


Primary multifocal anaplastic large cell lymphoma with eyelid involvement: a case report

Linfoma anaplásico de grandes células primário multifocal com envolvimento de pálpebra: relato de caso

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ABSTRACT | Ocular adnexal involvement in CD30+ lymphoproliferative disorders is rare. We report the case of a 73-year-old woman with a relapsing primary cutaneous anaplastic large cell lymphoma on her eyelid. A systemic extension study excluded extracutaneous involvement. Systemic chemotherapy resulted in an optimal response, with complete regression of the cutaneous lesions. There has been no recurrence during the 2 years of follow-up.

Keywords: Eyelid neoplasms; Lymphoma, primary cutaneous anaplastic large cell; Lymphoma, T-cell, cutaneous; Case reports

RESUMO | O acometimento ocular adicional nos distúrbios linfoproliferativos CD30+ é raro. Relatamos o caso de uma mulher de 73 anos com linfoma de grandes células anaplásicas primárias recidivantes em sua pálpebra. A avaliação sistêmica excluiu envolvimento extracutâneo. A quimioterapia sistêmica resultou em uma resposta ótima, com regressão completa das lesões cutâneas. Não houve recidiva durante 2 anos de acompanhamento.

Descritores: Neoplasias palpebrais; Linfoma anaplásico cutâneo primário de células grandes; Linfoma cutâneo de células T; Relatos de casos

INTRODUCTION

CD30+ lymphoproliferative disorders are type T lymphomas. They are characterized by large, anaplas-

tic morphology and the CD30+ immunophenotype of tumoral cells and include lymphomatous papulosis, anaplastic large cell primary cutaneous lymphoma, and anaplastic large cell systemic lymphoma. Eyelid involvement in anaplastic large cell lymphoma is rare and usually appears secondary to systemic anaplastic large cell lymphoma.

CASE REPORT

A 73-year-old woman presented with a multinodular lesion in her right upper eyelid that had grown slowly over the previous 8 weeks. Examination showed diffuse eyelid edema and redness. In addition, there were five erythematous subcutaneous nodules firmly adhered to deep planes, one of them located on the eyelid margin, another with central ulceration, and a medial nodule with ulceration and secondary hyperkeratosis (Figure 1). An ophthalmic examination revealed mechanical ptosis secondary to palpebral involvement, with preserved visual acuity. An intraocular pressure test, fundus examination, and test of extraocular motility revealed no pathologic findings. Examination results for the other eye were all within the normal range.

The patient's previous medical history included an anaplastic large cell primary cutaneous lymphoma on her lower limb. This had relapsed locally following first-line therapy with R-CHOP administered two years ago (Figure 2).

A CT scan revealed a circumscribed hyperdense eyelid mass, 15 × 7 mm in size, which did not compromise the eye; no orbital lesions were observed. An incisional biopsy of the lesion and a systemic extension study were

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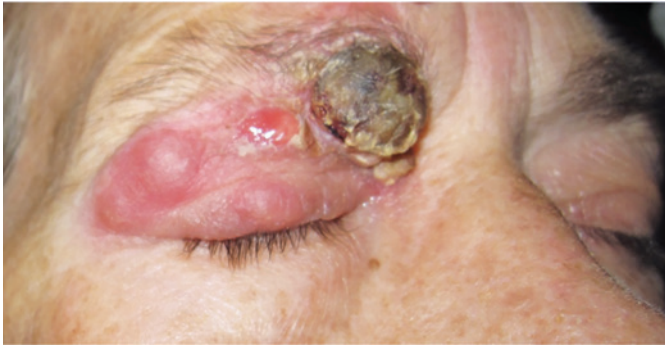


Figure 1. The multinodular lesion in the patient's upper eyelid, with ulceration and secondary hyperkeratosis.



Figure 2. Ulcerative lesions on the patient's lower limb.

