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About Lymphoma of the Skin

Get an overview of lymphoma of the skin and the latest key statistics in the US.

Overview and Types

If you have been diagnosed with lymphoma of the skin or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- [What Is Lymphoma of the Skin?](#)
- [Types of Lymphoma of the Skin](#)

Research and Statistics

See the latest estimates for new cases of lymphoma of the skin in the US and what research is currently being done.

- [Key Statistics for Lymphoma of the Skin](#)
- [What's New in Skin Lymphoma Research?](#)

What Is Lymphoma of the Skin?

When a non-Hodgkin lymphoma starts only in the skin (not in other organs or tissues) it is called a **skin lymphoma** (or **cutaneous lymphoma**). A lymphoma that starts in lymph nodes or another part of the body and then spreads to the skin is not a skin lymphoma (because it didn't start there).

- [What is lymphoma?](#)
- [The lymph system and lymphoid tissue](#)

What is lymphoma?

Lymphoma is a cancer that starts in white blood cells called **lymphocytes**, which are part of the immune system. The main types of lymphomas are:

- **Hodgkin lymphoma** (also known as Hodgkin's lymphoma, Hodgkin disease, or Hodgkin's disease)
- **Non-Hodgkin lymphoma** (also known as non-Hodgkin's lymphoma, NHL, or sometimes just lymphoma), which includes all skin lymphomas

Lymphocytes are in the lymph nodes (small, bean-sized collections of immune cells throughout the body) and other lymphoid tissues (such as the spleen, bone marrow, and some other organs, including the skin). Lymphomas can start in any of these places.

[Hodgkin lymphoma](#)¹ and other types of [non-Hodgkin lymphoma](#)² are discussed separately.

[What Is Cancer?](#)³

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer cells. Learn more here.

Lymphatic System

[Anatomy Gallery: Lymphatic System](#)⁴

Explore our 3D interactive tour of the lymphatic system.

[Anatomy Gallery: Skin](#)⁵

Explore our 3D interactive tour of the skin system.

The lymph system and lymphoid tissue

To understand what lymphoma is, it helps to know something about the lymph system (also known as the lymphatic system). The lymph system is part of the immune system, which helps fight infections and some other diseases. The lymph system also helps fluids move around the body.

Lymphocytes

The lymph system is made up mainly of lymphocytes, a type of white blood cell. The main types of lymphocytes are:

- **B lymphocytes (B cells):** B cells normally help protect the body against germs (bacteria or viruses) by making proteins called antibodies. The antibodies attach to the germs, marking them for destruction by other parts of the immune system.
- **T lymphocytes (T cells):** There are several types of T cells, each with a special job. Some T cells destroy germs or abnormal cells in the body. Other T cells help boost or slow the activity of other immune system cells.

Some other types of cancer, such as lung cancer or colon cancer, can spread to lymph tissue such as the lymph nodes. But cancers that start in these places and then spread to the lymph tissue are not lymphomas.

Hyperlinks

1. www.cancer.org/cancer/types/hodgkin-lymphoma.html
2. www.cancer.org/cancer/types/non-hodgkin-lymphoma.html
3. www.cancer.org/cancer/understanding-cancer/what-is-cancer.html
4. www.cancer.org/cancer/understanding-cancer/anatomy-gallery/lymphatic-system.html
5. www.cancer.org/cancer/understanding-cancer/anatomy-gallery/skin.html
6. www.cancer.org/cancer/types/skin-lymphoma/detection-diagnosis-staging/how-diagnosed.html
7. www.cancer.org/cancer/types/skin-lymphoma/treating.html

References

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Querfeld C, Rosen ST. Chapter 107: Cutaneous T-cell lymphoma and cutaneous B-cell lymphoma. In: Niederhuber JE, Armitage JO, Dorshow JH, Kastan MB, Tepper JE, eds. Abeloff's Clinical Oncology. 5th ed. Philadelphia, Pa. Elsevier: 2014.

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Types of Lymphoma of the Skin

There are many types of skin lymphomas. Classifying them can be confusing (even for

many doctors) because many of them are not very common.

The main system used to classify skin lymphoma is from the World Health Organization (WHO), which was last updated in 2016. It is based mainly on:

- Whether the lymphoma starts in T lymphocytes (T cells) or B lymphocytes (B cells)
 - How the lymphoma looks under the microscope
 - Whether certain proteins are on the lymphoma cells (based on lab tests)
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- [T-cell skin lymphomas](#)
 - [B-cell skin lymphomas](#)

T-cell skin lymphomas

Most skin lymphomas are T-cell lymphomas. These are sometimes called **cutaneous T-cell lymphomas (CTCLs)**.

Mycosis fungoides: Nearly half of all skin lymphomas are mycosis fungoides (MF). MF can occur in people of any age, but most who get it are in their 50s or 60s. Men are almost twice as likely as women to develop this lymphoma.

The first sign of this disease is one or more patchy, scaly, red lesions (abnormal areas) on the skin. MF lesions can be very itchy. Often these lesions are the only symptom of MF. But in some people the disease can progress to more solid, raised tumors on the skin (called **plaques**). Because MF can be confused with other skin problems, it can be hard to diagnose at first. Several [biopsies](#)¹ of the lesions might be needed before the diagnosis is confirmed.

