

## Anaplastic Large Cell Lymphoma

Expert review by:

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### Non-Hodgkin Lymphoma Overview

Lymphoma is a cancer of the white blood cells, namely lymphocytes, that happen to constitute the lymphatic system. Of the more than 67 types of lymphoma, over 61 are classified as non-Hodgkin lymphoma (NHL). Nearly all non-Hodgkin lymphoma cases occur in adults, with the average age of diagnosis in the 60s. While scientists do not know the exact causes of non-Hodgkin lymphoma, they do know that it is not caused by injury or by coming into contact with someone else with the disease. Most people diagnosed with non-Hodgkin lymphoma have no known risk factors, although increasingly many scientists believe infections may play an important role in causing select types of non-Hodgkin lymphoma to develop.

### Anaplastic Large Cell Lymphoma Overview

Anaplastic large cell lymphoma (ALCL) is a rare type of aggressive T-cell lymphoma, comprising about 3 percent of all non-Hodgkin lymphomas in adults and between 10 percent and 30 percent of all non-Hodgkin lymphomas in children. This type of lymphoma can present in either the systemic (throughout the body) or cutaneous (skin) form. When ALCL presents in the cutaneous form it's called primary cutaneous anaplastic large cell lymphoma. There are two subtypes of ALCL, ALK (anaplastic lymphoma kinase) positive and ALK negative. The ALK positive subtype usually affects children and young adults and ALK negative is more commonly found in older patients over age 60. This lymphoma is more often found in men than women.

Symptoms of ALCL include weight loss, night sweats, enlarged lymph nodes throughout the body, especially in the neck or armpits and skin lesions in the cutaneous form.

A chronic skin disease called lymphomatoid papulosis (LyP), in which papules arise usually on the trunk and limbs, has been linked to the development of ALCL. Although the World Health Organization and the European Organization for Research and Treatment of Cancer have classified LyP among the indolent (slow growing) cutaneous T-cell lymphomas, other experts do not consider the skin condition a true malignancy, because of its benign clinical course. The condition is extremely rare.

### How Anaplastic Large Cell Lymphoma Is Diagnosed and Staged

As with other types of NHL, diagnosing anaplastic large cell lymphoma requires taking a small sample of the tumor tissue, called a biopsy, and looking at the cells under a microscope. Anaplastic large cell lymphoma has a unique appearance under the microscope characterized by cells of different shapes and sizes and the uniform expression of a special marker on the lymphoma cells called CD30. Once a diagnosis has been made, a series of other diagnostic tests, such as blood, CT (computerized axial tomography), MRI (magnetic resonance imaging) and PET (positron emission tomography) scans and a bone marrow biopsy, will be done to determine the extent, or stage, of the disease.

The four stages of ALCL include:

- Stage I (early disease)—The cancer is located in a single lymph node region or in one organ or area outside the lymph node.
- Stage II (locally advanced disease)—The cancer is found in two or more lymph node regions on the same side of the diaphragm (the breathing muscle that separates the abdomen from the chest).
- Stage III (advanced disease)—Disease is found in lymph nodes both above and below the diaphragm.

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- Stage IV (widespread disease)—The lymphoma has spread beyond the lymph nodes and spleen to one or more organs such as the bone, bone marrow, skin or liver.

It is important to remember that being diagnosed with advanced or widespread ALCL is common and, unlike a stage III or IV diagnosis in solid tumor cancers in which the primary cancer has spread to other sites, advanced stages of NHL can be successfully treated and may have a favorable outcome.

### How Anaplastic Large Cell Lymphoma Is Treated

Although standard chemotherapy, such as CHOP (cyclophosphamide, doxorubicin, vincristine and prednisone), is given to newly diagnosed ALCL patients, disease outcome is usually very different depending on the subtype. The ALK positive type responds extremely well to CHOP, rendering over a 70 percent long-term disease-free survival, whereas the ALK negative type has a poorer prognosis with 45 percent to 50 percent achieving long-term disease-free survival. Higher doses of chemotherapy is usually prescribed for relapsed patients, followed by a stem cell transplant once remission is achieved. Primary cutaneous ALCL is usually treated with radiation therapy if the disease is localized and with systemic chemotherapy if it's in multiple areas of the skin.

### Treatments Under Investigation

Because ALCL is such a rare disease, finding enough patients to enroll in clinical trials is difficult. However, there are several new drugs now in clinical trials that are showing promising results, including:

- SGN30, an anti-CD30 monoclonal antibody
- Pralatrexate
- PEGS (cisplatin, etoposide, solimedeol and gemcitabine) chemotherapy
- CHOP chemotherapy, plus bevacizumab (Avastin)

### Complementary and Alternative Therapies

Complementary and alternative medicines are nonstandard therapies that may help patients cope with their cancer and its treatment, but that should not be used in place of standard treatment. No alternative therapy has ever been proven effective against lymphoma. However, complementary therapies such as meditation, yoga, acupuncture, exercise, diet and relaxation techniques have been shown to be effective in combating some treatment side effects. Before embarking on any complementary therapies, patients should discuss the matter with their healthcare team. Certain unproven treatments, including some herbal supplements, can interfere with standard lymphoma treatments or may cause serious side effects.

### How to Prepare for Follow-Up Appointments

It is important for patients both during and after treatment to be proactive in their healthcare, including keeping a master file of medical records, asking questions, reporting new symptoms, exercising and eating a balanced diet. In addition, patients who smoke should strongly consider stopping. Follow-up visits, usually scheduled every few months, typically include physical examinations, blood tests and occasionally CT scans. Since lymphoma symptoms may resemble those of less serious illnesses, like colds or viral infections, maintaining regular medical care is imperative. Besides looking for signs of a recurrence of cancer, follow-up care can help identify and resolve unusual side effects of treatment.