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Malt Lymphoma of the Eyelid: Management of A Rare Entity

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Abstract **Case Report**

Mucosa-associated lymphoid tissue (MALT) lymphoma is a rare subtype of non-Hodgkin lymphoma that originates in the mucosa-associated lymphoid tissue. The most commonly affected organ is the stomach, although it can involve nearly any organ throughout the body. When a suspicious lesion is present, an excisional biopsy may be performed for diagnosis. For staging, blood tests and imaging studies such as abdominal and chest CT scans are typically conducted. A bone marrow biopsy may also be performed if necessary. This patient initially presented with a palpable mass on the left upper eyelid. An extensive excisional biopsy was performed under general anesthesia. It revealed MALT lymphoma upon histopathological examination. This case highlights a diagnosis of MALT lymphoma in the subcutaneous tissue of the left upper eyelid. Lymphoma of the eyelid is an area that has received limited research attention, with few large-scale studies or recent reviews available in the literature.

Keywords: Lymphoid hyperplasia, Non-Hodgkin lymphoma, Mucosa-associated lymphoid tissue, MALT.

INTRODUCTION

Lymphoma of the eyelid is defined as lymphoma infiltrating the preseptal tissues, meaning lymphoma infiltrating the skin, subcutaneous tissue and/or 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, where lymphoma of the eyelid constitutes 6-24% of OAL [1,2,3]

When lymphoma of the eyelid is suspected, a thorough ophthalmological examination must be carried out, as well as a 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 sub-classification. If lymphoma is diagnosed, a full staging procedure must be carried out. This procedure includes imaging such as MRI or PET-CT to evaluate spread to the orbit, the lacrimal gland, or intracranial structures, as well as characterization of disseminated lymphoma. The staging procedure must also include a bone marrow biopsy. [4]

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. [5,]

Lymphoma of the eyelid is an area that has received limited research attention, with few large-scale studies or recent reviews available in the literature. The prognosis for eyelid lymphoma may be considerably poorer than that of other ocular adnexal lymphomas, particularly conjunctival lymphoma. [5,6]

Our objective is to discuss and assess the clinical features and findings associated with eyelid lymphoma, as well as to explore the treatment options available for this condition.

CASE PRESENTATION

A 57-year-old male was presented with a palpable mass with 2×1-cm-sized on his left upper eyelid. Upon examination, an elevated, mobile, pinkish lesion was found extending to the entire upper eyelid. (Fig. 1,2) With excisional biopsy, it was diagnosed as MALT lymphoma on pathologic test.

