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Table of Contents

Original Articles

Review Article

Case Reports

Conference Highlights/Scientific Contributions

Case Report

Lymphoid proliferation in Eyelid: A Primary Follicular Lymphoma Case

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Abstract

Ocular adnexal lymphoma (OAL) constitutes 8% of all extranodal lymphomas, being rare in the eyelids (5–20%). The objective was to report a case of eyelid follicular lymphoma in a young adult woman. A 35–year–old female with swelling in left lower eyelid with development for 2 months. The visual acuity was 20/20 in both eyes, the tumor mass was 3 cm wide and 1.5 cm long. The contrast–enhanced computed tomography showed a soft–tissue injury in the left lower eyelid but had no signs of local spread according to description. Grade 2 follicular lymphoma was diagnosed by biopsy. Oncological evaluation assessed positron emission tomography scan and bone marrow aspiration with negative results for systemic spread of disease. Patient started radiotherapy. This case highlights that young patients may present painless swelling lesions that may be largely underdiagnosed and should take into consideration some oncological pathologies.

Keywords: Case Report; Eyelid neoplasms; Lymphoma; Lymphoma, Follicular

Introduction

Ocular adnexal lymphoma (OAL) represents 8% of all extranodal lymphomas and it is the most common malignant tumor of the orbit [(1)]. The frequency of occurrence is higher in older adults [(2,3,4)]; however, there have been few cases reported in young adults [(5)]. These lymphomas compromise different sites of the eye. In particular sites, the reported frequency of involvement are: 5–20% in the eyelids, 20–33% in the conjunctiva, and 46–74% in the connective orbital tissue and lacrimal and orbital structures [(4.6).]

The aim of the study is to document a case of eyelid follicular lymphoma in a young adult woman.

Case report

The patient is a 35–year–old woman, born in Lima. She has no medical history relevant to the case. The corrected visual acuity was 20/20. There was a tumor in the lower left eyelid and it has had a slow and progressive growth for the last two months. In the left lower eyelid region, a tumor mass that was 3 cm wide and 1.5 cm long was evident. It was adhered to deep plane and had a defined edge. Additionally, the tumor had a hard consistency and extended to the anterior orbit, (Figure 1A) but it was painless to the patient. There was normal fundoscopy in both eyes. The rest of the physical examination went without any complication.

The contrast–enhanced computed tomography showed a soft–tissue injury in the left lower eyelid but had no signs of local spread according to description (Figure 1B).

The patient underwent transconjunctival incisional biopsy, which diagnosed follicular lymphoma grade 2 through the detection of positive immunohistochemical markers for Bcl–2, Bcl–6 and CD20 (Figure 2C–E), CD3 negative (Figure 2F).

Oncological evaluation included assessing the patient through a positron emission tomography (PET) scan (Figure 1C). Additionally, a bone marrow aspiration was conducted but the results were negative for the spread of disease. The patient started with radiotherapy.

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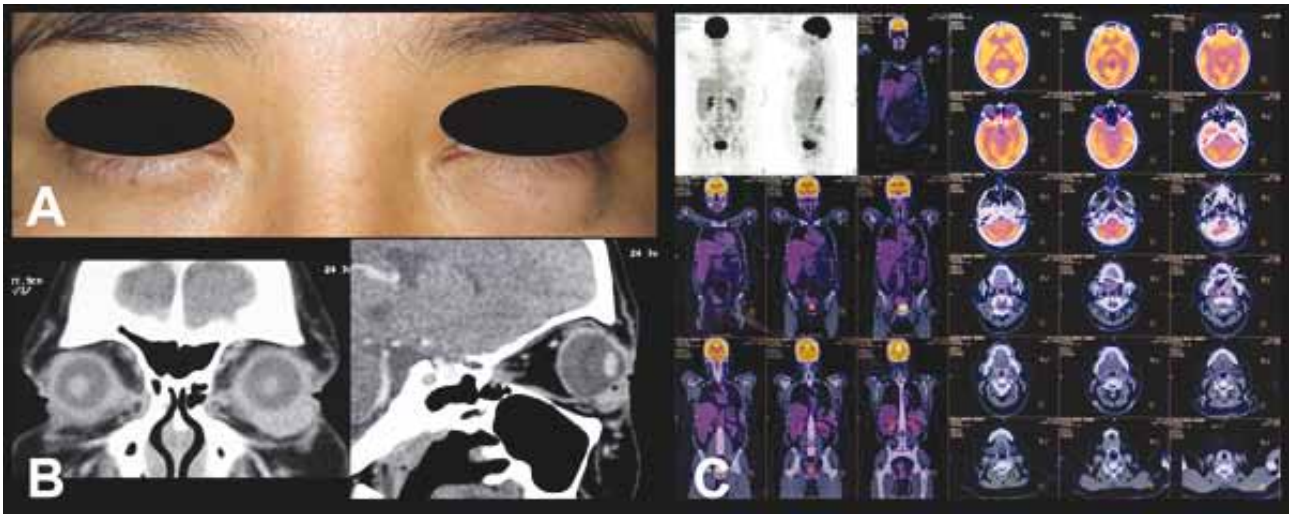

Figure 1. Clinical photographs. A, tumor in the left lower eyelid. B, Contrast–enhanced computed tomography revealed a tumor mass in the left lower eyelid. C, positron emission tomography (PET Scan) showed normal metabolic activity.

