



The Cutaneous Lymphoma Foundation is an independent, non-profit patient advocacy organization dedicated to supporting every person with cutaneous lymphoma by promoting awareness and education, advancing patient care, and facilitating research.

CUTANEOUS LYMPHOMA FOUNDATION

Fast Facts: Sézary Syndrome

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What is Lymphoma?

Lymphoma is a cancer of the white blood cells, namely lymphocytes, which constitute the lymphatic system. The two main types of lymphoma are Hodgkin lymphoma and non-Hodgkin lymphoma (NHL). Lymphoma is the most common blood cancer and the third most common cancer of childhood. Lymphoma occurs when lymphocytes grow abnormally.

The body has two main types of lymphocytes: B lymphocytes, or B-cells, and T lymphocytes, or T-cells. B-cells play an important role in making antibodies to fight bacterial infections, while T-cells play a role in fighting viruses and organ rejection in transplant patients. Although both cell types can develop into lymphomas, B-cell lymphomas are more common, comprising nearly 85 percent of all non-Hodgkin lymphomas. Like normal lymphocytes, those that become malignant can grow in any part of the body, including the lymph nodes, spleen, bone marrow, blood or other organs, including the liver, lung and kidney. Lymphoma has been shown to grow in virtually every organ of the body.

What is Non-Hodgkin Lymphoma?

Together, Hodgkin lymphoma and non-Hodg-

kin lymphoma comprise more than 67 subtypes of lymphoma. There are six primary types of Hodgkin lymphoma and as many as 61 types of non-Hodgkin lymphoma.

Nearly all non-Hodgkin lymphoma cases occur in adults. The average age of diagnosis is usually in the early- to mid-60s, but lymphomas can develop at any age. While scientists do not know the exact causes of non-Hodgkin lymphoma, they do know that it is not caused by injury or by coming in contact with someone with the disease. Most people diagnosed with NHL have no known risk factors, although increasingly many scientists believe infections may contribute to the development of certain types of non-Hodgkin lymphoma. According to the American Cancer Society, there were approximately 66,120 new cases of NHL diagnosed in 2008. The disease is more common in men than in women.

What Is T-Cell Lymphoma?

T-cell lymphoma is a disease in which T lymphocyte cells become cancerous. These lymphomas account for between ten percent and 15 percent of all cases of non-Hodgkin lymphoma in the United States (approximately 5,000 to 6,000 cases) a year, although some forms of T-cell lymphoma

are more common in Asia and in other parts of the world. There are many different types of T-cell lymphoma and some are extremely rare, occurring in only a few patients per year throughout the world. Most T-cell lymphomas can be classified into two broad categories: aggressive (fast-growing) or indolent (slow-growing).

One of the most common forms of T-cell lymphoma is cutaneous, or skin, lymphoma, because it starts in the lymphocytes in the skin. Cutaneous lymphoma actually describes many different disorders with various signs and symptoms, outcomes and treatment considerations.

What Is Sézary Syndrome?

The two most common types of cutaneous T-cell lymphomas (CTCL) are mycosis fungoides, which is often indolent, appears as reddish skin patches and can progress over many years, and an advanced and leukemic form of mycosis fungoides called Sézary syndrome. Sézary syndrome is distinguished from mycosis fungoides by the presence of malignant lymphocytes in the blood and is characterized by extensive thin red, itchy rashes covering over 80 percent of the body. In some cases, thicker, red patches (or plaques) and tumors may also appear. In addition, these symptoms may be accompanied by changes in the nails, hair or eyelids or the presence of enlarged lymph nodes.

There are about 3,000 new cases of mycosis fungoides each year in the U.S. and approximately 15 percent of those are diagnosed as Sézary syndrome. Although this type of NHL can affect people of any age, Sézary syndrome usually occurs in adults ages 50 and over and is slightly

more common in men than women. There are no known risk factors for this type of cutaneous T-cell lymphoma.

How Sézary Syndrome Is Diagnosed and Staged

Many of the same procedures used to diagnose and stage other types of cutaneous T-cell lymphomas are used in Sézary syndrome, including a physical exam and history; blood tests to identify antigens, or markers, on the surface of the cells in the blood; a skin and/or lymph node biopsy (removal of a small piece of tissue) for examination under the microscope by a pathologist (a doctor who studies tissues and cells to identify diseases); and a series of imaging tests such as CT (computerized axial tomography), MRI (magnetic resonance imaging) and/or PET (positron emission tomography) scans to determine if the cancer has spread to lymph nodes or other organs. In addition to these diagnostic tests, occasionally a bone marrow biopsy may be necessary to verify complete staging.