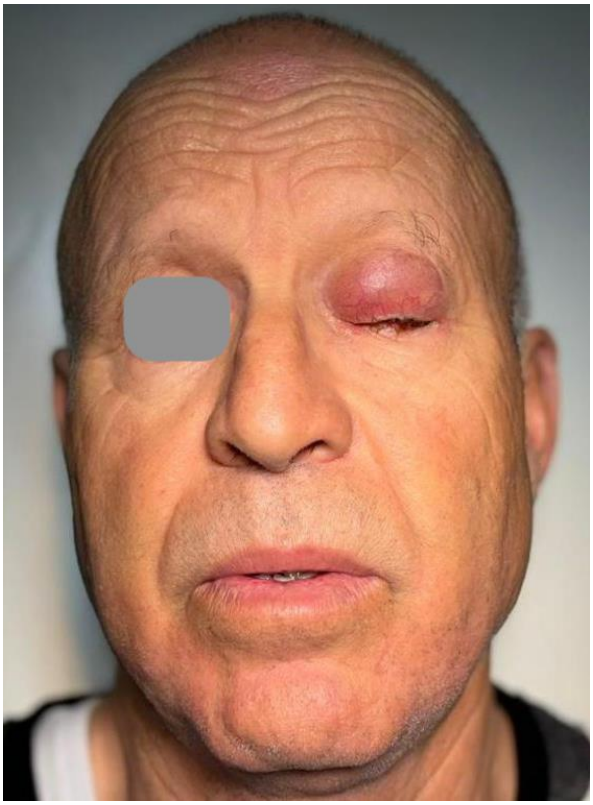

Figure 1 : Frontal view showing the left upper eyelid swelliing

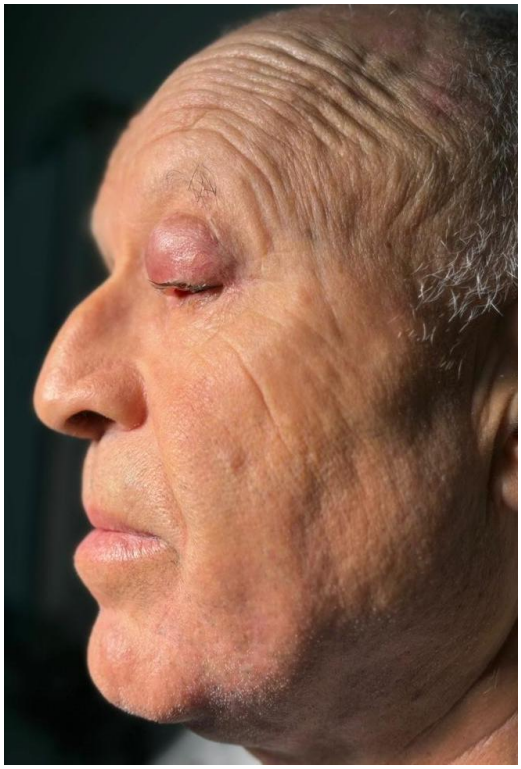

Figure 2: Side view showing the left upper eyelid swelliing

During his visit, the patient reported a recurrent, enlarging palpable mass and requested an excisional biopsy to determine its exact nature. He subsequently underwent excisional biopsy of the same site on the left upper eyelid under general anesthesia. Intraoperatively, a yellowish, oval-shaped mass was found adherent to the orbicularis oculi muscle. Initially suspected to be a fat mass from the upper eyelid, histopathological examination revealed it to be MALT lymphoma. (Fig. 3)

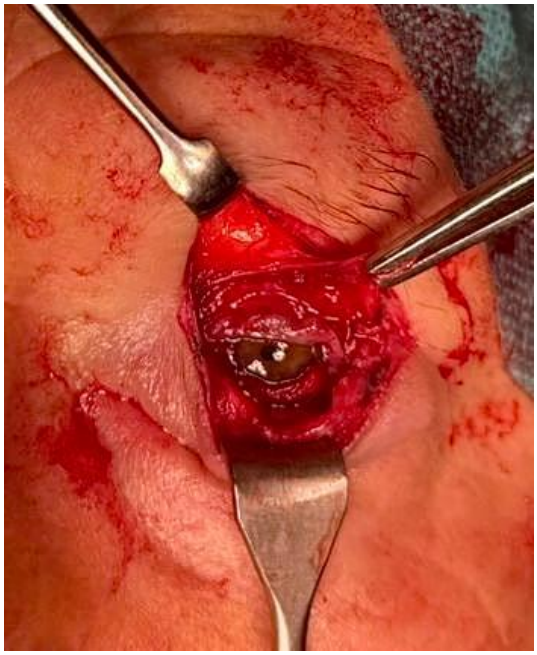

Figure 3: Operative view after the excision of the lesion

Immunohistochemical staining was positive for CD79a, CD20, and reactive CD3-positive T cells, while cyclin D1 was negative. Laboratory tests showed normal complete blood count, including leukocyte and neutrophil levels. Liver function tests, lactate dehydrogenase (LDH), beta-2 microglobulin, and serum protein electrophoresis were all within normal limits. Serologic testing for HIV, hepatitis B, and hepatitis C was negative. Additionally, serologic studies for *Chlamydia psittaci*, *Chlamydia trachomatis*, and *Chlamydia pneumoniae* were negative. Immunoglobulin E (IgE) levels were normal, and there was no clinical or laboratory evidence of a pre-existing autoimmune disorder.

The patient underwent positron emission tomography-computed tomography (PET-CT) and contrast-enhanced neck CT for staging. Bone marrow biopsy showed a normal karyotype and no evidence of bone marrow involvement.

