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Giant Eyelid Tumor Secondary to T-Cell Lymphoma in a 40 Years Old Woman: A Rare Presentation and Literature Review

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Authors' contributions

This work was carried out in collaboration among all authors. Author RZ designed the study, performed the statistical analysis and wrote the protocol. Author MK wrote the first draft of the manuscript. Authors AE and ZC managed the analyses of the study, as well as the literature searches. Authors KR and AO approved and supervised the work. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Lymphoma is a malignant neoplasm originating from monoclonal B-lymphocytes, T lymphocytes or from natural killer (NK) cells; Lymphoma of the eyelid is defined as lymphoma infiltrating the preseptal tissues, meaning lymphoma infiltrating the skin, subcutaneous tissue and the orbicularis muscle. We report a case of a 40-year-old young woman with a higher eyelid giant tumor with Literature Review.

Keywords: Lymphoma; neoplasm; orbicularis muscle.

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1. INTRODUCTION

Lymphoma of the eyelid constitutes 5% of ocular adnexal lymphoma. In previously published cases, 56% of lymphomas of the eyelid are of B-cell origin and 44% are of T-cell origin. Peripheral T-cell lymphoma (PTCL) represents a broad spectrum of disease with several different manifestations. PTCL often occurs in the extremities, but rarely in the eyelid. Periocular involvement by PTCL usually occurs in the setting of the Sézary syndrome or mycosis fungoides, which is a variant of PTCL [1-3].

We report a case ofa 40-year-old young woman presenting with a higher eyelid giant tumor.

2. CASE REPORT

We report the case of a 40-year-old patient with no significant pathological antecedents who presented a year ago a tumefaction of the upper right eyelid associated with another mass of the soft parts of the right leg. A biopsy was made and Objective primary cutaneous T-cell lymphoma with mean cellularity: CD4 +. The patient was admitted to a clinical hematology service to benefit from chemotherapy. The first course was received on 28/11/18 and the fourth course on 12/02/2019 with progression of the two lesions under treatment. A second line of chemotherapy is planned in the patient after resection of the palpebral tumor.

On examination we find a patient in good general condition without clinical tumoral syndroma with an ulcero-necrotic palpebral mass of 12 centimeters in diameter (Fig. 1) with a second ulcero-necrotic lesion of the right leg (Fig. 2).

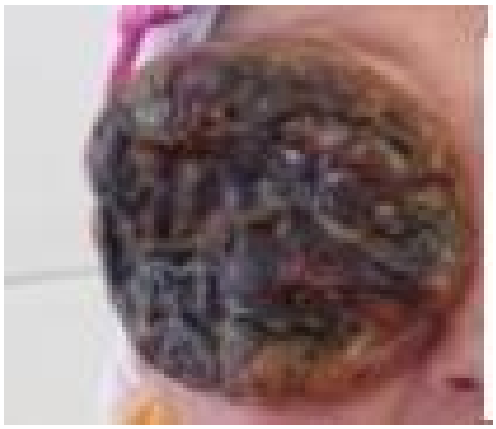

Fig. 1. Ulcero-necrotic palpebral mass

Orbital MRI has objectified a tumoral ulcero tumbling process of soft tissues extending in intra-orbital and coming into close contact with the eyeball (Figs. 3 and 4). The biological assessment was normal. The patient was transferred to maxillofacial surgery for resection with anatomopathological and immuno-histochemical study.

