Article Series Lymphoma

Peripheral T-cell lymphomas (PTCLs)

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Peripheral T-cell lymphomas (PTCLs) are a group of rare and often fast-growing blood cancers. They start in the lymphatic system, developing from mature T cells and natural killer (NK) cells. T cells and NK cells are types of white blood cells that work to keep you healthy and kill infected cells, like cancer cells. PTCLs cause your T cells and NK cells to grow and develop abnormally.

PTCLs are classified as a rare disease, accounting for just 5%–10% of all non-Hodgkin lymphoma cases diagnosed in the United States. PTCLs generally affect people aged 60 years and older and occur slightly more often in men than in women. However, younger adults and children can sometimes be diagnosed with a

type of PTCL as well.

What are the different types of peripheral T-cell lymphoma?
There are over 27 different types of PTCL. Some types are more common in the United States, while some are more common in places like Asia and the Caribbean.

Common subtypes of PTCL include:

- PTCL-not-otherwise specified (PTCL-NOS).
- Anaplastic large cell lymphoma (ALCL)
- Primary cutaneous anaplastic large cell lymphoma (pcALCL)
- Angioimmunoblastic T-cell lymphoma (AITL)
- Extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENK/TCL)
- Adult T-cell leukemia/lymphoma (ATLL)
- Enteropathy-associated T-cell lymphoma (EATL)
- Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL)
- Hepatosplenic T-cell lymphoma (HSTCL)
- Subcutaneous panniculitis-like T-cell lymphoma (SPTCL)
- Primary cutaneous gamma/delta T-cell lymphoma (PCGD-TCL)

What are the signs and symptoms of peripheral T-cell lymphomas?
PTCLs can affect various parts of the body including the lymph nodes, skin, gastrointestinal tract, liver and spleen. The signs and symptoms can vary depending on the type of PTCL but there are some common signs and symptoms that many people with PTCL experience.

Signs and symptoms may include:

- **Enlarged lymph nodes:** lymph nodes in the neck, armpit or groin may become swollen and may or may not be painful
- **Fever:** fevers that persist for several days or weeks and may be accompanied by night sweats
- **Fatigue:** extreme tiredness that does not improve with rest
- **Skin rash:** a skin rash or lesions that may be itchy and appear anywhere on the body
- **Abdominal pain or swelling:** enlargement of abdominal organs can cause pain or a feeling of fullness
- **Weight loss:** unintentional weight loss that is significant or rapid and may be accompanied by a loss of appetite

What happens if you have signs or symptoms of peripheral T-cell lymphoma?

Your doctor will start by examining your lymph nodes and other areas of the body that may be affected including the skin, the spleen and the liver. If they suspect lymphoma, a biopsy will come next.

Biopsy

A biopsy is a procedure to remove a sample of cells from the body so that it can be examined under a microscope. The biopsy tissue samples will be sent to a hematopathologist. A hematopathologist is a doctor who has special training in diagnosing blood diseases.

Subtype

If lymphoma is found, additional tests are done on the samples to determine the subtype of lymphoma. Because PTCLs are rare, and there are numerous types, an accurate diagnosis can be challenging. This type of diagnosis needs an experienced hematopathologist. A second opinion may be needed to confirm a diagnosis. If you or a loved one are going through this process, **ask your doctor if they can get a second hematopathologist opinion**. A correct diagnosis is essential in determining how your disease will progress and an appropriate treatment plan.

Stage

If your doctors conclude that you have a type of PTCL, finding out the stage of your cancer comes next. Staging is the process of determining how much cancer is in the body and where it is located. This may be done through blood tests, additional biopsies, and/or imaging tests.

Treatment

Once the type and stage of your cancer is determined, you will talk to your doctor about treatment options. A hematologist-oncologist is a doctor who specializes in treating blood cancers. **It is important to find a treatment center with hematologist-oncologists specializing in PTCLs.**

If your current medical center does not have hematologist-oncologists with PTCL experience, ask your doctor for a referral. Nonprofit patient support groups such as The Leukemia and Lymphoma Society® may also be able to help connect you with a treatment center. Visit www.LLS.org/InformationSpecialists for more information.