

CNS 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).

NHL is broadly categorized as B-cell lymphomas or T-cell lymphomas. B-cell lymphomas develop from abnormal B-cells and account for 85 percent of all NHLs. T-cell lymphomas develop from abnormal T cells and account for the remaining 15 percent of all NHLs. NHLs may also be classified as *indolent* (slow-growing) or *aggressive* (fast-growing).

Primary central nervous system (CNS) lymphoma is an aggressive form of NHL in which *malignant* (cancer) cells form in the lymph tissue of the brain and/or spinal cord. It may develop in the brain, spinal cord, eye, or *leptomeninges* (two of the membranes that surround the brain and spinal cord). When lymphoma has originated in other parts of the body and subsequently has spread to the CNS, it is referred to as secondary CNS lymphoma. The main symptoms of CNS lymphoma are focal neurological deficits (i.e., problems with nerve, spinal cord, or brain function), but headaches, vomiting, confusion, seizures, personality changes, and blurred vision can also occur.

The cause of CNS lymphoma is unknown, but there are some factors that may increase the risk of developing it, such as infection with Epstein-Barr virus (EBV) or human immunodeficiency virus (HIV); a compromised immune system (which may be the case for people with acquired immunodeficiency syndrome [AIDS] or patients who have undergone organ transplant); exposure to chemicals such as pesticides, solvents, or fertilizers; and a family history of NHL. Having one or more of these risk factors does not mean a person will develop NHL. Most people with risk factors never develop the disease and many people diagnosed have never been exposed to any clearly identifiable risk factors.

Treatment Options

Until the mid-1990s, radiation was the standard therapy for patients with CNS lymphoma. Now high-dose methotrexate-based therapy is recommended for most patients in combination with other agents such as vincristine (Oncovin) and procarbazine (Matulane) plus or minus cytarabine (Cytosar-U), plus or minus rituximab (Rituxan); or with ifosfamide (Ifex) plus or minus rituximab; or with cytarabine plus or minus rituximab; or with temozolomide (Temodar) and rituximab. Furthermore, this *initial treatment* (induction) is often followed by other therapy regimens (“consolidation”) to reduce the risk of recurrence. These consolidation therapies include high-dose chemotherapy alone such as cytarabine; cytarabine and etoposide; high-dose chemotherapy with stem cell transplant; or whole brain radiation therapy (WBRT). WBRT is seldom used in patients over the age of 65 because of progressive neurological complications (e.g., loss of memory or muscle coordination).

If lymphoma cells are found in the spinal fluid, chemotherapy is often delivered directly into the spinal fluid, in addition to the above chemoimmunotherapy regimens.

Treatments Under Investigation

Various agents are being investigated in clinical trials for patients with newly diagnosed CNS lymphoma and those who are *relapsed* (disease returns after treatment)/*refractory* (disease does not respond to treatment). Some of the agents being investigated for the treatment of patients with CNS include:

- Bendamustine (Treanda)
- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Nivolumab (Opdivo)
- Pembrolizumab (Keytruda)
- Pemetrexed (Alimta)
- Pomalidomide (Pomalyst)
- PQR309
- Temsirolimus (Torisel)

Treatment options are changing as new therapeutics become available and current treatments are improved. Because today’s scientific research is continuously evolving, 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.

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Supported through grants from:



Genentech
A Member of the Roche Group



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Last Updated June 2016

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Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. The Lymphoma Research Foundation (LRF) provides a Clinical Trials Information Service to increase awareness about investigational treatments for lymphoma being evaluated at cancer treatment centers nationwide. Our LRF Helpline staff will conduct a search for potential lymphoma treatment trials based upon medical information that is provided. Patients are strongly encouraged to discuss with their physician the summaries emailed or mailed by LRF. The patient's cancer specialist will be familiar with their medical history and can best evaluate all of the study criteria to determine if the clinical trial is appropriate.

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.

Support

A lymphoma diagnosis often triggers a range of feelings and concerns. In addition, cancer treatment can cause physical discomfort. One-to-one peer support programs, such as LRF's Lymphoma Support Network, connects patients and caregivers with volunteers who have experience with CNS lymphoma, 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.

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 computed tomography [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.

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 for people with CNS lymphoma, as well as disease-specific websites, videos, and e-Updates 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.