Because mycosis fungoides and Sézary syndrome are such rare cancers, it is important to confirm a diagnosis by a dermatopathologist or a hematopathologist, a pathologist who is an expert in diagnosing lymphomas.

The following staging system is used to determine the extent of mycosis fungoides:

- Stage IA—Less than 10 percent of the skin is covered in red patches and/or plaques.
- Stage IB—Ten percent or more of the skin surface is covered in patches and/or plaques.

- Stage IIA—Any amount of the skin surface is covered with patches and/or plaques; lymph nodes are enlarged, but the cancer has not spread to them.
- Stage IIB—One or more tumors are found on the skin; lymph nodes may be enlarged, but the cancer has not spread to them.
- Stage III—Nearly all of the skin is reddened and may have patches, plaques or tumors; lymph nodes may be enlarged, but the cancer has not spread to them.
- Stage IVA—Most of the skin area is reddened and there is involvement of the blood with malignant cells or any amount of the skin surface is covered with patches, plaques or tumors; cancer has spread to the lymph nodes and the lymph nodes may be enlarged.
- Stage IVB—Most of the skin is reddened or any amount of the skin surface is covered with patches, plaques or tumors; cancer has spread to other organs; and lymph nodes may be enlarged whether cancer has spread to them or not.

How Sézary Syndrome Is Treated

There are many effective therapies available to treat Sézary syndrome. Because the disease is chronic and systemic (affecting the entire body), Sézary syndrome is usually not treated with skin-directed therapies alone. The specific treatment for individual patients is based on a variety of factors, including the patient's general health and stage of the disease.

There are several types of standard treatment for Sézary syndrome:

- Biologic, or immunotherapy, therapy is a treatment used to stimulate a patient's own immune system to fight the cancer
- Chemotherapy, a drug given either orally or through an infusion in a vein, to stop the growth of rapidly dividing cancer cells
- Extracorporeal photopheresis (ECP), a procedure used to expose the blood to ultraviolet light
- Histone deacetylase inhibitors, a class of drugs that cause a chemical change that stops tumor cells from dividing
- Phototherapy, for example, the drug psoralen and ultraviolet-A light radiation (PUVA) directed to the skin or skin-directed ultraviolet-B (UVB) or narrow band ultraviolet-B (NBUVB)
- Radiation therapy, which uses high-energy X-rays or other types of radiation to kill cancer cells or keep them from growing
- Retinoids, which are drugs related to vitamin A and can slow certain types of cancer cells

Some specific drugs include:

- Alemtuzumab (Campath), a monoclonal antibody
- Bexarotene (Targretin), a retinoid
- Denileukin diftitox (Ontak), a monoclonal antibody
- Gemcitabine (Gemzar), an antimetabolite chemotherapy
- Interferon alfa or interleukin-2, immune stimulants that bind to specific cell-surface receptors
- Liposomal doxorubicin (Doxil), a chemo

therapy that binds to DNA

- Methotrexate (Trexall), an antimetabolite chemotherapy, which blocks the metabolism of cells
- Vorinostat (Zolinza), a histone deacetylase inhibitor

Some common combination therapies include:

- Bexarotene (Targretin) and interferon alpha
- Bexarotene and phototherapy
- ECP (extracorporeal photopheresis) and bexarotene
- ECP and interferon alpha
- ECP, interferon alpha and bexarotene
- Phototherapy and interferon alpha

Some second-line chemotherapies for relapsed (the recurrence of disease) or refractory (disease that is resistant to treatment) patients include:

- Bexarotene (Targretin) and denileukin difitox (Ontak)
- Chlorambucil (Leukeran)
- Cyclophosphamide (Cytosan)
- Pentostatin (Nipent)

Treatments Under Investigation

There are several single agent therapies and drug combination regimens being studied in clinical trials in the treatment of mycosis fungoides and Sézary syndrome. Some of those treatments include:

- Bortezomib (Velcade)
- Enzastaurin
- PDX (Pralatrexate)
- PUVA therapy, plus interferon alfa

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs, prognostic strategies and determining optimal doses for lymphoma patients. Because mycosis fungoides and Sézary syndrome are such rare diseases, finding enough patients to enroll in clinical trials is often difficult. If you are interested in participating in a clinical trial, talk to your doctor about an appropriate trial for you. To learn more about clinical trials, visit the Cutaneous Lymphoma Foundation at www.clfoundation.org.