Two weeks follow-up was satidfactory both on functional and esthetic level. (Fig. 4,5)

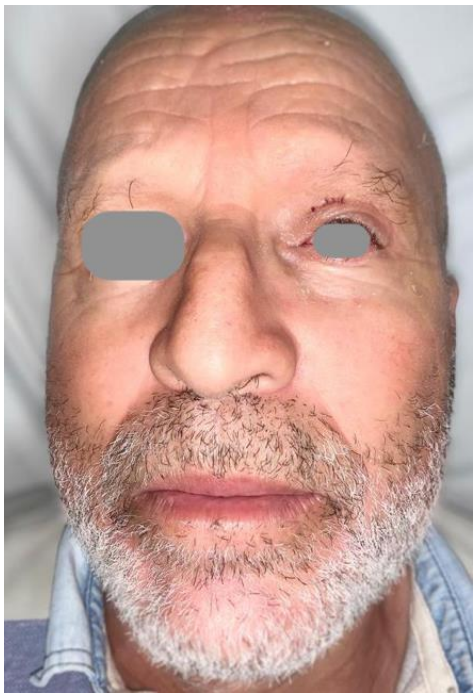

Figure 4 : Two weeks after surgery (eyes open)

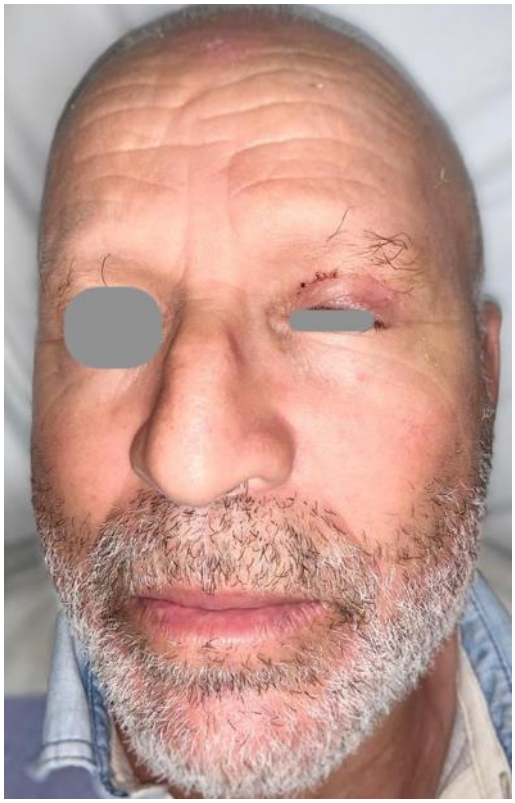

Figure 5: Two weeks after surgery (eyes closed)

The patient was treated with eight cycles of cyclophosphamide, vincristine, rituximab and prednisone, achieving complete remission.

DISCUSSION

Lymphoma is the most common primary malignant tumor of the orbit in adults, accounting for approximately 11% of all orbital masses and 34% of malignant tumors located in the orbit. Ocular adnexal lymphomas (OALs) remain rare, representing only 1–2% of all non-Hodgkin lymphomas (NHLs) and 7–8% of extranodal forms [16,17]. Among the histological subtypes, MALT lymphoma (mucosa-associated lymphoid tissue) is the most frequent. Initially described in the gastric mucosa, it alone accounts for 50–80% of OALs [18]. This type of lymphoma typically arises in the context of chronic inflammation, leading to the development of acquired lymphoid tissue. Its localization in the eyelid is uncommon, observed in approximately 6–24% of cases [19]. It manifests as infiltration of the preseptal tissues, including the skin, subcutaneous tissue, and/or the orbicularis muscle.

This type of lymphoma primarily affects middle-aged to older individuals, generally between the ages of 50 and 70, with a median age around 60 years. A slight female predominance has been reported, with a female-to-male sex ratio estimated at 1.3 [20]. MALT lymphoma arises from B lymphocytes in tissues that are initially devoid of lymphoid structures, under the influence of chronic antigenic stimulation, whether of infectious or autoimmune origin [21,22]. Among the suspected infectious agents, Chlamydia psittaci has been identified in some cases, particularly in forms affecting the ocular adnexa [23].

The clinical presentation of palpebral MALT lymphoma is typically insidious. It most often presents as a firm, painless, non-inflammatory eyelid mass with slow progression. Associated functional signs may include mechanical ptosis, tearing, or eyelid edema. In contrast, proptosis is generally absent in purely palpebral forms, and visual acuity is usually preserved. This non-specific symptomatology can be misleading and mimic various benign conditions such as a recurrent chalazion, chronic blepharitis, basal cell carcinoma, or other eyelid tumors [24,25], which increases the risk of diagnostic delay. The involvement is usually unilateral, although bilateral forms can occur, particularly in the context of systemic or secondary lymphomas [25].

