Getting the Facts

LYMPHOMA RESEARCH • FOUNDATION

Cutaneous T-Cell Lymphoma

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). Lymphoma occurs when lymphocytes, a type of white blood cell, grow abnormally. The body has two main types of lymphocytes that can develop into lymphomas: B-lymphocytes (B-cells) and T-lymphocytes (T-cells). Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs, and can accumulate to form tumors.

T-cell lymphomas, which occur when T-lymphocyte cells become cancerous, account for 10 percent to 15 percent of all NHL cases in the United States. There are many different forms of T-cell lymphoma, some of which are extremely rare. 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 T-cell lymphoma (CTCL), a general term for T-cell lymphomas that involve the skin. CTCL also can involve the blood, the lymph nodes and other internal organs.

Types of Cutaneous T-Cell Lymphoma

CTCL describes many different disorders with various symptoms, outcomes and treatment considerations. The two most common types of CTCL are mycosis fungoides and Sezary syndrome.

Mycosis fungoides is the most common type of CTCL, with approximately 16,000 to 20,000 cases across the United States. The disease does not look the same for all patients and may present itself as patches, plaques or tumors. Patches are usually flat, possibly scaly and look like a rash. Patches are often mistaken for eczema, psoriasis or non-specific dermatitis. Plaques are thicker, raised lesions. Tumors are raised bumps, which may or may not ulcerate. A common characteristic is itching, although some patients do not experience itching. It is possible to have one or all three types of lesions.

The disease course for mycosis fungoides is based upon the stage at diagnosis. Most patients with early stage CTCL have slow-growing disease that often does not progress to higher stages. While many patients progress slowly, or not at all, some will progress rapidly.

Most patients experience skin symptoms without serious complications. However, approximately 10 percent who experience progressive disease with lymph node and/or internal involvement develop 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.

Mycosis fungoides is difficult to diagnose in its early stages, as the symptoms and skin biopsy findings are similar to those of other skin conditions. A medical history, physical exam and skin biopsy are essential for diagnosis. A physician will examine lymph nodes, order various blood tests and may conduct other screening tests, such as a chest x-ray or CT (computerized tomography) scan. Scans are usually not needed for those with the earliest stages of the disease.

Sezary syndrome is an advanced, variant form of mycosis fungoides, which distinguishes itself by the presence of malignant lymphocytes in the blood. It is usually characterized by extensive thin, red, itchy rashes covering over 80 percent of the body. In certain cases, patches and tumors appear. These symptoms may be accompanied by changes in the nails, hair or eyelids or the presence of enlarged lymph nodes.

Many of the same procedures used to diagnose and stage other types of cutaneous T-cell lymphomas are used in Sezary syndrome, including a physical exam and history, blood tests, 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. A bone marrow biopsy may be done, but is usually not necessary to complete staging.

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The Lymphoma Research Foundation offers the following patient education and support programs:

- *Lymphoma Helpline*
- Clinical Trials Information Service
- Lymphoma Support Network
- Publications
- Teleconferences
- Webcasts & podcasts
- In-person conferences

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Treatment Options

Selecting a treatment for a patient depends on the symptoms, the patient's general health and stage of disease. For mycosis fungoides, treatment is either directed at the skin or the entire body (systemic). Because Sezary syndrome is chronic and systemic (affecting the entire body), it is usually not treated with skin-directed therapies alone. Treatments may be prescribed alone or in combination to achieve the best long-term benefit.

Skin-Directed Therapies

- Ultraviolet light (PUVA, UVB, narrow-band UVB)
- Topical steroids
- Topical chemotherapies (nitrogen mustard, carmustine)
- Topical retinoids (bexarotene gel)
- Local radiation
- Total skin electron beam therapy

Systemic Therapies

- Denileukin diftitox (Ontak)
- Extracorporeal photopheresis
- Gemcitabine (Gemzar)
- Interferon alfa
- Liposomal doxorubicin (Doxil)
- Methotrexate (Trexall)
- Oral retinoids (bexarotene capsules)
- Romidepsin (Istodax)
- Vorinostat (Zolinza)

Some second-line therapies, in addition to those listed above, for relapsed (recurrence of the disease) or refractory (disease that is resistant to treatment) patients include: bortezomib (Velcade), chlorambucil (Leukeran), cyclophosphamide (Cytoxan), etoposide (Toposar), pentostatin (Nipent), and temozolomide (Temodar).

Treatments Under Investigation

There are several treatments being tested in clinical trials for CTCL.

- Allogeneic stem cell transplant
- Autologous dendritic cell vaccine
- Bortezomib (Velcade)
- Enzastaurin
- Forodesine (BCX-1777)
- Lenalidomide (Revlimid)
- Mogamulizumab (KW-0761)
- Pralatrexate (Folotyn)
- Vorinostat (Zolinza)
- Zanolimumab (HuMax-CD4)

Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with the Lymphoma Research Foundation or their physician for any treatment updates that may have recently emerged.

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for lymphoma patients. Patients interested in participating in a clinical trial should talk to their physician. Contact the Lymphoma Research Foundation's *Helpline* for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Resources

The Lymphoma Research Foundation (LRF) offers a range of resources addressing treatment options, research advances and coping with lymphoma. For a more comprehensive source of NHL information, visit LRF's website to view or order the publication entitled *Understanding Non-Hodgkin Lymphoma: A Guide for Patients, Survivors and Loved Ones.* LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts. For more information, visit lymphoma.org, e-mail helpline@lymphoma.org or call (800) 500-9976.