based on the development of the diesease polyradiotherapy might be requied.

Course and prognosis

Five-year survival of about 90%.

Lymphomatoid papulomatosis

Lymphomatoid papulomatosis is a chronic disease. It often resolves and relapses spontaneously..

Symptomatology

Presence of lesions that are papulomatous or even necrotic and nodular in various stages of development on the trunk, buttocks and limbs.

Histopathological diagnosis

1. Type A: contains Reed-Sternberg cells.

1. Type B: identical to Mycosis fungoides (Epidermotropism and Pautrer's microabscesses in the epidermis).
2. Type C: large atypical CD30 + cells.

Therapy

There is no effective treatment. PUVA is used for suppression, after discontinuation of treatment, recurrence.

Course and prognosis

Chronic course, mostly benign .Healing by scarring and depigmentation.

Links

Related Articles

- Lymphoma
- CNS lymphomas
- Hodgkin's lymphoma

Used literature

- STORK, George. *Dermatovenerology.* 1. edition. Prague : Galén, Karolinum, 2008. pp. 502. ISBN 978-80-7262-371-6.