

Cutaneous T-Cell Lymphoma (CTCL) – Advanced Stage



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).

Cutaneous T-cell Lymphoma (CTCL) is a rare form of lymphoma that involves the skin and this fact sheet will refer to advanced stage disease. The mutated T-cells migrate to the skin, often first presenting as a rash which can be itchy and hard to diagnose. The skin lesions can be varied and most skin lymphoma is indolent and stays localised to the skin. It is usually not aggressive and does not usually need to be treated aggressively. Most commonly patients have red scaling flat patches. Sometimes these become raised to form plaques or even large lumps that are referred to as tumours. Some patients can even be red all over and this is called erythroderma. These lymphomas are more common in men than women and more common in older patients but can also affect children. There are many rare subtypes of CTCL, so we have described a few of these in further detail below.

TYPES OF CTCL

Mycosis Fungoides (MF) is the most common subtype of CTCL, accounting for over 50% of all cases. MF is generally an indolent lymphoma that is often unpredictable (over years to decades). MF starts in the skin and most remain only in the skin and can appear as patches, plaques or tumours (raised bumps which may or may not ulcerate). It can mimic many common skin conditions such as eczema or psoriasis. It often occurs in areas of the skin that are protected from sun by clothing.

Uncommonly mycosis fungoides can progress to lymph nodes, blood and internal organs. Most patients live normal lives and have a normal life span with this lymphoma. Treatments are tailored to meet an individual's specific needs. Most patients just require topical therapies such as cortisone ointment or ultraviolet light therapy. Some patients need low dose radiation treatment. When the disease is more problematic and/or extensive then systemic therapies may be needed. The first line systemic therapies are of low toxicity and are used in many other diseases. One example is low dose methotrexate that is also used in benign conditions such as psoriasis and rheumatoid arthritis.

Sezary Syndrome (SS) is a rare subtype of CTCL, second to MF, that affects both the skin and circulating blood. Patients with this condition present with extensive itchy red skin that covers 80% or more of their body. Diagnosis is often difficult and delayed. Treatments are designed to normalise the immune system. Three main modalities are used in this condition including extracorporeal photopheresis, interferon and total skin electron beam therapy.

Primary Cutaneous Anaplastic Large-Cell Lymphoma (PCALCL) is a rare subtype of CTCL that generally only affects the skin, although in rare cases can spread to other organs. This condition behaves differently to the more aggressive systemic type of anaplastic large cell lymphoma. PCALCL is usually an indolent lymphoma that can affect people of all ages, including children but is more common in 45-60 years old. The condition is characterised by the appearance of solitary or uncommonly multiple raised red skin lesions, nodules or tumours that do not go away and can ulcerate.

Subcutaneous Panniculitis-like T-cell Lymphoma (SPTCL) is rare and is characterised by nodules that involve the subcutaneous fat. This CTCL more commonly presents in young adults. It responds well to steroid tablets and other forms of immune system modulation. There are many other subtypes of CTCL that are best dealt with by experienced specialists.

DIAGNOSIS AND STAGING

Diagnosis requires careful correlation of clinical and pathological findings by experienced practitioners. Several major hospitals have multidisciplinary clinics that employ the skills of a diverse

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range of practitioners including dermatologists, haematologists, radiotherapy oncologists, pathologists and specialist nurse practitioners. It is critical that an accurate diagnosis be made so that the patient can receive appropriate therapy.

Once a diagnosis of CTCL is made, further tests may be needed to be performed to see where else in the body the lymphoma is located. Most patients with CTCL do not require extensive testing. Clinical and physical examination combined with biopsy results will often suffice. If a patient is suspected to have or is in a higher risk for systemic disease, then other investigations may be ordered including;

- Physical examination and photography of the skin
- Positron emission tomography (PET) scan
- Computed tomography (CT) scan
- Bone marrow biopsy
- Blood tests

TREATMENT OPTIONS

CTCL can often be brought under good control with the current available therapies. Most cases are considered incurable but even though they can be chronic conditions patients can live an otherwise normal life.

The prognosis depends on the type of CTCL, your age, general health and how much of your skin is affected. Your specialist team will educate you on your specific condition and its implications. People can live normal lives with CTCL, however symptoms can become debilitating and painful and the skin involvement can cause body image issues which need to be managed. The more aggressive forms of CTCL need more intensive treatment sometimes more urgently. There are a range of treatment options available for people diagnosed with these more aggressive lymphomas including clinical trial therapies. Psychological support is also an important facet of management.

Most patients learn to manage their condition together with their specialist team. They generally use topical treatments in addition to systemic treatments if they require them. Some patients will use baths or other agents to reduce the infective load on their skin such as bleach baths and some patients require wound care based on what their skin is doing at the time. Skin infections and skin pain are things that often need managing. As a general principle chemotherapy is reserved for late stage refractory disease. There are several non-chemotherapy treatments that are effective in CTCL.

There are a range of treatment options available for people diagnosed with these lymphomas including;

- Alemtuzumab (Campath™)

- Bexarotene (Targretin™) gel and capsules
- Brentuximab Vedotin (Acedtris™)
- Carmustine (BCNU™)
- CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine & prednisolone)
- Corticosteroids (topical and oral)
- Extracorporeal Photopheresis
- Gemcitabine (Gemzar™)
- Interferons
- Methotrexate (Methoblastin™)
- Pralatrexate (Folotyne™)
- Phototherapy
- Radiotherapy
- Rituximab (MabThera™)
- Romidepsin (Istodax™)
- Stem cell transplant (autologous or allogenic)
- Vorinostat (Zolinza™)

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 CTCL including;

- Anti CD47 monoclonal antibodies
- Carfilzomib (Kyprolis™)
- Lenalidomide (Revlimid™)
- Mogamulizumab (Poteligeo™)

CLINICAL TRIALS

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

1. Speak to their specialist to see what options are available
2. For more information see "Understanding Clinical Trials" fact sheet: <http://bit.ly/2B83sBR>

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.
- See "Cutaneous T-cell Lymphoma" presentation from Dr Carrie Van Der Weyden, Peter Mac in Lymphoma Australia YouTube: <http://bit.ly/2VBObIX>
- Cutaneous Lymphoma Foundation (USA), promote awareness and education for those affected. For more information please visit their website www.clfoundation.org

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