

Anaplastic Large Cell Lymphoma (ALCL)



OVERVIEW

Lymphoma is the 6th most common cancer in Australia in adult men and women. It can affect people of all ages and is the most common blood cancer. Lymphoma is a cancer of the immune system and affects lymphocytes which are a type of white blood cell. When lymphocytes gain DNA mutations they divide and grow uncontrollably resulting in lymphoma.

There are two main types of lymphocytes called B lymphocytes (B-cells) and T lymphocytes (T-cells). Lymphomas caused by B-cells are more common and account for around 85% of lymphoma cases and lymphomas caused by T-cells account for around 15% of lymphoma cases. The first lymphoma to be discovered was called "Hodgkin lymphoma" (around 15% of all B-cell lymphomas), after Thomas Hodgkin, who described it. All subsequent lymphomas discovered were called "non-Hodgkin lymphoma" (around 90% of all lymphomas, both B-cell & T-cell lymphomas).

There are over 80 different subtypes of lymphoma, that are classified according to its clinical behaviour. "Aggressive" (high grade or fast growing) lymphomas are those that grow quickly, usually weeks to months and need treatment immediately. "Indolent" (low grade or slow growing) lymphomas usually develop over years and often are not treated straight away but are monitored. It is important to know your subtype of lymphoma. Lymphoma cells can travel to any part of the body and be found in lymph nodes, the bone marrow, the spleen, blood, bone, skin and almost any organ or tissue.

Anaplastic large cell lymphoma (ALCL) is a rare subtype of T-cell lymphoma. Anaplastic refers to the appearance of the lymphoma cells, that look quite different from normal lymphocytes. The presence of the protein, 'CD30' on the surface of lymphoma cells is the hallmark of the disease. ALCL can occur in two different forms depending on which parts of the body are involved.

TYPES OF ALCL

The two types of ALCL are:

Primary Cutaneous – which presents and is usually confined to the skin, follows a less aggressive course. It is usually

managed as an indolent (slow-growing) lymphoma as long as it is confined to the skin. If the primary cutaneous ALCL extends beyond the skin to the lymph nodes or organs it is usually managed as a systemic ALCL. This occurs in about 10 percent of people diagnosed with primary cutaneous ALCL. Patients with primary cutaneous ALCL usually have a single lump or ulcerating tumour in the skin. Lymph nodes in the area may also become involved. People may experience spontaneous remissions with this disease. However, the remissions are inevitably followed by relapses (the lymphoma returns). Primary cutaneous ALCL most commonly presents in people aged between 50 and 60 and is rare in children. It is 2-3 times more common in men than in women. There is a strong link between cutaneous ALCL and other forms of T-cell lymphoma such as mycosis fungoides.

Systemic ALCL – can affect all organs in the body and is aggressive. Patients with systemic ALCL often present with painless enlarged lymph nodes in the neck, armpit or groin. Other parts of the body can be affected including bones, skin, bone marrow, the lungs and the liver. Night sweats, fevers and unexplained weight loss may also be present. In more than half of people with systemic ALCL, there will be elevated blood levels of the enzyme lactate dehydrogenase (LDH).

There are two subtypes: ALK-negative and ALK-positive, depending on whether the lymphoma cells produce a protein called 'anaplastic large cell kinase' (ALK). ALK-positive ALCL is more likely to affect children and young adults (median age 34), although there is a group who present later in life. People with ALK-negative ALCL present at a later age (median age 58 years). Systemic ALCL is slightly more common in men than in women.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of ALCL. A biopsy is an operation to remove a lymph node or other abnormal tissue to look at it under the microscope. The biopsy can be done under local or general anaesthetic depending on what part of the body is being biopsied.

Once a diagnosis of ALCL is made, further tests are needed to be performed to see where else in the body the lymphoma is located (or the "stage"). Examples of staging tests include;

FACT SHEET

- Positron emission tomography (PET) scan
- Computed tomography (CT) scan
- Bone marrow biopsy

Patients may also undergo a number of baseline tests prior to any treatment commencing to check their organ function and these baseline tests may include a heart scan, kidney scan, breathing tests and blood tests.

TREATMENT OPTIONS

The two types of ALCL are treated differently.

Systemic ALCL treatments are similar to regimens appropriate for other aggressive lymphomas, such as diffuse large B-cell lymphoma (DLBCL). With combination chemotherapy, many patients with ALCL will get a lasting remission. People with ALK-positive ALCL generally respond well to chemotherapy.

- CHEOP or CHOP (combinations of cyclophosphamide, doxorubicin, etoposide, vincristine and prednisolone)
- Radiotherapy
- Stem cell transplant
- steroid therapy

Primary cutaneous ALCL may go into a spontaneous remission (the disease goes away without treatment), although this is usually followed by a relapse (the disease comes back). If no spontaneous remission occurs or the disease relapses the most common treatments include:

- Radiotherapy or surgery to remove the area of the skin affected
- Systemic chemotherapy is only used in people who have extensive involvement that cannot be treated with localized therapies

TREATMENTS UNDER INVESTIGATION

Clinical trials for ALCL is challenging, as it is a rare disease and therefore finding enough people diagnosed to enrol can be difficult. There are several new drugs that are showing some promise in clinical trials around the world including:

- brentuximab vedotin (Adcetris™) is approved for patients with relapsed/refractory ALCL
- Pralatrexate (Folotyn™)

CLINICAL TRIALS

Clinical trials are essential in identifying effective medicines and determining optimal doses of these medicines for people diagnosed with lymphoma. People who are interested

in participating in a clinical trial can find one using the following methods:

1. Speak to their specialist to see what options are available
2. Go to the ClinTrial Refer website www.clintrial.org.au to search available clinical trials
3. See 'Understanding Clinical Trials' fact sheet

Download the ClinTrial Refer app from the Apple or Android stores for your smart phone or device. The ClinTrial Refer service was developed to connect patients, health professionals and clinical trial sites to improve access to clinical trials for patients in Australia.

FOLLOW UP

Once treatment is completed, people with lymphoma need to be followed up by their specialist with regular appointments to monitor:

- Evaluate the effectiveness of the treatment
- Ongoing treatment side effects
- Recovery from treatment
- Signs of the lymphoma relapsing
- Potential late effects caused by treatment that can occur months or years later, that can vary based on the duration and frequency of treatment, age, gender and overall health of each person.

RESOURCES AND SUPPORT

Lymphoma Australia offer a wide variety of resources and support for people with lymphoma and their carers. Please visit our website www.lymphoma.org.au for further information.

SOME QUESTIONS TO ASK YOUR DOCTOR

- What type of ALCL do I have?
- What are the treatment options for my type of ALCL?
- Are there any treatment options that are better for my type of lymphoma but are yet to be funded by the PBS in Australia?
- Are there any clinical trials currently available to me?
- If you think my ALCL has relapsed, will you do another tissue biopsy to confirm this?