

Cutaneous T-Cell Lymphomas (CTCLs)

Types, Treatment and Research

Most people have never heard of cutaneous T-cell lymphomas (CTCLs). Compared to lung and breast cancers, CTCLs are virtually unknown to the public. But to those who live with these diseases, they are all too familiar.

CTCLs are “visible” diseases with rashes that might cover 80 percent of a person’s body. The itching and dry skin can be extremely hard to control, making people feel uncomfortable. Because CTCLs are rare and sometimes misdiagnosed, patients often go years with little to no relief.

What are CTCLs?

CTCLs are a family of non-Hodgkin lymphomas. The most common forms of CTCLs are mycosis fungoides (MF) and Sézary syndrome (SS). In CTCLs, a white blood cell, also known as a T-cell, becomes activated and makes copies of similar cells (clones) which invade the skin in MF and the blood in SS. They appear as skin lesions.

Mycosis Fungoides

MF, which is the most common form of CTCL, affects about 1,500 people in the United States per year. The number of new cases has increased 300 percent over the last three decades. MF often remains confined to the skin for many years, acting more like a chronic skin disease, such as eczema or psoriasis, than a lymphoma. Doctors often miss the correct diagnosis, even after sending skin samples for testing in a lab. MF can look like:

- Dry, itchy skin.
- Pink, scaly patches of eczema, a condition that causes itching and inflamed skin.
- Flat, raised patches of psoriasis, a chronic condition that causes scaly patches of skin.
- A fungal infection, ringworm or light or dark-colored areas of skin (especially in skin of color).

When MF is diagnosed early and covers less than 10 percent of the skin (Stage IA), patients have an excellent prognosis of healing. Early MF can be treated with creams, chemotherapy on the skin, sunlight, electron beam radiation or PUVA, which is a combination of Psoralen (P) and ultraviolet A (UVA) light used to treat many different skin conditions. Psoralen is a drug taken by mouth that makes the skin disease more responsive to ultraviolet light. Because UVA light is low in energy, it is not very beneficial when used alone. MF can also be treated with ultraviolet B (UVB) light.

When MF is not diagnosed properly or treated well, it can get worse, forming tumors and ulcers that can become infected. The disease can also involve the lymph nodes, blood or bone marrow. Once this happens, treatment is more aggressive, requiring chemotherapy or experimental methods.

Sézary Syndrome

SS is named after the French skin doctor who discovered the condition. He found patients with this skin rash also have cancer cells in the blood. Patients with SS have dry, itchy, red skin over 80 percent of their bodies. The itching may be severe and hard to manage. They may also have thick, yellow hands and feet (keratoderma), a fungal infection or red skin.

Patients with SS often carry staphylococcus, a type of bacteria, on their skin. This makes the skin more red and itchy. While patients are treated with certain medications, such as retinoids and interferon, or light therapy (photopheresis), we can often improve the skin and symptoms by using:

- Antibiotic soap.
- Acetic acid to keep the pH of the skin low.
- Moisturizers and steroid creams.

Patients who do not find relief may talk to their doctor about taking part in a research study using antibodies or small molecules. Some of these treatments are still experimental and are being studied.

The Other Types of CTCLs

CD30+ lymphoproliferative disorders are the second most common group of CTCLs. These disorders are made up of large abnormal T-cells, which express a protein called CD30+.

One form is lymphomatoid papulosus (LYP), which appears as a small pink or red bump that may itch or tear open but goes away on its own within several months. When larger lesions form and do not get better, the diagnosis is anaplastic large cell lymphoma (ALCL) and radiation treatment may be needed. CD30+ expression can also be found in tumors of MF, Hodgkin lymphoma and with a viral infection like shingles. There is a new antibody for CD30+.

Some rare forms of CTCL invade the body's fat. These include panniculitic T-cell lymphoma, CD8+ T-cell lymphoma and gamma/delta T-cell lymphoma. Other forms, such as peripheral T-cell lymphoma, may need more aggressive treatment than MF.

B-Cell Lymphomas

B-cell lymphomas can also affect the skin. They appear as pink patches, papules or nodules. Follicular B-cell lymphoma most often appears on the head and neck with glassy pink papules or nodules. Marginal zone lymphomas, such as mucosa-associated lymphoid tissue (MALT), usually appear as small pink patches on the arms or trunk. Both have a good prognosis and can be treated locally.

Diffuse large B-cell lymphoma, especially the leg type, can spread and is treated with chemotherapy.

What are the Symptoms of CTCLs?

Symptoms usually occur in four stages. The diseases can start at any stage and do not necessarily go through the stages in any order.

T Staging:

- T1 – Patches/plaques on less than 10 percent of skin.
- T2 – Patches/plaques on more than 10 percent of skin.
- T3 – Tumors.
- T4 – Erythroderma.

Patch Stage:

- Flat, pink, red, brown or white lesions.
- Patches may persist or resolve on their own.
- Vary in size, shape and scale.
- Resemble eczema or ring worm.
- Often appear in non-sun-exposed areas.

Plaque Stage:

- Lesions are raised and may have scaling.
- Deeper skin layers involved.
- Patches may be round, oval or ring shaped.
- Color is generally pink, red, brown or grey.
- May be confused with psoriasis.

Tumor Stage:

- One or more nodules with or without skin tearing.
- Pink to red or brown. If purple, more aggressive cancer.
- Often first appears on the head and neck.
- Tumors with broken skin are often infected.

Generalized Erythroderma Stage:

- More than 80 percent of skin is pink to red.
- Scaling can be fine to coarse.
- Palms and soles may be thick and crack, making it difficult to walk.
- Fungus infection present.
- Rule out other causes of erythroderma.

Sézary Syndrome:

- More than 80 percent of skin is pink to red.
- More than 1,000 cancerous Sézary cells in the blood.
- Severe itching, enlarged lymph nodes (adenopathy), thickening of the skin (keratoderma) and infections.