LYMPHOMA
RESEARCH • FOUNDATION

Getting the Facts

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

Mantle 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 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).

Mantle cell lymphoma (MCL) is a rare B-cell NHL that most often affects men over the age of 60. The disease may be aggressive (fast-growing), but it can also behave in a more indolent (slow-growing) fashion in some patients. MCL comprises about five percent of all NHLs. The disease is called "mantle cell lymphoma" because the tumor cells originally come from the "mantle zone" of the lymph node. MCL is usually diagnosed as a late-stage disease that has typically spread to the gastrointestinal tract and bone marrow.

A diagnosis of MCL requires taking a small sample of tumor tissue, called a biopsy, and looking at the cells under a microscope. A blood test may also be necessary to measure the white blood cell count and certain proteins, which help to diagnose MCL. Other tests, such as a bone marrow biopsy, computed axial tomography (CAT) scan, or positron emission tomography/computerized tomography (PET/CT) scan may be used to confirm a diagnosis and to determine what areas of the body are involved by the cancer.

Overproduction of a protein called cyclin D1 is found in more than 90 percent of patients with MCL. Identification of excess cyclin D1 from a biopsy is considered a very sensitive tool for diagnosing MCL. One-quarter to one-half of patients with MCL also have higher-than-normal levels of certain proteins that circulate in the blood, such as lactate dehydrogenase (LDH) and beta-2 microglobulin. Measuring these and other proteins can help physicians determine how aggressive an individual patient's MCL is and may guide therapy decisions.

Treatment Options

The type of treatment selected for a patient with MCL depends on multiple factors, including the stage of disease, the age of the patient, and the patient's overall health. For the subset of patients who do not yet have symptoms and who have a relatively small amount of slow-growing disease, "watchful waiting" and monitoring the disease for progression may be an acceptable option. MCL is usually diagnosed once it has spread throughout the body, and the majority of these patients will require treatment. Initial treatment approaches for aggressive MCL in younger patients include combination chemotherapy, typically in combination with the monoclonal antibody rituximab (Rituxan), as first-line treatment, followed by autologous stem cell transplantation (in which patients receive their own stem cells). HyperCVAD (cyclophosphamide, vincristine, doxorubicin, and dexamethasone alternating with methotrexate and cytarabine) plus rituximab are recommended as aggressive induction therapy and are associated with durable remissions in newly diagnosed patients. For older patients, chemotherapy followed by a prolonged course of rituximab alone, known as maintenance therapy, is often recommended. A common chemotherapeutic treatment approach used to treat MCL is called R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone). Bendamustine (Treanda) in combination with rituximab is another common first-line treatment option. Several additional intensified chemotherapy combinations are also used in combination with rituximab, particularly in younger patients.

Although allogeneic stem cell transplantation (in which patients receive stem cells from a donor) is very intensive and causes various side effects, including possibly graft-versus-host disease, it may increase response times for selected younger patients, especially in those whose disease has relapsed (returned after treatment).

Bortezomib (Velcade) is approved by the U.S. Food and Drug Administration (FDA) for the treatment of patients with MCL. Studies with bortezomib show that the drug may be effectively combined with conventional chemotherapy.

Lenalidomide (Revlimid) is another treatment for MCL approved by the FDA for patients who have relapsed or progressed after two prior therapies, one of which included bortezomib. Lenalidomide is an immunomodulatory agent that affects the growth and survival of tumor cells by altering the body's immune cells.

Ibrutinib (Imbruvica) is approved by the FDA for treatment of MCL in patients who have received at least one prior therapy and is also used to treat chronic lymphocytic leukemia. This therapy is a tyrosine kinase inhibitor, which stops signals in cancer cells that are responsible for growth and survival.

Treatment options may change as new treatments are discovered and current treatments are improved. Therefore, it is important that

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:
Ann S. LaCasce, MD
Dana-Farber Cancer Institute

Supported through grants from:

Celgene

Genentech
A Member of the Roche Group

Biogen

Takeda
ONCOLOGY

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 July 2015

Stay Connected through our social media

patients check with their physician or with the Lymphoma Research Foundation (LRF) for any treatment updates that may have recently emerged.

Treatments Under Investigation

Many new drugs used alone or in combination are being studied in clinical trials for MCL, including abemaciclib, bendamustine (Treanda) as first-line treatment, idelalisib (Zydelig), VR-CAP (bortezomib, rituximab, cyclophosphamide, doxorubicin, and prednisone), vorinostat (Zolinza), ofatumumab (Arzerra), everolimus (Afinitor), panobinostat (Farydak), and temsirolimus (Torisel). Some approved treatments are also being investigated for efficacy as maintenance treatments, given to patients in remission who have no signs of lymphoma to prolong remission. Bortezomib is being investigated for this use following treatment with the chemotherapy regimen EPOCH-R (prednisone, etoposide, doxorubicin, cyclophosphamide, vincristine, and rituximab).

Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because the optimal initial treatment of MCL is not clear and it is such a rare disease, clinical trials are very important and will identify the best treatment options in this disease. Patients interested in participating in a clinical trial should view the *Understanding Clinical Trials* fact sheet on LRF's website at www.lymphoma.org, talk to their physician, or contact the LRF Helpline for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.

Follow-up

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 CAT scans) may be required at various times during remission to evaluate the need for additional treatment.

Patients 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 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 websites at www.FocusOnMCL.org or www.lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.