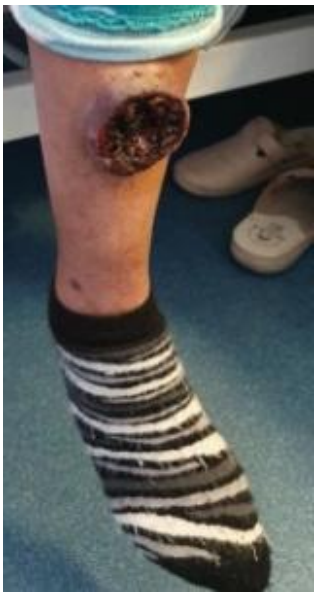

Fig. 2. Ulcero-necrotic lesion of the right leg

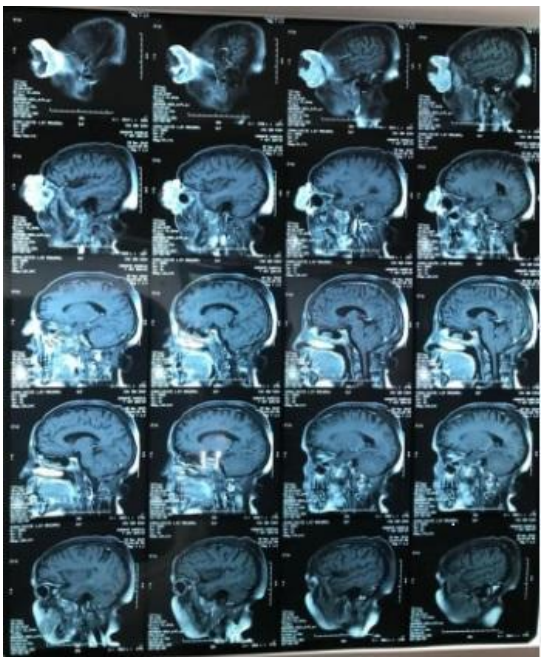

Fig. 3. Images of orbital MRI

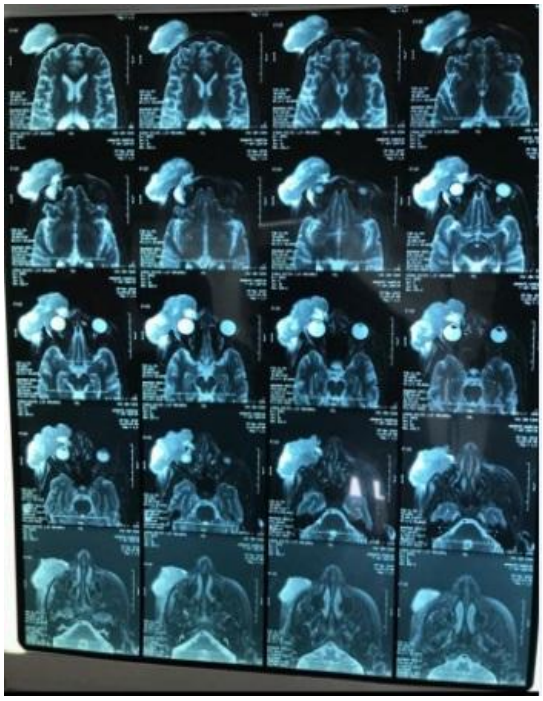

Fig. 4. Images of orbital MRI close contact with the eyeball

3. DISCUSSION

Lymphoma is a malignant neoplasm originating from monoclonal B-lymphocytes, T lymphocytes or, less commonly, from natural killer (NK) cells. Lymphoma can be divided into Hodgkin lymphoma and non-Hodgkin lymphoma (NHL). NHL is a large heterogeneous group consisting mostly of B-cell lymphomas (80%) and, less frequently, T-cell lymphomas (14%) or NK-cell lymphomas (6%) [4].

Lymphoma of the eyelid is defined as lymphoma infiltrating the preseptal tissues, [5] meaning lymphoma infiltrating the skin, subcutaneous tissue and the orbicularis muscle. This corresponds to the anterior lamella of the eyelid. We do not include lymphoma of the lacrimal sac. Ocular adnexal lymphoma (OAL) is rare, constituting 1-2% of NHL and 7-8% of extranodal lymphomas, [6] where lymphoma of the eyelid constitute 6-24% of OAL [7-8].

Primary lymphoma is defined by the following: 1) proven lymphoma of the eyelid with no evidence of concurrent systemic disease and 2) no prior history of lymphoma. Patients with concurrent or prior lymphoma are considered to have secondary lymphoma [9].

We describe a case of aggressive secondary ocular adnexal lymphoma (OAL) of T-cell origin that presented as an ulcero necrotic palpebral mass of 12 cm in diameter.Ocular manifestations of PTCL can include both intraocular and extraocular conditions [10].

Cook and colleagues [11] reportedthat eyelid ectropion was the most common findings in theirpatients with PTCL, though other ocular manifestations,including eyelid thickening or edema, placoid tumor, tightskin, blepharitis and corneal abnormalities, were also found.

Stenson and Ramsay [12] found eyelid tumors in eight of 30 consecutive patients with mycosis fungoides diagnosed bybiopsy. Isolated PTCL of the eyelid is rare.

In thistype of lymphoma, relapses are common and can producean indolent course. Kirsch and colleagues reported a patientwith multiple recurrences of systemic T-cell lymphoma afterinitial presentation with only an eyelid mass that was treated with systemic chemotherapy and local radiation.

