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## Cutaneous B-cell lymphoma

### What are cutaneous B-cell lymphomas?

Lymphomas are tumours of the lymph nodes and lymphatic system. Extranodal lymphomas are tumours that occur in organs or tissues outside of the lymphatic system. When lymphomas occur in the skin with no evidence of extracutaneous disease at the time of diagnosis, they are called 'primary' cutaneous lymphomas. There are many different types of primary cutaneous lymphomas but they can be broadly divided into two categories, [cutaneous T-cell lymphomas](#) and B-cell lymphomas. Primary cutaneous lymphomas of B cell type comprise approximately 20% of cutaneous lymphomas.

Cutaneous B-cell lymphomas occur when there is a malignant proliferation of lymphocytes of the B-cell type. Mutation occurring at different points in B cell development leads to differing forms of lymphoma.

#### Cutaneous B-cell lymphoma



FC lymphoma

### Classification of cutaneous B-cell lymphomas

Recently the World Health Organisation (WHO) and European Organization for Research and Treatment of Cancer Classification (EORTC) reached a consensus classification for cutaneous lymphomas<sup>1</sup>. Primary cutaneous B-cell lymphomas include:

- Primary cutaneous follicle centre lymphoma
- Primary cutaneous marginal zone B- cell lymphoma
- Primary cutaneous diffuse large B-cell lymphoma, leg type
- Primary cutaneous diffuse large B-cell lymphoma, other

### Primary cutaneous follicle centre (FC) lymphoma

These skin lymphomas typically present as solitary or grouped nodules, tumours, or infiltrative plaques. They occur most commonly in the scalp, forehead, neck, trunk and back. The tumours have an indolent (slow growing) course but progress with time. Neoplastic follicle centre cells, usually a mixture of centrocytes (small and large cleaved follicle centre cells) and centroblasts (large noncleaved follicle centre cells with prominent nucleoli) are seen in the dermis in at least part of the infiltrate, but usually spare the overlying epidermis. Cellular morphology

may vary with the age and size of the lesion, with care required to differentiate this group from Primary cutaneous diffuse large B-cell lymphoma, leg type.

Primary cutaneous FC lymphoma needs to be differentiated from a secondary cutaneous lymphoma, which is where a nodal follicular lymphoma has spread to involve the skin ([skin metastasis](#)). It appears from molecular biology studies that Primary cutaneous FC lymphoma usually lack expression of the bcl-2 protein, and, the t(14;18) translocation, which if present may suggest systemic follicular lymphoma.

Primary cutaneous FC lymphomas may be treated with [radiotherapy](#) or combination chemotherapy. Radiotherapy is the treatment of choice for localised tumours whereas multiple tumours on the skin of different parts of the body (multifocal) may respond better to combination chemotherapy. Patients with primary cutaneous FC lymphomas have a good overall survival rate. Five-year survival is reported to be more than 95%.

### **Primary cutaneous marginal zone B-cell lymphoma**

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These are low-grade malignant B-cell lymphomas of the MALT (mucosa-associated lymphoid tissue) type, that occur in the skin as solitary or multiple red to brownish red papules, plaques, or nodules. They are usually located on the trunk or extremities, especially the arms. Histologically, these lymphomas are characterised by the presence of nodular or diffuse infiltrates with small lymphocytes, marginal zone B cells, lymphoplasmacytoid cells and plasma cells localised in the dermis and subcutaneous fat. This group also includes primary cutaneous immunocytomas.

Primary cutaneous immunocytoma and marginal zone B-cell lymphoma are classed as indolent lymphomas with excellent prognosis. Five-year survival rate is close to 100%. Dissemination to extracutaneous sites is very rare, however there are reports of it spreading to other organs and the bone marrow.

Radiotherapy or surgical excision is effective treatment for patients with a solitary or few lesions. Multifocal skin lesions are treated with chemotherapy agents such as chlorambucil, interferon alpha and anti-CD20 antibody (rituximab). As this is an indolent lymphoma, observation is a reasonable approach in some cases.

### **Primary cutaneous large B-cell lymphoma, leg type**

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These cutaneous B-cell lymphomas occur mainly in elderly females and present as erythematous red or bluish-red nodules or tumours usually on one or both lower legs. These tumours are more aggressive than large B-cell lymphomas of the head and neck, often disseminate to extracutaneous sites and have a more unfavourable prognosis.

Histologically these lymphomas show a diffuse monotonous population of centroblasts and immunoblasts. This picture may be seen in lesions not arising on the leg, and these can be classified into this group.

Only some localised small tumours may be treated using radiotherapy. Anthracycline-based chemotherapy is used for most skin tumours or when tumours have spread to other parts of the body. Some cases have shown response to the anti-CD20 antibody rituximab. Death usually occurs with disseminated disease.

### **Primary cutaneous diffuse large B-cell lymphoma, other**

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This group contains large B-cell lymphomas arising in the skin, which do not belong to the primary cutaneous follicle centre lymphoma group or large B-cell lymphoma, leg type. These lymphomas commonly present with skin lesions on the head, the trunk or the extremities and have an excellent prognosis.

Intravascular large B-cell lymphoma is a well-defined subtype of large B-cell lymphoma. Malignant B lymphoid cells proliferate within the lumen of small blood vessels, primarily in the skin and central nervous system. Lesions appear as erythematous, tender nodules, tumours, and telangiectases mainly on the trunk and lower legs.

Cutaneous lesions may be confused with [mycosis fungoides](#), [sarcoidosis](#), [blood vessel tumours](#), or secondary cutaneous involvement by lymphoma or leukaemia. As the disease progresses secondary organ involvement often occurs. Prognosis is poor, particularly if the disease has spread to other parts of the body.

## Related information

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### References:

- 1. Blood, 15 May 2005, Vol. 105, No. 10, pp. 3768–3785: [WHO–EORTC classification for cutaneous lymphomas](#)
- Book: Textbook of Dermatology. Ed Rook A, Wilkinson DS, Ebling FJB, Champion RH, Burton JL. Fourth edition. Blackwell Scientific Publications.
- Cutaneous B-cell Lymphoma. The Doctor's Doctor.
- [Connors JM, His ED, Foss FM. Lymphoma of the Skin, Hematology 2002. American Society of Hematology Education Program Book.](#)

### On DermNet NZ:

- [Skin lesions, tumours and cancers](#)
- [Cutaneous T-cell lymphoma](#)
- [Lymphocytoma cutis](#)

### Other websites:

- [Cutaneous CD30+ \(Ki-1\) Anaplastic Large-Cell Lymphoma](#) - emedicine dermatology, the online textbook
- [Cutaneous Lymphoma Foundation](#)

### Books about skin diseases:

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