Over time, MF can spread across the skin or invade lymph nodes and organs like the liver. In many people this disease grows slowly, but it can sometimes grow more quickly, especially in older people. Some people with MF go on to develop Sezary syndrome.

Rare variants of MF include **folliculotropic MF**, **pagetoid reticulosis**, and **granulomatous slack skin**.

Sezary syndrome (SS): This is often thought of as an advanced form of mycosis fungoides, but these are actually different diseases. In SS, most or all of the skin is affected, instead of just patches of skin. People with SS typically have a very itchy, scaly red rash that can look like a severe sunburn. This is called **generalized**

erythroderma. The skin is often thickened. Lymphoma cells, called **Sezary cells**, can be found in the blood (as well as in the lymph nodes).

Whereas MF is usually slow growing, SS tends to grow and spread faster, and is harder to treat. People with SS also often have further weakened immune systems, which increases their risk of serious infections.

Adult T cell leukemia-lymphoma (ATLL): This rare type of T-cell lymphoma is more

Extranodal NK/T-cell lymphoma, nasal type: This rare type of lymphoma can start in T-cells or in other lymphocytes known as natural killer (NK) cells. It typically starts in the nose or sinuses, but sometimes it can start in the skin. This lymphoma has been linked to infection with the Epstein-Barr virus (EBV), and is more common in Asia and Central and South America. It tends to grow quickly.

Primary cutaneous peripheral T-cell lymphoma, rare subtypes: This is a group of rare skin lymphomas that don't fit into any of the above categories. There are several types.

- **Primary cutaneous gamma/delta T-cell lymphoma** develops as thickened plaques (raised lesions) or actual tumors, mainly on skin of the arms and legs, but sometimes in the intestines or lining of the nose. This type of lymphoma tends to grow and spread quickly.
- **Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma** develops as widespread patches, nodules and tumors that often break open in the middle. This type of lymphoma can sometimes look like mycosis fungoides, but a [biopsy](#)² can tell them apart. This lymphoma tends to grow and spread quickly.
- **Primary cutaneous acral CD8+ T-cell lymphoma** is very rare, and typically starts as a nodule on the ear, although it can also start on other parts of the body, such as the nose, hand, or foot. It tends to grow slowly and can often be cured with treatment.
- **Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder** often starts as a single area of thickening of the skin or a tumor on the head, neck, or upper body. This disease tends to grow slowly and can often be cured with treatment.

B-cell skin lymphomas

These lymphomas are sometimes referred to as **cutaneous B-cell lymphomas (CBCLs)**.

Primary cutaneous marginal zone B-cell lymphoma: This is a very slow-growing lymphoma that is usually curable. In Europe (but not in the United States), it is sometimes linked to an infection with *Borrelia*, the germ that causes Lyme disease.

This lymphoma can occur at any age, although it tends to occur more often in older

adults. It appears as skin lesions that are red to purplish large pimples, plaques (raised or lowered, flat lesions), or nodules (bumps) on the arms or upper body. There may be only a single lesion, but there can sometimes be a few.

Primary cutaneous follicle center lymphoma: This is the most common B-cell lymphoma of the skin. It tends to grow slowly. The early lesions are groups of red pimples, nodules, or plaques that form on the scalp, forehead, or upper body. Less often they are found on the legs. Sometimes the pimples grow into nodules.

This type of lymphoma is typically found in middle-aged adults. It tends to respond well to radiation therapy, and most patients have an excellent outlook.

Primary cutaneous diffuse large B-cell lymphoma, leg type: This is a fast-growing lymphoma that begins as large nodules, mainly on the lower legs. It occurs most often in older people, and is more common in women than men. In some people, this lymphoma spreads to lymph nodes and internal organs, causing serious problems.

These lymphomas often require more intensive treatment. The outlook is better if there is only one lesion at the time of diagnosis.

Hyperlinks

1. www.cancer.org/cancer/types/skin-lymphoma/detection-diagnosis-staging/how-diagnosed.html
2. www.cancer.org/cancer/types/skin-lymphoma/detection-diagnosis-staging/how-diagnosed.html

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Jacobsen E, Freedman AS, Willemze R. Primary cutaneous large B cell lymphoma, leg type. UpToDate. Accessed at www.uptodate.com/contents/primary-cutaneous-large-b-cell-lymphoma-leg-type on February 8, 2018.

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Key Statistics for Lymphoma of the Skin

What's New in Skin Lymphoma Research?

1. www.cancer.org/cancer/types/skin-lymphoma/causes-risks-prevention/what-causes.html
 2. www.cancer.org/cancer/types/skin-lymphoma/detection-diagnosis-staging/how-diagnosed.html
 3. www.cancer.org/cancer/types/skin-lymphoma/treating.html
 4. www.cancer.org/cancer/types/skin-lymphoma/detection-diagnosis-staging/how-diagnosed.html
 5. www.cancer.org/cancer/managing-cancer/treatment-types/radiation/photodynamic-therapy.html
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