FACT SHEET

Angioimmunoblastic T-Cell Lymphoma (AITL)

OVERVIEW

Lymphoma is the 6th most common cancer in Australia 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 effects lymphocytes which are a type of white blood cell. When lymphocytes gain certain genetic changes, 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 and 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.

Angioimmunoblastic T-cell lymphoma (AITL) is accounts for around 20 per cent of cases of T-cell lymphoma. AITL is often considered a fast-growing lymphoma that requires more urgent treatment, but some patients have a form of disease that is slower to grow. AITL is more common in older people but can affect younger adults as well.

Symptoms of AITL include high fevers, night sweats, skin rash, loss of more than 10 per cent of your body weight without trying, also known as B symptoms. It can also be associated with autoimmune disorders such as autoimmune haemolytic anaemia, where you have fewer red cells circulating in your blood stream and immune thrombocytopenia where you have less platelets circulating in your blood stream. The skin rash that can occur with AITL is often non-specific and can result in papules, nodules, plaques, ulcers, petechiae and rarely erythroderma.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of AITL. A biopsy is a surgical procedure to remove part of or all of an affected lymph node or other abnormal tissue to look at it under the microscope in the laboratory to see what the cells look like. The biopsy can be done under local or general anaesthetic depending on what part of the body is being performed on. Sometimes the diagnosis will be made on a bone marrow biopsy alone.

Once a diagnosis of AITL is made there are further tests that need to be performed to see where else in the body the lymphoma may be and is referred to as staging. Because AITL is a blood cancer the lymphoma can travel all over the body, so it is important that a check of the entire body is done looking for the lymphoma. Staging tests may include:

- Positron emission tomography (PET) /CT scan
- Computed tomography (CT) scan
- Bone marrow biopsy
- Skin biopsy
- Lumbar puncture (if lymphoma suspected in the brain or spinal cord)

The majority of patients with AITL are diagnosed with advanced stage 3 or 4 lymphoma. Patients will 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 and blood tests.

TREATMENT OPTIONS

AITL is a fast-growing lymphoma and therefore needs to be treated urgently. A combination of chemotherapy is used to treat AITL. Some standard treatments for AITL include.

- CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone)
- CHOEP (cyclophosphamide, doxorubicin, vincristine, etoposide and prednisolone)
- Participation in a clinical trial

Due to the increased chance of this lymphoma relapsing the person may also be recommended to undergo an autologous stem cell transplant to consolidate the response of the initial chemotherapy and to increase their chances of the lymphoma never returning.

If the lymphoma does return, second line treatment may include:

- Pralatrexate (Folotyn™)
- Romidepsin (Istodax™) (not currently funded by the PBS)
- Combination chemotherapy
- Autologous stem cell transplant (own cells)
- Allogeneic stem cell transplant (donor cells)
- Radiotherapy
- Participation in a clinical trial

TREATMENTS UNDER INVESTIGATION

Many new individual and combination medicines are currently being tested in clinical trials around the world for both newly diagnosed and relapsed/refractory AITL including:

- Alemtuzumab (Campath™)
- Belinostat (Beleodaq™)
- Brentuximab vedotin (Adcetris™)
- Lenalidomide (Revlimid™)
- Azacitadine
- Chimeric antigen receptor (CAR) T-cell therapy

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. See 'Understanding Clinical Trials' fact sheet for websites to find a clinical trial; https://bit.ly/2V8fNjf

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 lymphoma relapsing
- Potential late effects caused by treatment that can occur months or years later, that can be based on the duration and frequency of treatment, age, gender and overall health of each person

RESOURCES AND SUPPORT

Organisation	How can they help?

Lymphoma Australia	• Lymphoma Australia offers a wide variety of resources and support for people with lymphoma or CLL and their caregivers. Please visit our website lymphoma.org.au for further information:
	• Lymphoma Australia Fact sheets & booklets including:
	  • Booklet: What you need to know about lymphoma
	  • Peripheral T-cell lymphoma
	  • Understanding Clinical Trials
	  • Emotional impact of a lymphoma diagnosis & treatment
	• lymphoma.org.au/page/1218/fact-sheets
	• Lymphoma Australia YouTube Channel: Presentations and interviews on a variety of topics about lymphoma subtypes, management and supportive care.
	• youtube.com/user/LymphomaAustralia
	• Lymphoma Nurse Support Line: 1800 953 081 or email: nurse@lymphoma.org.au
	• Online private Facebook group: **Lymphoma Down Under** http://bit.ly/2mrPA1k

SOME QUESTIONS TO ASK YOUR DOCTOR

- What stage of AITL do I have?
- Is there any additional testing that can be done to give you a greater insight into how to treat my type of lymphoma?
- What are my treatment options for AITL?
- 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?
- Am I a candidate to have a stem cell transplant in first remission or are there any other maintenance treatment options for my AITL?

This resource was last reviewed and updated November 2020