

Cutaneous B-Cell Lymphoma (CBCL)



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 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 & 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.

Lymphomas that arise in tissues or organs outside of the lymphatic system are called extra nodal lymphomas. When extra nodal lymphomas originate in the skin and there is no evidence of lymphoma outside of the skin, they are called primary cutaneous lymphomas. Primary cutaneous B-cell lymphomas (CBCL) occur when the lymphoma cells originate from B lymphocytes.

TYPES OF CBCL

Primary Cutaneous Follicle Cell Lymphoma: This is the most common type of CBCL, and these lymphomas are typically indolent developing slowly over months to many years. They usually appear on the head, neck, or torso of the body as a red-

brown bumpy skin eruption, nodule or tumour.

Primary Cutaneous Marginal Zone B-cell Lymphoma: This is the second most common type of CBCL, and these lymphomas can have a similar appearance to follicle centre lymphoma. They usually appear on the torso or arms as a pink-red bumpy skin eruption, nodule or tumour.

Primary Cutaneous Diffuse Large B-cell Lymphoma Leg Type: This is less common than other CBCL but is usually more aggressive and fast growing developing over weeks to months. This lymphoma usually appears as solitary or multiple tumours on the legs but can also involve non leg areas such as arms and torso. The lesions may even ulcerate and spread outside of the skin more frequently than slow growing CBCL.

Primary Cutaneous Diffuse Large B-cell Lymphoma - Other: This is a very rare group of CBCLs including intravascular large B cell lymphoma, T-cell rich B-cell lymphoma, plasmablastic lymphoma, and anaplastic B cell lymphoma. These lymphomas usually appear on the head, torso and extremities.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of CBCL. A biopsy is a procedure to remove a part the 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 CBCL 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 as staging. Because CBCL 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:

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

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, breathing test and blood tests.

TREATMENT OPTIONS

Treatment options for CBCL depends on the type of CBCL and whether the skin lesion is solitary or multifocal (wider spread). For the rare aggressive CBCL the patient's overall health and condition are crucial to assess prior to deciding upon treatment and with the more common indolent CBCL although managed in a similar fashion there is more room to move with treatments as they are generally tolerated better.

Primary cutaneous follicle centre lymphoma and primary cutaneous marginal zone B-cell lymphoma are treated very similarly and if there is a solitary lesion the most common treatment is local radiotherapy. Some of these slow growing lymphomas can be managed by the watch and wait approach where immediate treatment is not required where patients are closely monitored through regular checkup visits with their medical team and laboratory and imaging tests.

Symptomatic lesions can be treated by radiotherapy, intralesional steroids, topical therapies or even surgical treatment can be considered. If lesions are widespread, systemic therapies such as the monoclonal antibody rituximab that targets B-cells in the body may be appropriate. Regular skin examinations are very important especially for the slow growing lymphomas as the skin is the most common site for the lymphoma to appear. General laboratory tests will also be followed although not all patients require imaging tests unless there is a concern of the lymphoma being widespread.

For aggressive types of CBCL such as diffuse large B-cell Lymphoma leg type, systemic chemotherapy with the monoclonal antibody rituximab is required. Baseline imaging is performed prior to this treatment starting as well as during treatment to check effectiveness and at the end of treatment. Some patients will also go on to have radiotherapy after the chemotherapy treatment to decrease the risk of the lymphoma returning.

The details of your treatment will vary depending on the stage of the CBCL and general fitness. The side effects from treatment varies between the different types of treatment. CBCL usually responds well to treatment but in some people the lymphoma comes back (relapses) and further treatment is needed. Treatments available for relapsed CBCL can vary and include:

- Combination chemotherapy
- Rituximab
- Targeted therapies
- Radiotherapy
- Radioimmunotherapy

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 on the website to find out more. www.lymphoma.org.au

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

RESOURCES AND SUPPORT

Lymphoma Australia offers 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.

Cutaneous Lymphoma Foundation (USA), promote awareness and education for those affected. For more information please visit their website www.clfoundation.org

SOME QUESTIONS TO ASK YOUR DOCTOR

- What type of CBCL do I have?
- Is there any additional testing that can be done to give you greater insight into how to treat my type of lymphoma?
- What are the treatment options for my type of CBCL?
- 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 CBCL has relapsed, will you do another tissue biopsy to confirm this and what treatment options do I have?