

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 sample of tumor tissue, called a biopsy, and looking at the cells under a microscope. Other tests of the lymphoma cells are necessary to verify a diagnosis of MCL and distinguish it from other subtypes of NHL. A bone marrow biopsy, computed tomography (CT) scan, or positron emission tomography/CT (PET/CT) scan may be used to determine what areas of the body are involved by the cancer.

Overproduction of a protein called cyclin D1 in the lymphoma cells 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” may be an acceptable option. With this strategy, patients’ overall health and disease are monitored through regular checkup visits and various evaluating procedures,

such as laboratory and imaging tests. Active treatment is started if the patient begins to develop lymphoma-related symptoms or there are signs that the disease is progressing based on testing during follow-up visits. 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), though rituximab is not specifically approved by the U.S. Food and Drug Administration (FDA) for MCL. Combination chemotherapy containing cytarabine plus the immunotherapeutic monoclonal antibody rituximab are recommended as aggressive induction therapy and are associated with durable remissions in newly diagnosed patients. Consolidation high-dose chemotherapy followed by autologous stem cell is often utilized to prolong remission in younger, medically fit patients. For older or less fit patients, less intensive 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 related or unrelated donor) is very intensive and causes various side effects, including possibly graft-versus-host disease, it may increase response times for selected younger patients whose disease has *relapsed* (returned after treatment).

Bortezomib (Velcade) is approved by the 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. It may be given in combination with rituximab.

Ibrutinib (Imbruvica) is approved by the FDA for treatment of MCL in patients who have received at least one prior therapy and is also

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used to treat chronic lymphocytic leukemia and Waldenström macroglobulinemia. 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 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 as initial induction therapy for MCL, including acalabrutinib (ACP-196), venetoclax (Venclexta), ofatumumab (Arzerra), and temsirolimus (Torisel). Please view the *Mantle Cell Lymphoma: Relapsed/Refractory* fact sheet for information about treatments being evaluated for relapsed/refractory (disease no longer responds to treatment) MCL.

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 to 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 PET/CT 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 concerns. In addition, cancer treatment can cause physical discomfort. LRF's Lymphoma Support Network is a one-to-one peer support program that connects patients and caregivers with volunteers who have experience with MCL, similar treatments, or challenges, for mutual emotional support and encouragement. You may find this useful whether you or a loved one is newly diagnosed, in treatment, or in remission.

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 and MCL, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts for people with MCL, as well as our *Mantle Cell Lymphoma* e-Updates that provide the latest disease-specific news and treatment options. For more information 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.