When lymphoma of the eyelid is suspected, a thorough ophthalmological examination must be carried out, as well asa general examination for lymphadenopathy and extraocular involvement, especially of the skin in cutaneous T-cell lymphomas. A biopsy should be performed as the basis for diagnosis and further subclassification. If lymphoma is diagnosed,a full staging procedure must be carried out. This procedureincludes imaging such as magnetic resonance imaging orpositron emission tomography-computed tomography toevaluate spread to the orbit, the lacrimal gland, or intracranialstructures, as well as characterization of disseminated lymphoma. The staging procedure must also include a bonemarrow biopsy [9].

Histopathologic subclassification of lymphoma is a complexprocedure involving evaluation of the tumor's morphologyand immunohistochemistry. The tissue from the biopsy isformalin-fixed, paraffin-embedded, and stained withhematoxylin-eosin. The tissue is also analyzed immuno histochemically using antibodies against CD3, CD5, CD20, andCD79a. The reactivity of the tumor cells with these antibodiesdetermines whether it is a B-cell or a T-cell lymphoma. B-celllymphomas are further characterized using CD10, CD23, CD30, cyclin D1, Bcl-2, Bcl-6,

MUM-1, k light chain, and l lightchain antibodies. T-cell lymphomas are characterized using antibodies against CD4, CD8, CD30, CD56, ALK-1, TIA, andgranzyme B.

To determine the correct treatment for lymphoma of theeyelid, a thorough examination and grading of each patient isnecessary. This must be carried out by a team of hematologists, radiotherapists, and ophthalmologists. A number offactors must be considered when determining treatment forlymphoma of the eyelid. These include the histopathologic subtype, the extent of the lymphoma, disease-specific andgeneral prognostic factors and the impact of the lymphomaon the eye. The most common treatments for lymphoma of the eyelidare radiotherapy, chemotherapy and surgery, either alone orin combination. Less commonly used treatments include corticosteroids and monoclonal antibodies, as well as very infrequently used methods such as PUVA, PDT, brachytherapy, plasmapheresis, interferon alpha and bone marrow transplant [13].

In many of the cases included in this study, long-term followup is missing, making an estimate of prognosis difficult. Sixty-one cases (31%) of lymphoma of the eyelid reported recurrence or progression, whereas 69 cases (35%) did reportfollow-up without recurrence or progression. Twenty-six ofthese (13%) had follow-up periods of 2 years or more. The most important prognostic factor for lymphoma of theeyelid is its histological subtype. Some subtypes, especially NKTL, have a very poor prognosis, whereas others, such asC-ALCL and EMZL, have a goodprognosis .Secondary disease and high-stage disease are predictors ofpoorer prognosis. The suggestion that the prognosis of aspecific lymphoma of the eyelid is worse than that of the samelymphoma located in other structures of the ocular adnexalregion does not seem to be true, at least for several subtypesof lymphoma of the eyelid.

The overall recurrence/progression rate of T-cell lymphomas of the eyelid is 36%. High-grade NKTL has a poorprognosis, with 10 of 12 patients experiencing recurrence orprogression. Eight of the 12 cases died of the disease. NKTL occurring outside the nasal cavity is aggressive, withshort survival time and poor response to therapy. MF in theeyelid has a recurrence/ progression rate of 44%. The prognosisof MF in general depends on its clinical stage; patients withlow-stage tumors have a good prognosis, whereas patientswith advanced disease have a poor prognosis [14]. The othercutaneous T-cell lymphomas of the eyelid seem to have agood prognosis, with C-ALCL recurring in only 1 of 12 patients.

4. CONCLUSION

Lymphoma of the eyelid is a rare disease; only 199 cases ofknown cell origin have been reported. Because lymphoma of the eyelid is a rare disease, its diagnosis is often difficult, andcorrect treatment is often delayed, with a first tentative diagnosis of infection or inflammation being common. With the exception of a few subtypes, lymphoma of theeyelid commonly affects elderly patients. The presenting symptoms of lymphoma ofthe eyelid are most commonly tumor and swelling of theeyelid. Ulceration and erythema are frequent in T-celllymphoma but very infrequent in B-cell lymphoma. When choosing treatment and handling of lymphoma of the eyelid, the histopathological subtype and the clinical stage of the tumor serve as the best indicators of prognosis. WHO studies describe lymphoma subtype with different clinical features, behavior, and prognosis and thelymphoma must be handled accordingly. Radiotherapy is thetreatment of choice for solitary, low-grade lymphomas and chemotherapy is the treatment of choice for disseminated and high-grade lymphomas.

Further studies of the genetics and pathogenesis of lymphoma of the eyelid are needed to provide a better understanding of the disease and to make itpossible to optimize treatment and thereby improve theprognosis even further.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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