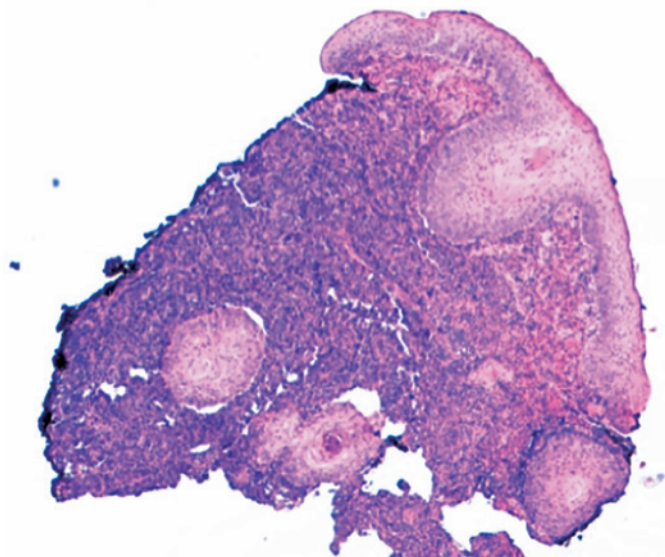


Figure 3. Incisional biopsy, showing pseudoepitheliomatous epidermal hyperplasia and giant atypical cells with dermic tumoral infiltrate.

performed. The anatomopathological study revealed pseudoepitheliomatous epidermal hyperplasia associated with giant atypical cells and dermic tumoral infiltrate that was positive for CD30 and CD45 and negative for CD20, CD3, CD2, CD5, CD8, CD7, AE1/AE3, S100, EMA, and ALK (Figure 3). The systemic extension study ruled out extracutaneous involvement. The diagnosis was multifocal primary cutaneous large cell anaplastic lymphoma, with a TNM stage of T3aN0M0.

Second-line systemic chemotherapy (six cycles of navelbine and gemcitabine) achieved complete regression of the eyelid and limb lesions (Figure 4). After two years, the patient remains in complete remission.

DISCUSSION

Lymphomas are the most common type of ocular adnexal neoplasia, affecting the conjunctiva, lacrimal gland, eyelids, and orbital tissues. Non-Hodgkin B lymphomas account for more than 90% of adnexal ocular lymphomas, whereas only 1%-2% exhibit the T immunophenotype^(1,2). A great many studies have focused on the most common subtypes. In order of frequency, these are mucosa-associated lymphoid tissue lymphoma, follicular lymphoma, giant cell diffuse B lymphoma, and mantle cell lymphoma. Studies of the uncommon subtypes have been scarce and describe either small groups within larger series of B lymphomas or sporadic case reports. Because they are uncommon, adnexal lymphomas can be difficult to diagnose and manage⁽³⁾.

Because of the clinical presentation and the multinodular and ulcerative aspects of our patient's lesion, the differential diagnosis considered epidermoid carcinoma, Merkel cell carcinoma, sebaceous cell carcinoma, and even metastasis. Her previous history of anaplastic large cell primary cutaneous lymphoma suggested the possibility of a lymphoid tumor.

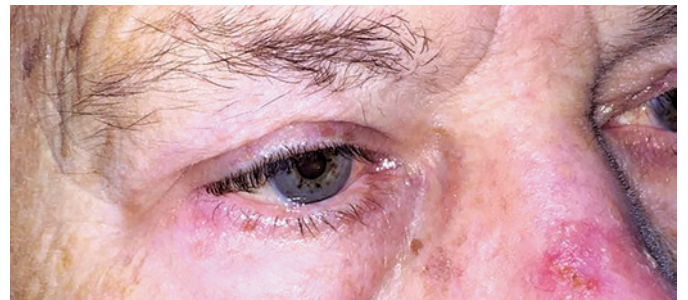


Figure 4. The eyelid after systemic chemotherapy, showing complete regression of the lesion.

CD30+ anaplastic large cell primary cutaneous lymphomas typically present in elderly patients as ulcerated solitary nodules on the limbs, with over 20% being multifocal. Eyelid involvement in an anaplastic large cell primary cutaneous lymphoma is rare and usually appears secondary to systemic anaplastic large cell lymphoma. Even though eyelid involvement by primary cutaneous anaplastic large cell lymphoma, as shown in our clinical case, presents rarely, there have been previous reports of this^(1,2,4).

The mean survival rate after 5 years is 90%, but relapses are frequent, so strict follow-up of this patient group is essential. Extracutaneous dissemination, particularly to regional lymph nodes, occurs in approximately 10% of cases⁽⁵⁾. A variant of keratoacanthoma-like cutaneous anaplastic large cell lymphoma has been described^(6,7); this could have been a diagnostic option for our case, given that these lesions are prone to ulceration and hyperkeratosis.

The definitive diagnosis is made anatomopathologically, especially for lymphoid tumors, where immunohistochemistry is essential. A differential diagnosis that includes other CD30+ lymphoproliferative disorders, especially anaplastic large cell lymphoma with secondary cutaneous involvement, is fundamental. A systemic workout is essential; although the lesions are similar, the prognosis and therapeutic approach differ significantly.

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