

Transformed Mycosis Fungoides

Overview

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

One of the most common forms of T-cell lymphoma is cutaneous T-cell lymphoma (CTCL), a general term for T-cell lymphomas that cause skin lesions. Although the skin is involved, the skin cells themselves are not cancerous, but rather the T cells that move from the blood to the skin tissue are. CTCL describes many different disorders with various symptoms, outcomes, and treatment considerations. Mycosis fungoides (MF) is the most common type of CTCL, with approximately five cases per one million persons across the United States.

Several forms of lymphoma can undergo a transformation, a process by which an indolent (slow-growing) lymphoma transforms into a typically more aggressive (fast-growing) lymphoma. Transformed MF occurs when a specific subset of MF tumor cells undergo molecular and/or genetic changes that cause them to become larger. It is the presence of these larger cells (in a specific quantity) that differentiates transformed MF from MF. Only some MF patients develop transformed MF, making the disease very rare. The only sure way to know if a patient has transformed MF is to do a biopsy and have it reviewed by a pathologist (a doctor who specializes in making a disease diagnosis by looking at tumor tissue under the microscope).

Transformed MF typically, but not always, acts aggressively. Physicians of patients with transformed MF may see signs that include the presence of a new solitary nodule within a classic MF patch or plaque, an abrupt onset of multiple scattered papules and/or nodules that do not go away on their own, and new or enlarging tumors. The type of treatment recommended for any individual patient depends on the extent of the disease, and the number and

types of prior treatments. Aggressive disease is usually treated with higher potency therapies, while less aggressive disease or earlier stage is usually treated with lower potency therapies.

Treatment Options

For most patients with transformed MF, physicians utilize approaches that have been proven effective for other forms of aggressive T-cell lymphomas. A patient whose MF has transformed should be seen by an experienced specialist to develop an individualized treatment plan. The U.S. Food and Drug Administration (FDA) has approved several therapies for the treatment of patients with transformed MF.

Therapeutic options for transformed MF patients with limited areas of skin involvement include:

- Mechlorethamine topical gel (e.g., Valchlor)
- Local radiation
- Post-radiation maintenance therapy with skin-directed treatments such as low-dose bexarotene (Targetin), methotrexate (Trexall), or interferon
- Continuation of the skin-directed therapy that the MF patient was receiving prior to transformation, which could include steroids, methotrexates, or cyclophosphamide (Cytoxan) among others

Transformed MF patients with multiple areas of skin involvement are usually treated with systemic therapy (treatment directed at the entire body), which can include:

- Denileukin diftitox (Ontak)
- Romidepsin (Istodax)

In addition, combination therapy may be used for patients with more advanced disease. Patients may also benefit from an allogeneic stem cell transplant (in which patients receive stem cells from a donor) if it is determined that they are good candidates for this procedure. For more information on transplantation, view the *Transplant in Lymphoma* fact sheet on the Lymphoma Research Foundation's (LRF's) website at www.lymphoma.org.

Treatments Under Investigation

Treatments under investigation for transformed MF include bortezomib (Velcade), brentuximab vedotin (Adcetris), everolimus

National Headquarters

115 Broadway, Suite 1301
New York, NY 10006
(212) 349-2910
(212) 349-2886 fax

Helpline: (800) 500-9976
helpline@lymphoma.org

Website: www.lymphoma.org

Email: LRF@lymphoma.org

Medical reviewer:

Francine Foss, MD
Yale Cancer Center

Supported through grants from:



Genentech
A Member of the Roche Group



© 2015 Lymphoma Research Foundation

Getting the Facts is published by the Lymphoma Research Foundation (LRF) for the purpose of informing and educating readers. Facts and statistics were obtained using published information, including data from the Surveillance, Epidemiology, and End Results (SEER) Program. Because each person's body and response to treatment is different, no individual should self-diagnose or embark upon any course of medical treatment without first consulting with his or her physician. The medical reviewer, the medical reviewer's institution, and LRF are not responsible for the medical care or treatment of any individual.

Last Updated September 2015

Stay Connected through our social media



(Afinitor), lenalidomide (Revlimid), and pralatrexate (Folotyn). Other treatments include immunotherapy with genetically engineered T cells, which is being tested in early trials for patients with transformed MF. Various other trials are investigating combinations of agents with and without stem cell transplant in patients with transformed MF. It is critical to remember that today's scientific research is continuously evolving. Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that patients check with their physician or LRF for any treatment updates that may have recently emerged.

Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for lymphoma patients. Because transformed MF is such a rare disease, clinical trial enrollment is critical to establishing more effective, less toxic treatments. The rarity of the disease also means that the most novel treatments are often available only through clinical trials. Patients interested in participating in a clinical trial should talk to their physician or contact LRF's Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Follow-up

Because transformed MF is generally characterized by multiple disease relapses after responses to a variety of treatments, patients in remission should have regular visits with a physician who is familiar with their medical history and the treatments they have received. Medical tests (such as blood tests and computed axial tomography [CAT] scans) may be required at various times during remission to evaluate the need for additional treatment.

Some treatments can cause long-term effects or late effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. The physician will check for these effects during follow-up care.

Survivors and their caregivers are encouraged to keep copies of all medical records and test results as well as information on the types, amounts, and duration of all treatments received. This documentation will be important for keeping track of any effects resulting from treatment or potential disease recurrences.

Support

A lymphoma diagnosis often triggers a range of feelings and raises concerns. In addition, cancer treatment can cause physical and emotional discomfort. Support groups and online message boards can help patients connect with other people who have lymphoma. One-to-one peer support programs, such as the LRF Lymphoma Support Network, match lymphoma patients (or caregivers) with volunteers who have gone through similar experiences.

Resources

LRF offers a wide range of resources that address treatment options, the latest research advances, and ways to cope with all aspects of lymphoma, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts, as well as disease-specific websites, videos, and eNewsletters for current lymphoma information and treatment options. To learn more about any of these resources, visit our website at www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.