

Getting the Facts

Marginal Zone 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).

Marginal zone lymphomas are a group of indolent (slow-growing) NHL B-cell lymphomas, which account for approximately 12 percent of all B-cell lymphomas. The median age for diagnosis is 65 years old.

There are three types of marginal zone lymphoma:

Extranodal marginal zone lymphoma or mucosa-associated lymphoid tissue (MALT) is the most common form of marginal zone lymphoma. It occurs outside the lymph nodes, in places such as the stomach, small intestine, salivary gland, thyroid, eyes, and lungs. MALT lymphoma is divided into two categories: gastric, which develops in the stomach, and non-gastric, which develops outside of the stomach. This form of lymphoma makes up about nine percent of all B-cell lymphomas.

In many cases of MALT lymphoma, there is a previous medical history of inflammation or autoimmune disorders. For example, *Helicobacter pylori* (*H. pylori*), a microbial pathogen linked to chronic gastritis, has been associated with a significant portion of patients with gastric MALT lymphoma.

Nodal marginal zone lymphoma (sometimes called monocytoid B-cell lymphoma) occurs within the lymph nodes and accounts for about two percent of all B-cell lymphomas.

Splenic marginal zone lymphoma occurs most often in the spleen and blood. It has been associated with Hepatitis C. This form of lymphoma makes up about one percent of all B-cell lymphomas.

Treatment Options

The type of treatment selected for a marginal zone lymphoma patient

depends on the stage of disease, the age of the patient, the patient's overall health, any signs or symptoms related to the lymphoma, and the location of the disease.

Treatment Options for Gastric MALT

For gastric MALT lymphoma, the initial treatment is antibiotic therapy, which is typically given for two weeks. Approximately 70 percent to 90 percent of patients respond to antibiotic therapy, and approximately half of the patients require no further treatment. If the lymphoma relapses after antibiotic therapy, there are many additional treatment options available, including (in alphabetical order):

- Bendamustine (Treanda)
- Bortezomib (Velcade)
- Chemotherapy
- Fludarabine (Fludara)
- Radiation (low-dose)
- Rituximab (Rituxan)
- Surgical excision

Treatment Options for Non-gastric MALT

Non-gastric MALT can appear in a variety of areas throughout the body. Therefore, treatment is usually based on the exact location and extent of spread. Treatment typically includes surgery for certain sites (lung, breast) or radiation therapy with or without chemotherapy. More advanced disease usually includes immunoradiotherapy with chemotherapy. Among the common first-line treatments are bendamustine plus rituximab and R-CHOP (rituximab, cyclophosphamide, doxorubicin vincristine, prednisone). Recently, antibiotic therapy such as doxycycline has been shown to be effective in marginal zone lymphoma that affects the area around the eye ("ocular adnexal marginal zone lymphoma").

Treatment Options for Nodal Marginal Zone Lymphoma

Because nodal marginal zone lymphoma is most often a slow-growing disease, doctors may defer treatment until symptoms appear, an approach called "watch and wait" or "watchful waiting." When treatment is necessary, options include radiation therapy, chemotherapy, and other treatments commonly used in other types of slow-growing lymphomas, such as follicular lymphoma.

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Treatment Options for Splenic Marginal Zone Lymphoma

Several treatment options exist for splenic marginal zone lymphoma. Some patients receive a splenectomy (surgical removal of the spleen) or, for those patients who are not surgical candidates, low-dose radiation of the spleen. Other patients may receive rituximab, a monoclonal antibody, with or without chemotherapy.

Treatments Under Investigation

Several new drugs and drug combinations are being studied in clinical trials for marginal zone lymphoma and other slow-growing lymphomas, including:

- B-cell receptor signaling blockers
- Ibritumomab tiuxetan (Zevalin)

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 LRF or with their physician 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. 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

Patients in remission should have regular visits with a physician who is familiar with their medical history as well as the treatments they have received. Patients will likely have some medical tests (such as blood tests and computed axial tomography [CAT] scans) at various times during remission so that the physician can evaluate whether additional treatment is needed.

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.

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. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts. For more information about any of these resources, visit the website at www.lymphoma.org or contact the Helpline at (800) 500-9976 or helpline@lymphoma.org.