Initial evaluation is based on a comprehensive ophthalmologic examination, combined with a systemic staging workup to assess for lymph node involvement or extra-ocular localization, particularly in cutaneous T-cell forms [21]. Imaging plays a central role, with orbital MRI as the reference modality. It typically reveals homogeneous, well-circumscribed masses, isointense to muscle on T1-weighted sequences, slightly hyperintense on T2, and showing moderate, homogeneous enhancement after gadolinium injection. No signs of bone lysis are usually observed.

The definitive diagnosis of palpebral MALT lymphoma relies on an excisional biopsy, allowing for histopathological analysis. This typically shows a diffuse infiltration of monomorphic lymphocytes, residual germinal centers, and plasmacytoid differentiation. Immunohistochemistry is essential for confirming the diagnosis and guiding classification. MALT lymphomas characteristically express CD20+, CD79a+, and Bcl-2+ markers, while being negative for CD5, CD10, and Cyclin D1. This immunoprofile helps exclude other entities such as mantle cell or follicular lymphomas [22].

Once the diagnosis is confirmed, a full staging workup is mandatory. This includes a cervico-thoraco-abdomino-pelvic CT scan, a PET scan—although its sensitivity is limited for MALT lymphomas—as well as a bone marrow biopsy, as part of the staging according to the Ann Arbor classification. The workup is complemented by virologic assessment, including standard viral serologies and testing for *Chlamydia psittaci* by PCR or serology [21,23]. The goal is to detect possible orbital, lacrimal, or meningeal involvement, particularly in advanced forms [21,23].

The therapeutic management of palpebral MALT lymphoma primarily depends on the extent of the disease at diagnosis. In localized forms, low-dose external radiotherapy (between 20 and 30 Gy) is the treatment of choice. It achieves complete response rates exceeding 90–95% [26,27]. However, it is not without side effects, the most common being dry eye, superficial punctate keratitis, and, in the longer term, cataract formation.

In cases of bilateral or disseminated disease, or when radiotherapy is contraindicated, immunotherapy with rituximab (a monoclonal anti-CD20 antibody) may be proposed, either as monotherapy or in combination with chemotherapeutic agents such as chlorambucil, bendamustine, or within an R-CHOP protocol [28]. Moreover, a watchful waiting approach may be considered in certain situations, particularly in cases of indolent, asymptomatic lymphoma or in frail elderly patients [26].

Finally, in regions where Chlamydia psittaci is endemic, antibiotic treatment with doxycycline (100 mg twice daily for three weeks) has shown tumor response rates of 40 to 60%, suggesting a potential role for bacterial eradication in disease control [29,30].

Palpebral MALT lymphoma is generally an indolent form with an excellent prognosis. The 5-year overall survival exceeds 90–95%, and the risk of transformation into a more aggressive form remains low, estimated between 1% and 3%. Nevertheless, local recurrences are not uncommon, which underscores the need for regular and long-term follow-up. Clinical surveillance every six months is recommended during the first two years after treatment, followed by annual monitoring. This should ideally be supplemented by annual orbital MRI to detect any early recurrence or extension [16,17,18].

It is crucial to distinguish primary forms, strictly limited to the eyelid without systemic involvement or prior lymphoma history, from secondary forms, which reflect dissemination from a pre-existing or concurrent systemic lymphoma [31]. This distinction carries significant prognostic and therapeutic implications. Some studies suggest that palpebral lymphomas may have a slightly less favorable prognosis compared to conjunctival forms, possibly due to later diagnosis or specific biological features of the eyelid location [20,24].

CONCLUSION

MALT lymphoma can be slow-growing, and delayed diagnosis can occur due to subtle early symptoms. The eyelid localisation is rare in literature. Regular eye exams, especially for those with risk factors, can help with early detection. When a suspicious lesion is present, an excisional biopsy may be performed for diagnosis. For staging, blood tests—including a blood smear—and imaging studies such as abdominal and chest CT scans are typically conducted.

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Conflicts of Interest: The authors declare no conflicts of interest.

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