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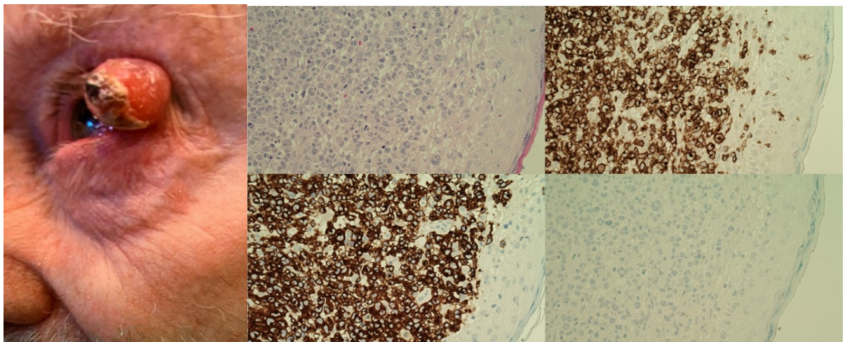
A rare case of primary cutaneous-anaplastic large cell lymphoma (PC-ALCL) localized to the eyelid



A 78-year-old gentleman presented with a three-months history of a rapidly enlarging ulcerated eyelid nodule, which was subsequently excised with incomplete margins. Positron emission tomography/computed tomography (PET/CT) demonstrated no avid sites, indicating low-volume localized disease. Histopathology revealed CD30-positive lymphoproliferative T cells, suggesting differential diagnoses that include lymphomatoid papulosis (LYP) and primary cutaneous-Anaplastic Large Cell Lymphoma (PC-ALCL). The tumour cells infiltrating the dermis were large with anaplastic morphology (top left) and stained for T cell markers such as CD2 (top right) with expression of CD30 (bottom left) but were negative for ALK1 (bottom right). The cells stained for CD2, CD4, CD5, CD45; negative for CD7, CD8, CD10, CD20, CD22, CD79a, CD138 and ALK1. Clinico-pathological correlation lead to the diagnosis of PC-ALCL. Post-operative radiotherapy (RT) treatment was then delivered given positive margins.

PC-ALCL is a rare T-cell lymphoma subtype of non-Hodgkin Lymphoma with only circa 501 cases recorded from 2005 to 2016.[1] Comprising 25–30% of primary cutaneous lymphomas, it is the second most common type of cutaneous T-cell lymphoma.[2] Around 80% of patients present with a localized cutaneous nodule. Distinguishing PC-ALCL from LYP requires clinical judgement due to overlapping histopathologic, phenotypic, and genetic characteristics. PC-ALCL patients often present with erythematous ulcerated nodules, while LYP is characterized by recurrent eruption of multiple papulonodular lesions spontaneously regressing after a few weeks. The median age of PC-ALCL patients is 60 years-old, in contrast to that of LYP, which is 45 years-old.[3]

The prognosis of PC-ALCL is favourable with a 5-year survival rate of 80–90%.[4] Excision and RT are first line therapies in patients with solitary nodules. Those with recurrent disease or multifocal lesions at presentation require systemic therapies. Several case reports have demonstrated the effectiveness of first-line low dose methotrexate. Brentuximab (anti-CD30 monoclonal antibody) and a regimen containing cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) are options in those with increased nodal involvement or who have failed other treatment modalities.

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