LYMPHOMA
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Getting the Facts

Follicular Lymphoma: Relapsed/Refractory

Overview

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma 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).

Follicular lymphoma (FL), a B-cell lymphoma, is the most common indolent (slow-growing) form of NHL, accounting for approximately 20 percent to 30 percent of all NHLs. Common signs of FL include enlargement of the lymph nodes in the neck, underarm, stomach, or groin, as well as fatigue, shortness of breath, night sweats, and weight loss. Often, people with FL have no obvious symptoms of the disease at diagnosis.

After treatment, many patients will have a remission that lasts for years; however, the disease does return in most patients. For those patients who relapse (disease returns after treatment) or become refractory (disease does not respond to treatment), second-line therapies are often successful in providing another remission.

Treatment Options

Treatment for relapsed FL is based on the duration of remission from the last treatment received and the symptoms of the cancer. Chemotherapy, radiation, and monoclonal antibodies, such as rituximab (Rituxan) may be used alone or in combination to treat relapsed or refractory follicular lymphoma. Common second-line regimens include:

- R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone)
- R-CVP (rituximab, cyclophosphamide, vincristine, prednisone)
- Bendamustine (Treanda) alone or with rituximab

Radioimmunotherapy (RIT) may also be used alone or in combination with chemotherapy to treat relapsed or refractory FL. Currently, two radioimmunotherapy drugs are commercially available: Iodine 131 tositumomab (Bexxar) and Y90 ibritumomab tiuxetan (Zevalin). While there are some differences between these two drugs, both produce similar clinical results with durable remissions for appropriate patients. To learn more about RIT, view the Lymphoma Research Foundation (LRF) Radioimmunotherapy fact sheet.

For some patients with relapsed FL, high-dose chemotherapy and an autologous stem cell transplant (in which patients receive their own stem cells) or an allogeneic stem cell transplant (in which patients receive stem cells from a donor) may provide a prolonged disease-free interval.

Approximately 30 percent to 40 percent of patients with FL will eventually develop a transformed lymphoma, which is often more aggressive and usually treated with high-dose chemotherapy along with an autologous stem cell transplant.

Treatments Under Investigation

Many treatments are currently being tested in clinical trials as single-agent therapy or as part of a combination therapy regimen in patients with relapsed/refractory FL. These treatments include:

- Bortezomib (Velcade)
- Everolimus (Afinitor)
- Idelalisib (GS-1101, formerly CAL-101)
- Ibrutinib (PCI-32765)
- Lenalidomide (Revlimid)
- Ofatumumab (Arzerra)
- Panobinostat

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

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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

Because FL is generally characterized by multiple disease relapses after responses to a variety of treatments, patients in remission should have regular visits with a physician who is familiar with their medical history as well as with the treatments they have received. Medical tests (such as blood tests and computed axial tomography [CAT] scans) may be required at various times during remission to evaluate the need for additional treatment.

Some treatments can cause long-term effects or late effects, which can vary based on duration and frequency of treatments, age, gender, and the overall health of each patient at the time of treatment. The doctor will check for these effects during follow-up care.

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.

Support

A lymphoma diagnosis often triggers a range of feelings and 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 LRF's Lymphoma Support Network, match lymphoma survivors (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. 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 www.FocusOnFL.org. You can also contact the Helpline at (800) 500-9976 or helpline@lymphoma.org.