FACT SHEET

Burkitt Lymphoma (BL)

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 effects 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 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.

Burkitt lymphoma (BL) is a rare but especially aggressive B-cell lymphoma. The disease commonly presents as swollen lymph nodes and abdominal swelling. Other common symptoms include night sweats, unexplained weight loss and fevers. BL accounts for approximately 1 - 2% of adult lymphomas, but up to 30% of childhood lymphoma globally. It is more common in men than in women. The average age of adults diagnosed with BL is between the ages of 30 and 50 years, whilst children diagnosed are between 5 and 10 years of age.

TYPES OF BURKITT LYMPHOMA

There are three main types of BL:

- **Endemic (African):** is the most common lymphoma of children in Africa and is associated with chronic malaria and Epstein-Barr virus (EBV). It particularly affects the jaw and other facial bones but can also occur in the abdomen, ovaries, kidney or breast. Endemic BL is rare outside of Africa.
- **Sporadic (Non-African):** occurs throughout the world and it accounts for 1 - 2% of adult lymphoma cases. In the sporadic BL, Epstein-Barr virus occurs in about 20% of the patients and the abdomen is the most common site of disease occurrence where it causes swelling and discomfort. BL may spread to the central nervous system (i.e., brain and spinal cord). Other organs and tissue may be affected such as the thyroid and tonsils, and the facial bones may also be involved.
- **Immunodeficiency-associated:** is most common in people with human immunodeficiency virus/ acquired immunodeficiency syndrome (HIV/AIDS). Immunodeficiency-related BL can also occur in patients who have inherited immune deficiencies or those who take immunosuppressive medications to prevent rejection after organ transplant.

DIAGNOSIS AND STAGING

A biopsy is always required for a diagnosis of BL. 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. The biopsy can be done under local or general anaesthetic depending on what part of the body is being biopsied.

A blood test may also be done to measure the white blood cell count and certain proteins, that helps to diagnose BL. A hallmark of BL is a gene called the MYC gene. Rearrangements of the chromosome at the site that harbours MYC are required for a diagnosis of BL. Abnormalities in this gene are found in other high-grade mature B-cell lymphomas as well.

The distinction between BL and other high-grade B-cell lymphomas (such as Diffuse Large B-cell Lymphoma, DLBCL)

also depends on the pathologist's opinion based the appearance of the biopsy under the microscope. Accurately diagnosing BL is critical because BL and DLBCL are treated differently.

Once a diagnosis of BL is made, further tests are needed to be performed to see where else in the body the lymphoma may be (stage I-IV). Because BL is a blood cancer it can be found anywhere in the body. Staging tests include:

- Positron emission tomography (PET) /CT scan
- Computed tomography (CT) scan – that may not be required if a PET scan was performed
- Bone marrow biopsy
- Lumbar puncture (if lymphoma is suspected in the brain or spinal cord)
- Other baseline tests done prior to treatment starting to check organ function: heart scan, kidney scan, respiratory tests and blood tests

TREATMENT OPTIONS

BL is a highly aggressive disease and it is often very responsive to the standard intensive combination chemotherapy regimens. Treatment however does needs to start very soon after diagnosis. Central nervous system (CNS) at diagnosis is recognised to be a strong risk factor for relapse (disease returns) so it is recommended that treatment regimens include treatment to protect the CNS (intrathecal: chemotherapy injected into the spinal fluid). The most common chemotherapy treatment for adults with BL include:

- CODOX-M: (cyclophosphamide, vincristine, doxorubicin, intrathecal cytarabine, methotrexate and rituximab) – **alternating with cycles of IVAC**
- IVAC: (ifosphamide, etoposide, cytarabine and intrathecal methotrexate) **OR**
- DA-EPOCH-R (dose adjusted etoposide, prednisolone, vincristine, cyclophosphamide, doxorubicin, intrathecal methotrexate and rituximab)

Different combination chemotherapy regimens are used to treat BL in children and adolescents. BL is one of the most common types of childhood lymphoma and younger patients tend to have excellent responses to chemotherapy and particularly high cure rates. For this reason, the current trend in the treatment of children is focused on decreasing toxicity by reducing the overall amount of chemotherapy used to treat the disease.

A high percentage of people achieve remission of their BL with standard treatment. However, for some people, the lymphoma returns (relapses) or in the rare cases does not respond to initial treatment (refractory). People with relapsed or refractory BL may have other treatment options including other chemotherapy combinations, biologic medicines or participation in a clinical trial.

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, 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
- 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

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 BL do I have? What is my stage?
- Do I have "High risk or low risk" Burkitt lymphoma?
- Do I require lumbar punctures with my treatment?
- What are the treatment options for my type of BL

This resource was last reviewed and updated July 2019
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