Are Complimentary and Alternative Therapies Safe and Effective?

Complementary and alternative medicines are nonstandard therapies that may help patients cope with their cancer and its treatment, but that should not be used in place of standard treatment. No alternative therapy has ever been proven effective against lymphoma. However, complementary therapies such as meditation, yoga, acupuncture, exercise, diet and relaxation techniques have been shown to be effective in combating some treatment side effects. Before embarking on any complementary therapies, patients should discuss the matter with their healthcare team. Certain unproven treatments, including some herbal supplements, can interfere with standard lymphoma treatments or may cause serious side effects.

How to Prepare for Follow-Up Appointments

It is important for patients both during and after treatment to be proactive in their healthcare, including keeping a master file of medical records, asking questions, reporting new symptoms,

exercising and eating a balanced diet. In addition, patients who smoke should strongly consider stopping. Follow-up visits for people with Sézary syndrome often depends on the stage of the disease and treatment and can range from as frequently as every few weeks when starting new therapies that require monitoring to as little as every six months.

Typically, follow-up visits include physical examinations, blood tests and occasionally imaging tests such as CT or PET scans. Besides determining disease recurrence, follow-up care can help identify and resolve unusual side effects of treatment.

Finding Support

Getting a lymphoma diagnosis can be frightening and treatment can cause physical and emotional discomfort. Connecting with other people who have Sézary syndrome can be extremely helpful. The Cutaneous Lymphoma Foundation offers information on patient assistance programs, free e-mail listservs, where to find clinical trials and the latest research on its website at www.clfoundation.org. Support groups and online message boards are often useful. One-to-one peer support programs, such as the Lymphoma Research Foundation's Lymphoma Support Network at www.lymphoma.org, matches lymphoma survivors (or caregivers) with volunteers who have gone through similar experiences.

Staying Informed

The Cutaneous Lymphoma Foundation is a patient advocacy organization dedicated to educating patients and caregivers about cutaneous lymphomas and improving access to safe and effective treatments. For more information about

these resources:

Call: 248-644-9014

Website: www.clfoundation.org

The Lymphoma Research Foundation offers a wide range of resources on the latest treatment and research advances, and services for coping with lymphoma. For more information about any of these resources:

Call: 800-500-9976

E-mail: helpline@lymphoma.org

Website: www.lymphoma.org

How to Support Us

Your contribution helps promote awareness and education about cutaneous lymphomas and supports research. When you send a donation of any amount to the Cutaneous Lymphoma Foundation, you will receive a subscription to the Cutaneous Lymphoma Foundation Forum, our quarterly newsletter.

To make a donation, send a check or money order to:

Cutaneous Lymphoma Foundation

P.O. Box 374

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You may also donate online at www.clfoundation.org

Who We Are

The Cutaneous Lymphoma Foundation is an independent, nonprofit patient advocacy organization dedicated to supporting patients with cutaneous lymphomas by promoting awareness and education, advancing patient care and facilitating research.

Glossary of Terms

Biopsy

Removal of a small piece of tissue (for example, a lymph node) for evaluation under a microscope.

Chemotherapy

Treatment with “chemo” drugs to stop the growth of rapidly dividing cancer cells, including lymphoma cells.

Chemotherapy regimen

Combinations of anti-cancer drugs given at a certain dose in a specific sequence according to a strict schedule.

CT or CAT (computerized axial tomography) scan

This imaging test provides a series of detailed pictures of the inside of the body using an X-ray machine linked to a computer.

Electron beam radiation

Radiation of the skin that does not penetrate to internal organs.

Lymph nodes

Small bean-shaped glands located in the small vessels of the lymphatic system. Thousands are located throughout the body and are most easily felt in the neck, armpits and groin.

Lymphatic system

The vessels, tissues and organs that store and carry lymphocytes that fight infection and other diseases.

Lymphocyte

A type of white blood cell.

PET (positron emission tomography) scan)

PET scans may be used instead of gallium scans

to identify areas in the body that are affected by lymphoma. This test evaluates metabolic activity in different parts of the body using a radioisotope.

Relapse

The return of cancer after treatment. Lymphoma may recur in the area where it first started or it may relapse in another area of the skin.

Refractory disease

A cancer that is resistant to treatment.

Stage

The extent of cancer on the skin or in the body, including whether the disease has spread from the original site to other body parts.



Contact Us

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Because each person's body and response to treatment is different, no individual should indulge in self-diagnosis or embark upon any course of medical treatment without first consulting with his or her physician.

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