Discussion

Ocular adnexal lymphoma infrequently represents approximately 10 to 15% of tumors affecting the orbit, conjunctiva, eyelid and lacrimal sac (1). This case report is unusual because of the age of onset, location and histological type.

With respect to the age of onset, ocular adnexal lymphomas rarely occur in people under 50 years old. According to a series of 7 cases documented by Frimmel et al [3], the age range in which ocular adnexal lymphoma is usually detected is between 51 and 88 years old. However, there are cases of eyelid lymphomas in children like the cases registered by Köksal et al. but they usually correspond to Burkitt's lymphoma [7]. Also, Winhoven et al. and Sanka et al. described some cases found in young people who reported multinodular injuries in the eyelids due to anaplastic large cell lymphoma [8,9].

The location of the tumor in the eyelid is unusual due to the fact that it corresponds to 5–20% of cases of OAL[6]. The indolent clinical presentation and progressive growth in this region poses different possible diagnosis. The patient could be diagnosed with a benign tumor such as lipomas, or less likely, malignant tumors such as sarcomas or lymphomas. Auxiliary tests, such as contrast–enhanced computed tomography imaging, allow the evaluation of the local extent of the disease, and the incisional biopsy of the lesion allows histopathological diagnosis, corresponding to grade 2 follicular lymphoma.

The most common histological type of OAL is the mucosa–associated lymphoma (MALT) [10]; the second most frequent type is follicular lymphoma [1]. According with the precedence set forth in the literature, immunohistochemical lesion phenotype corresponds

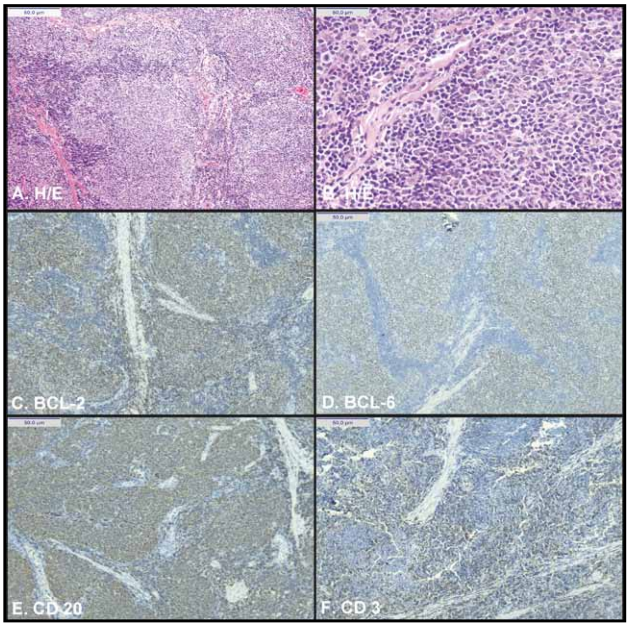

Figure 2. Histopathologic examination. A, Hematoxylin–eosine staining biopsy sample (H–E, 100X), predominantly follicular pattern (more than 75%). B, Higher magnification revealed small and medium centrocytes with inconspicuous nucleoli and scant cytoplasm (H–E, 400X). C, Positive for Bcl–2 antibodies. D, Bcl–6 nuclear immunoreactivity. E, Immunopositivity for CD20 antibodies. F, Negative for CD3 antibodies.

to follicular lymphoma. The patient had positive immunohistochemical B cell markers such as CD20 and negative to CD3, discarding T cell proliferation. Moreover, it showed positive immunoreaction for Bcl–2 and Bcl–6, exclusively found in follicular lymphoma patients [6]. Bcl–2 marker evidence high functionality of antiapoptotic Bcl–2 gene due to either the translocation of chromosome 14 and 18, t (14; 18) (q32; q21), or the translocation of

chromosome 2 and 18, t (2; 18) (p12; q21) [1]. The Ki–67 protein marker also determines the proliferative index in the samples, with a relative frequency of 30% found in the proliferative population.

PET scan test and bone marrow aspirate showed absence of active systemic disease. The patient was referred to treatment with localized radiation therapy using a linear accelerator.

Conclusion

This case report highlights that young patients may present painless swelling lesions that may be largely underdiagnosed and should take into consideration some oncological pathologies such as lymphomas which are rare, requiring biopsy for the correct diagnosis.

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