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Primary cutaneous follicle centre lymphoma

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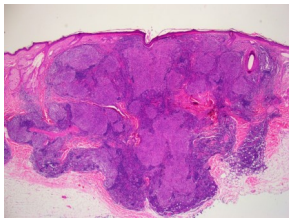
Primary cutaneous follicle centre lymphoma (PCFCL) is a low-grade B-cell lymphoma, comprising neoplastic follicle centre cells. It constitutes 50% of all primary cutaneous B-cell lymphomas. It is a disease of adults – mainly middle-aged – with men affected more often than women. Lesions most often present on the scalp, forehead or trunk. They are usually characterised by single or localised, erythematous/violaceous plaques, nodules or tumours, which are not ulcerated. Multifocal disease is uncommon, as is lower limb involvement.

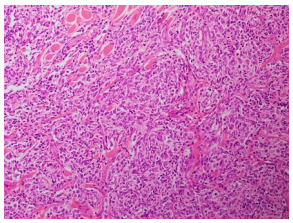
Histologically, the lesions can exhibit a follicular, follicular and diffuse, or diffuse growth pattern. This involves the dermis and often subcutis, with epidermal sparing. Neoplastic follicles in the follicular pattern are ill-defined and contain a monotonous population of follicle centre cells, in the absence of tingible-body macrophages. The diffuse pattern comprises large centrocytes with admixed centroblasts. Rarely, the centrocytes can assume a spindled morphology. Neoplastic cells are CD20+, CD79a+ and BCL6+. MUM1 is usually negative. In the follicular pattern, CD10 may be positive, while the Ki67 proliferation index is low within neoplastic follicles. In the diffuse pattern, CD10 is negative, while the Ki67 proliferation index is usually high. BCL2 is negative or very weakly positive in neoplastic B-cells. The presence of strong BCL2 and CD10 expression within the latter however, lends weight to the likelihood of secondary cutaneous involvement by nodal follicular lymphoma, and close staging in such circumstances is prudent.

Most cases of PCFCL show monoclonal rearrangement of the immunoglobulin genes. Some studies have reported t(14;18) translocation in a minority of cases, with both follicular and diffuse growth patterns, however this is generally seen in nodal follicular lymphoma.

The prognosis for PCFCL is generally excellent with a five-year survival of greater than 95%. This is independent of the growth pattern, number of blasts, the presence of t(14;18) or BCL2 expression. A caveat to this is cases of diffuse pattern PCFCL arising on the legs; these have a worse prognosis, similar to that of primary cutaneous diffuse large B-cell lymphoma, leg type.



Primary cutaneous follicle centre lymphoma (PCFCL): Follicular pattern



PCFCL: Cellular sheets predominantly comprising centrocytes, some with spindled morphology, as well as sparse, reactive small lymphocytes

References

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Dr Khamu is a Dermatopathologist who joined our Collingwood laboratory in July 2017. Dr Khamu graduated from the University of Queensland in 2005. He undertook his Anatomical Pathology training in Brisbane between 2007 and 2012, having completed rotations at various institutions including the Princess Alexandra Hospital, the Royal Brisbane and Women's Hospital and Sullivan Nicolaides Pathology. During this time he was also an associate lecturer in Pathology at the University of Queensland. Upon completion of his training, Dr Khamu joined Melbourne Pathology in early 2012 as a Consultant Pathologist. In 2013, he returned to Brisbane, having been given the opportunity to work with Professor David Weedon at Sullivan Nicolaides Pathology. Over the five years in this role, Dr Khamu gained valuable and broad dermatopathology experience, including GP and dermatology skin reporting. He also continued to enjoy teaching, playing an active role in the education of pathology and dermatology registrars, as well as GPs.