LYMPHOMA
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Getting the Facts

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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 approximately 90 percent of all NHLs. T-cell lymphomas develop from abnormal T cells and account for approximately 10 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. In more than 90 percent of cases, it is a B-cell lymphoma. 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 (such as 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 a compromised immune system, (for example, 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 most 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 with rituximab (Rituxan) is recommended for most patients. Sometimes, this is given in combination with other agents such as cytarabine (Cytosar), vincristine (Oncovin), procarbazine (Matulane), ifosfamide (Ifex), or temozolomide (Temodar). 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 (Etopophos, Toposar); 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 (for example, 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:

- Ibrutinib (Imbruvica)
- Lenalidomide (Revlimid)
- Nivolumab (Opdivo)
- Obinutuzumab (Gazyva)
- Pembrolizumab (Keytruda)
- PQR309

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.

Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for patients with lymphoma. Because CNS lymphoma is a rare disease, clinical trial enrollment is critical for 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 view the *Understanding Clinical Trials* fact sheet on LRF's website at lymphoma.org/publications, 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 with lymphoma 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, computed tomography [CT] scans, and positron emission tomography [PET] scans) may be required at various times during *remission* (disappearance of signs and symptoms) to evaluate the need for additional treatment.

Some treatments can cause long-term side effects or late side effects, which can vary based on the duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. A physician will check for these side effects during follow-up care. Visits may become less frequent the longer the disease remains in remission.

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 side effects resulting from treatment or potential disease recurrences. LRF's award-winning *Focus On Lymphoma* mobile app (lymphoma.org/mobileapp) and the *Lymphoma Care Plan* (lymphoma.org/publications) can help patients manage this documentation.

Patient and Caregiver Support Services

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*, connect patients and caregivers with volunteers who have experience with CNS lymphoma, similar treatments, or challenges, for mutual emotional support and encouragement. Patients and loved ones may find this useful whether the patient 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, including our award-winning mobile app. LRF also provides many educational activities, from in-person meetings to teleconferences and webcasts for people with lymphoma and CNS lymphoma, as well as patient guides and e-Updates that provide the latest disease-specific news and treatment options. To learn more about any of these resources, visit our websites at lymphoma.org/CNS or lymphoma.org, or contact the LRF Helpline at (800) 500-9976 or helpline@lymphoma.org.

Contact the Lymphoma Research Foundation
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