Anaplastic T-Cell Non-Hodgkin's Lymphoma: Mucocutaneous Presentation

Linfoma Não-Hodgkin Anaplásico de Células T: Apresentação Mucocutânea

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A 41-year-old male presented with a 4 cm nodular, vegetating and ulcerated lesion of the upper lip, with two weeks of evolution (Fig.s 1 and 2). Biopsy revealed primary cutaneous anaplastic T-cell non-Hodgkin's lymphoma (pc-ALCL). The immunohistochemical study revealed CD30 positivity and ALK negativity, and the CT staging showed localized disease only, LDH was high and bone marrow biopsy revealed no alterations - stage I-A. He underwent radiotherapy and chemotherapy with CHOP, with regression of the upper lip lesion. After 6 months, there was appearance of non-characteristic scattered skin lesions on the trunk and limbs, from papules to ulcerated nodules with <2 cm, associated with B symptoms, whose biopsy was compatible with recurrence. He then underwent second-line treatment with etoposide and subsequent autologous bone marrow stem cell transplantation. Four months after transplantation, he presented a new recurrence of cutaneous lesions, histologically compatible with lymphoma recurrence. Since the lesions disappeared spontaneously, leaving only a residual hyperpigmented smear and occasional itching, without B symptoms and, as both the PET-scan and the bone marrow biopsy did not show alterations, only clinical surveillance was decided.

ALCL represents mature T-cell neoplasms that express the lymphocyte activation marker CD30.[1] They can be

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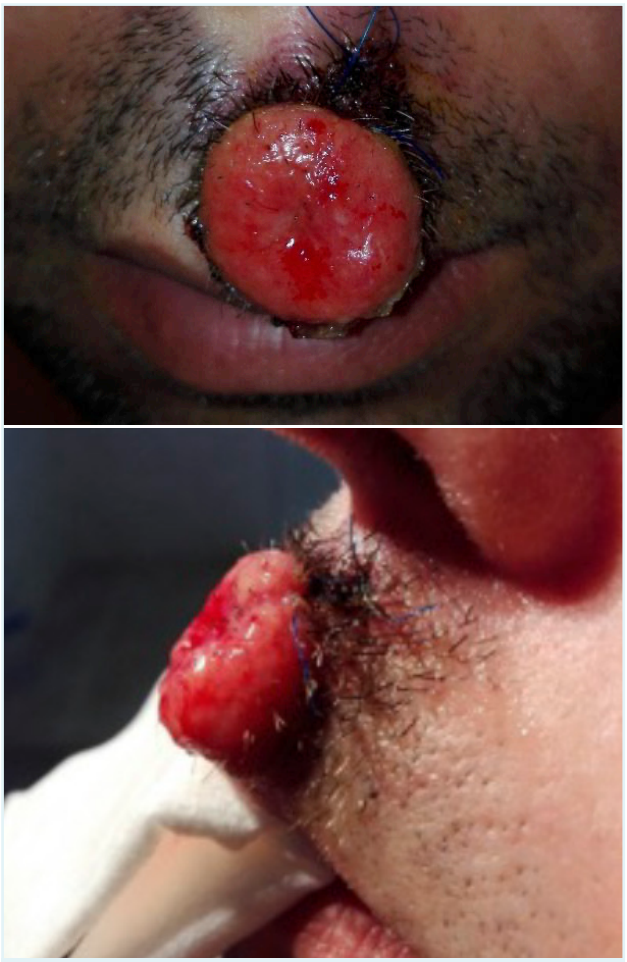

FIGURE 1 and 2. Nodular pc-ALCL lesion in upper lip.

grouped based on the expression of the ALK marker (positive or negative) and by clinical presentation (systemic or localized).[1] Localized forms of ALCL include primary cutaneous (pc) ALCL and breast implant-associated ALCL (BIA-ALCL).[1]

Pc-ALCL accounts for 8% of cutaneous T-cell lymphoma (CTCLs); it is seen mostly in adults, with males diagnosed more often than females.[2] Clinically, pc-ALCL presents with nodular cutaneous lesions measuring over 1.5 cm usually in the face, trunk, extremities and buttocks.[3] Although the majority of the patients present with solitary or localised nodules or papules that may demonstrate ulceration, multifocal lesions are see in 20% of patients.[3] These lesions may have partial or complete spontaneous regression, but in 10% of the patients, extracutaneous dissemination occurs, which usually involves lymph nodes.[3] Patients with pc-ALCL have an excellent prognosis, with a 5-year disease-specific survival over 90%.[3]

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