

Peripheral T-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 lymphocytes, a type of white blood cell, grow abnormally. The body has two main types of lymphocytes that can develop into lymphomas: B-lymphocytes (B-cells) and T-lymphocytes (T-cells). Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood or other organs, and can accumulate to form tumors.

Peripheral T-cell lymphoma (PTCL) comprises a group of rare and usually aggressive NHLs that develop from mature T-cells. Peripheral T-cell lymphoma represents a spectrum of T-cell lymphomas and accounts for approximately 10 percent to 15 percent of all NHL cases in the United States.

Types of Peripheral T-Cell Lymphoma

PTCLs are sub-classified into various specific diseases based on distinct clinical and/or pathological patterns.

Peripheral T-Cell Lymphoma Unspecified (PTCL-US) comprises a group of diseases that do not fit into any of the other subtypes of PTCL. PTCL-US is the most common subtype, making up about one quarter of all diagnosed PTCLs. It is also the most common of all the T-cell lymphomas. The term PTCL can be confusing as it can refer to the entire spectrum of mature T-cell lymphomas or sometimes to this specific subtype, PTCL-US, only. Although most patients with PTCL-US present with lymph node involvement, sites outside the lymph nodes, such as the liver, bone marrow, gastrointestinal tract and skin, may also be involved. This group of PTCLs is considered aggressive and requires standard combination chemotherapy upon diagnosis.

Anaplastic Large-Cell Lymphoma (ALCL) is a rare type of aggressive T-cell lymphoma comprising only 3 percent of all lymphomas in adults (about 15 percent to 20 percent of all PTCLs) and between 10 percent and 30 percent of all lymphomas in children. ALCL can appear in the skin or in other organs throughout the body (systemic ALCL). Prognosis and treatment

are different for each subtype. For more information, see LRF's ALCL Fact Sheet.

Angioimmunoblastic T-Cell Lymphoma (AITL) is an often fast-growing T-cell lymphoma that accounts for between 1 percent and 2 percent of all NHL cases (about 15 percent to 20 percent of all PTCLs) in the United States. This type of lymphoma often responds to milder therapies, such as steroids, although it often progresses and requires chemotherapy and other medications. In advanced cases, bone marrow transplantation may be used. For more information, see LRF's AITL Fact Sheet.

Enteropathy-Type T-Cell Lymphoma is an extremely rare subtype that appears in the intestines and is strongly associated with celiac disease. As with other rare cancers, patients should discuss treatment options with their medical team.

Nasal NK/T-Cell Lymphoma involves natural killer (NK) cells, which have specific jobs in the normal immune system. NK cells are closely related to T-cells and often have features that overlap with normal T-cells. If they become a cancer, they are called an NK or NK/T-cell lymphoma and are grouped with other forms of PTCL. Although this fast-growing lymphoma is very rare in the United States, it is more common in Asia and parts of Latin America, leading researchers to suspect that some ethnic groups may be more prone to this cancer. This type of lymphoma is associated with the Epstein-Barr virus and most often involves the nasal area, trachea, gastrointestinal tract or skin. As with other rare cancers, patients should consult with their medical team for treatment options and the availability of clinical trials.

Hepatosplenic Gamma-Delta T-Cell Lymphoma is an extremely rare and aggressive disease that starts in the liver or spleen. As with other rare cancers, patients should discuss treatment options with their medical team.

Cutaneous T-cell Lymphomas (CTCL) are a group of lymphomas that originate in the skin. CTCLs are a subset of PTCL as they are lymphomas of mature T-cells. However, these lymphomas are generally less aggressive, have a different prognosis, and have different treatment approaches than the aggressive PTCLs.

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Mycosis Fungoides is the most common type of cutaneous T-cell lymphoma. It is generally a slow-growing cancer that starts in the skin, appearing as a scaly, red rash in areas of the body that are not usually exposed to the sun. *Sézary Syndrome* is an advanced, variant form of mycosis fungoides, and affects both the skin and the peripheral blood. It can cause widespread itching, reddening and peeling of the skin as well as skin tumors. For more information, see LRF's CTCL Fact Sheet.

Treatment Options

For most subtypes of PTCL, the frontline treatment regimen is typically a combination chemotherapy, such as CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) or EPOCH (etoposide, vincristine, doxorubicin, cyclophosphamide, prednisone) or other multi-drug regimens. Because most PTCL patients will relapse, some oncologists recommend giving high-dose chemotherapy followed by an autologous stem cell transplant to some patients who had a good response to their initial chemotherapy program. While promising, there is no firm clinical data to support that undergoing a transplant in this setting is better than not undergoing a transplant.

In fall 2009, the United States Food and Drug Administration approved pralatrexate (Folotyn) for the treatment of patients with relapsed (disease returns after treatment) or refractory (disease does not respond to treatment) PTCL. Pralatrexate (Folotyn) was the first drug approved specifically for PTCL patients. Clinical trials are in development and underway to see if pralatrexate (Folotyn) is potentially synergistic with other drugs commonly used in the treatment of both T-cell and B-cell lymphomas, including gemcitabine (Gemzar), HDAC inhibitors and proteasome inhibitors.

Relapsed PTCL patients may also be treated with combination chemotherapy programs such as ICE (ifosfamide, carboplatin, etoposide), followed by an autologous or allogeneic stem cell transplant. Gemcitabine (Gemzar) appears effective against some forms of relapsed PTCL and is often given in combination with other chemotherapies, including vinorelbine (Navelbine) and doxorubicin (Doxil) in a regimen called GND. Other chemotherapy regimens include DHAP (dexamethasone, cytarabine, cisplatin) and ESHAP (etoposide, methylprednisolone, cytarabine and cisplatin). In June 2011, the U.S. Food and Drug Administration (FDA) granted accelerated approval of romidepsin (Istodax) for injection for the treatment of peripheral T-cell lymphoma (PTCL) in patients who have received at least one prior therapy.

Treatments Under Investigation

Many new drugs are being studied in clinical trials for the treatment of PTCL, including:

- ABT-262
- Bortezomib (Velcade)
- Obatoclox (GX15-070)
- PDX101 (Belinostat)
- Vorinostat (Zolinza)
- Zanolimumab
- AT-101
- Decapeptide
- Panobinostat (Faridak)
- Romidepsin (Istodax)
- Lenalidomide (Revlimid)
- Brentuximab Vedotin (Adcetris)

Participating in Clinical Trials

Clinical trials are crucial in identifying effective drugs and determining optimal doses for lymphoma patients. Because peripheral T-cell lymphomas comprise such a rare group of diseases, clinical trial enrollment is crucial to establishing more effective, less toxic treatments. The rarity of these diseases also means that the most novel treatments are often only available through clinical trials. Patients interested in participating in a clinical trial should talk to their physician. Contact the Lymphoma Research Foundation's *Helpline* for an individualized clinical trial search by calling (800) 500-9976 or emailing helpline@lymphoma.org.