

## Giant Primary Cutaneous Follicle Centre B-Cell Lymphoma

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### Case Presentation

A 67-year-old man presented with firm, smooth, violaceous scalp nodules in the frontal-parietal region, which had been present for 4 years. The lesions exhibited slow growth and had merged to form a mass with telangiectasia, necrosis, ulceration, and crusting. Histological evaluation showed a dense follicular proliferation separated from the epidermis by a grenz zone, consisting of enlarged centrocytes intermixed with centroblasts. Immunohistochemistry was positive for Bcl-6, CD20, CD79a, PAX-5, while CD10, Bcl-2, CD34, CD38, CD68, IRF4/MUM1, CD4 were negative. Subsequent staging CT scan showed infiltration of the periosteum, bone, and dura mater, but no evidence of nodal or extracutaneous visceral disease was found. A bone marrow biopsy did not reveal any signs of lymphoma. We made a diagnosis of primary cutaneous follicle center B-cell lymphoma (PCFCL). The patient was treated with the R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone) regimen and achieved complete remission after 3 cycles. No relapses were observed during 18 months of follow-up.

### Teaching Point

PCFCL accounts for 12% of cutaneous lymphomas and typically occurs between the ages of 50 and 70 [1]. It manifests as slow-growing erythematous papules, plaques, or tumors primarily on the head-neck region or trunk.

Histologically, PCFCL is characterized by the proliferation of centrocytes, centroblasts, and a few reactive lymphocytes. The immunophenotype of neoplastic cells shows positive expression of B-cell markers such as CD20, CD79a, and Bcl-6, while lacking CD5 or CD43 expression, which distinguishes it from systemic follicular lymphomas. These markers are also helpful in differentiating PCFCL from primary cutaneous diffuse large B-cell lymphoma, leg type (which is positive for Bcl-2 and MUM1) and primary cutaneous marginal zone lymphoma (which is positive for Bcl-2 but negative for Bcl-6 and CD10) [2].

Although PCFCL has an excellent prognosis, long-standing untreated masses can invade deeper structures, leading to increased morbidity and mortality. Nevertheless, even bulky and neglected disease can show rapid responses to treatment.



**Figure 1.** (A,B) Pictures of the giant mass of the scalp affecting the fronto-parietal area, in frontal (A) and lateral (B) view. (C) Detail of the scalp mass. It is deducible that the mass resulted from the confluence of nodules, as an isolated tumor is visible in the lower part of the pictures. Telangiectasia, crusting, necrosis with ulceration and fibrin deposits are also appreciable.

## References

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