# Cutaneous Lymphoma

# FAST FACTS

Cutaneous Lymphoma Foundation

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## What is Cutaneous Lymphoma?

Cutaneous lymphomas are types of non-Hodgkin's lymphomas (NHL) that originate in the lymphocytes (white blood cells). Unlike most other types of NHL, which develop in lymph nodes, cutaneous lymphomas develop primarily in the skin. They can be classified into cutaneous B-cell lymphomas and cutaneous T-cell lymphomas, depending on which type of lymphocyte they originate from.

There are many forms of cutaneous lymphoma with a range of symptoms, outcomes, and treatment considerations. The most common types are cutaneous T-cell lymphomas, which make up 75–80% of all cutaneous lymphomas. Mycosis fungoides and Sézary syndrome are the most frequently diagnosed subtypes of cutaneous T-cell lymphoma. CD30+ lymphoproliferative disorders, including lymphomatoid papulosis and anaplastic large cell lymphoma, account for 10–15% of all cutaneous T-cell lymphomas. Cutaneous B-cell lymphomas originate in skin-based B-cells. Cutaneous B-cell lymphomas make up 20–25% of all cutaneous lymphomas and include a variety of subtypes; primary cutaneous follicle center lymphoma is the most common form.

## What is Mycosis Fungoides?

Mycosis fungoides is the most common cutaneous T-cell lymphoma accounting for approximately half of all cutaneous T-cell lymphomas. It is typically slow growing, and it most often remains localized to the skin. Mycosis fungoides can be difficult to diagnose because it can be mistaken for other skin conditions. The disease appears differently in each patient, with skin symptoms or lesions that can appear as *patches*, *plaques*, or *tumors*. It is possible to have more than one type of lesion.

## What is Sézary Syndrome?

Sézary syndrome is a less common but more aggressive type of cutaneous T-cell lymphoma. Sézary syndrome affects the skin, blood, and lymph nodes. Sézary syndrome makes up approximately 15% of patients with cutaneous T-cell lymphoma. Sézary syndrome appears as widespread redness and scaling of the skin (erythroderma) with enlarged lymph nodes. Patients may also experience severe itching or changes in the nails, hair, or eyelids. It is a chronic and systemic disease.

## Who gets Cutaneous Lymphoma? How common is it?

As a group, cutaneous lymphomas are a relatively rare family of diseases that occurs more often in men than women. They are more common in people older than 50. There are an estimated 30,000 people in the United States and 3,000 people in Canada living with cutaneous lymphoma. Approximately 1,500–2,000 new cases are diagnosed each year across North America. There is general consensus that these numbers are low because they do not currently account for misdiagnosed or undiagnosed patients. It is difficult to diagnose the disease in its early stages, and there is not an accurate reporting system. It is important to know that cutaneous lymphomas are not contagious and cannot be passed from one person to another.

## What does Cutaneous Lymphoma look like?

Cutaneous lymphoma presents differently in each patient, with variation in the degree of severity. *Patches* usually resemble a rash. *Plaques* are thicker, raised lesions that are usually itchy and are often mistaken for eczema, psoriasis, or dermatitis. *Tumors* are raised bumps that may or may not ulcerate. Patients can have any or all of these symptoms, but some people can have cutaneous lymphoma for many years with only one of these characteristic signs. Patients with Sézary syndrome often have extensive thin, red, itchy rashes (erythroderma) covering over 80% of their body. Patients may have enlarged lymph nodes or experience changes such as thickening on the palms of their hands and soles of their feet, brittle fingernails, hair thinning or hair loss, or drooping eyelids.

## How does Cutaneous Lymphoma progress?

The course of this disease is not predictable, especially because it can be different for each individual patient. Most patients do not progress at all, while others can progress at varying rates. Most patients will have only skin symptoms and no serious complications. In approximately 10% of patients, the disease will progress to the lymph nodes or internal organs with serious complications. Many patients live normal lives while they treat their disease, and some are able to remain in remission for long periods of time.

