



THE AUSTRALASIAN COLLEGE
OF DERMATOLOGISTS

Primary Cutaneous B-Cell Lymphoma (PCBCL)

Also known as ... Cutaneous B-cell Lymphoma

What is primary cutaneous B-cell lymphoma (PCBCL)?

Tumours of the lymph nodes and lymphatic system are called “lymphomas”. B-cell lymphomas are caused by an excess of B-cell lymphocytes, a type of white blood cell. Primary cutaneous B-cell lymphomas (PCBCL) are a group of B-cell lymphomas that are localised to the skin, without evidence of disease beyond the skin at the time of diagnosis. PCBCL most commonly affects people over the age of 55 years.

There are several types of PCBCL:

- Primary cutaneous marginal zone lymphoma (PCMZL)
- Primary cutaneous follicle centre lymphoma (PCFCL)
- Primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT)
- Primary cutaneous diffuse large B-cell lymphoma, other (PCDLBCL-other)
- Primary cutaneous intravascular large B-cell lymphoma

PCDLBCL and other primary cutaneous intravascular large B-cell lymphoma are very rare and hence will not be discussed further.

What causes PCBCL?

The cause of PCBCL is largely unknown. Previous infection with *Borrelia burgdorferi* which causes Lyme disease has been associated with some cases of PCMZL. B-cell lymphomas comprise 20-25% of primary skin lymphomas.

What does PCBCL look like?

- **Primary cutaneous marginal zone lymphoma (PCMZL)**

PCMZL presents as red to purple-coloured lumps and bumps on the skin. The most common sites affected are the trunk, arms or head. The lesions may be single or multiple and usually cause no symptoms.

- **Primary cutaneous follicle centre lymphoma (PCFCL)**

PCFCL occur most commonly on the head, neck and trunk. Affected areas are reddish purple and raised.

- **Primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT)**

PCLBCL-LT presents with lumps and bumpy areas on one or both legs. Lesions are red to purple/blue in colour. Despite the name, this type of lymphoma can occur at other sites in 15% of cases.



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How are PCBLs diagnosed?

To make the diagnosis of PCBCL the following investigations are required:

- Skin biopsy – this will be sent to a pathologist for examination. If positive, a dense collection of abnormal appearing B-cells will be seen. The pathologist then performs special stains on the tissue (immunohistochemistry) to help identify which type of B-cell lymphoma is present.
- Investigations to exclude involvement in other organs:
 - Blood tests
 - Radiological tests (CT scan, PET scan)
 - Lymph node biopsy (if any lymph nodes are felt on examination)
 - A bone marrow biopsy is generally recommended for individuals with DLBCL and PCMZL, as well as individuals with more widespread PCFCL, and would be done under the guidance of a haematologist.
- Borrelia serology – Individuals with PCMZL may be investigated for Lyme disease if they have been to a geographical region where exposure is possible.

How are PCBCLs treated?

The method of treatment depends on the type of PCBCL present. Most cases would involve the expertise of a dermatologist and a haematologist in a specialised centre.

For PCFCL and PCMZL which are indolent (low-grade) lymphomas, management options include:

- Observation without treatment if no symptoms
- Radiotherapy – the most frequently used treatment method
- Surgical excision (removal)
- Corticosteroid injections into the areas affected
- Rituximab (anti-CD20 antibody) injections
- Oral or injected medications for more generalised disease such as rituximab, interferon, chlorambucil

For PCDLBCL-LT, which is a more aggressive (intermediate-grade) type of lymphoma, management options include:

- Radiotherapy
- Rituximab
- Chemotherapy R-CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone with rituximab).

What is the likely outcome of PCBCL?

The prognosis of PCBCL depends on the type of lymphoma diagnosed.

- **Primary Cutaneous Marginal Zone Lymphoma (PCMZL)**

This is an indolent (low-grade) lymphoma with an excellent prognosis. The 5-year survival rate is 99%. Spontaneous resolution (recovery) can occur. Following treatment recurrence (new skin lesions) is common (60%+). This does not imply a poorer prognosis.

- **Primary Cutaneous Follicle Centre Lymphoma (PCFCL)**

This is an indolent (low-grade) lymphoma with a very good prognosis. The 5-year survival rate is 95%. Spontaneous resolution (recovery) can occur. Recurrence is common following treatment (60%+) but does not imply a worse prognosis.

- **Primary Cutaneous Diffuse Large B cell lymphoma, leg-type (PCDLBCL-LT)**

This is a more aggressive (intermediate grade) lymphoma with a poorer prognosis and high risk of spread to other organs. The 5-year survival rate is less than 50%.

This information has been written by Dr Kate Newland, Dr Olivia Milne, Associate Professor Chris McCormack and Professor Miles Prince