## How is Cutaneous Lymphoma diagnosed, and what kinds of tests should a patient expect?

Cutaneous lymphoma is difficult to diagnose, especially in the early stages, because the symptoms and skin biopsy findings can be similar to those of other skin conditions. A medical history, physical exam, and skin biopsy are essential for diagnosis. A physician will examine the lymph nodes and order various blood tests and a skin biopsy. In the presence of more advanced disease, more testing may be done to determine if the cancer has spread. Other screening tests, which can include a chest x-ray, a CAT scan, a PET scan, or an MRI scan may be done. Scans are usually not needed for patients with the earliest stages of the disease. A bone marrow biopsy may be done to verify complete staging of the disease, but is more likely to be needed with cutaneous B-cell lymphomas than cutaneous T-cell lymphomas.

## What causes Cutaneous Lymphoma?

Although there is continuing research, currently no single factor is known to cause cutaneous lymphomas. They are acquired diseases with no clear genetic or hereditary link, and they are not contagious. Despite a number of anecdotal reports, there does not appear to be a clear and defined link between cutaneous lymphoma and chemical exposure, pesticides, radiation, allergies, the environment, or occupations. Exposure to Agent Orange may be a risk factor for developing cutaneous T-cell lymphoma (mycosis fungoides variant), but no direct causal relationship has been shown.

## Is there a cure for Cutaneous Lymphoma?

Cutaneous lymphoma can usually be managed for many years. The 70–80% of patients diagnosed with an early stage of cutaneous lymphoma live a normal life expectancy. In recent years, greater research efforts, greater treatment options, and more collaboration among physicians have contributed to better care and outcomes for patients.

## What are the treatment options for Cutaneous Lymphoma?

Treatment of cutaneous lymphoma depends on the stage and symptoms of the disease. A patient's age, health status, and goals for treatment are also factors in treatment selection. For some patients with slow-growing or static disease, "watchful waiting" may be the best clinical course. For patients with fast-growing or advanced disease, treatment is usually necessary.

Treatments are categorized as *skin-directed* or *systemic* (directed at the entire body). Skin-directed therapies include topical steroids, phototherapy, topical chemotherapy, and topical retinoids. Radiation therapies are focused on skin symptoms/lesions and are generally used for early-stage disease. They can be used alone or in combination, and selection is based on a patient's specific symptoms. Systemic therapies reach the bloodstream, and are distributed throughout the entire body, including the skin. Systemic therapies are generally used for advanced disease, and there are many systemic therapy agents that can be used alone or in combination. There are new agents approved for cutaneous lymphomas, including new biologic agents in development and in clinical trials. Additional clinical trials are being conducted to determine the most effective treatments. Combination chemotherapy regimens are generally reserved for patients who have not responded well to several single-agent therapies.

## Should patients consider a clinical trial?

Clinical trials are crucial in identifying effective drugs and finding the best treatments for patients with cutaneous lymphoma. Physicians may recommend clinical trials because standard treatments may not be effective. Sugar pills (also called placebos) are rarely used in cancer clinical trials.

## Questions patients should ask their doctor about Cutaneous Lymphoma:

- What type of cutaneous lymphoma do I have?
- What stage is my disease?
- What treatment do you recommend? Why?
- What are the side effects related to the treatment you recommend? Are they reversible after the treatment ends?
- Am I a candidate for and would I benefit from participation in a clinical trial?
- How often will I be receiving treatment?
- Do I have to receive my treatment at the specialty clinic, or can I receive my treatment from a clinical practice closer to my home?
- What are the costs of this treatment? Where can I get help with financial concerns?

## Abbreviations

**ALCL** anaplastic large cell lymphoma

**CAT** computed axial tomography

**CBCL** cutaneous B-cell lymphoma

**CL** cutaneous lymphoma

**CTCL** cutaneous T-cell lymphoma

**LyP** lymphomatoid papulosis

**MF** mycosis fungoides

**MRI** magnetic resonance imaging

**NHL** non-Hodgkin's lymphoma

**PCFCL** primary cutaneous follicle center lymphoma

**PET** positron emission tomography

**SS** Sézary syndrome

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