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CASE REPORT
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# Case Series of Orbital Lymphoma: Cardinal Presentations

Yordan A. Urrutia, MS*
Tyler Fezza, BS†
Kalie Kosek, PA-C†
John P. Fezza, MD†

**Summary:** Periorbital lymphomas may present with nonspecific symptoms and may disguise as other ocular conditions, which may result in a missed diagnosis. The purpose of his case series is to add to the existing literature and raise awareness of this rare and lethal cancer so early detection and treatment can be instituted. A retrospective review of four diverse cases of periorbital lymphoma is described, highlighting the important role of plastic surgeons in early recognition and prompt diagnosis through extensive examination, imaging studies, and biopsies. The histological subtype and site of lesion play a critical role in the prognosis of patients with periorbital lymphoma. This article also underscores the significance of early detection and a multidisciplinary team approach. *(Plast Reconstr Surg Glob Open 2024; 12:e5913; doi: 10.1097/GOX.0000000000005913; Published online 18 June 2024.)*

Lymphomas are malignant tumors that develop from clonal proliferation of B lymphocytes; T lymphocytes; and, less frequently, natural killer cells. They are divided into Hodgkin lymphoma (HL), which is of B-lymphocyte origin, and non-HL (NHL), which have different clonal origins and consist of B-cell lymphomas, T-cell lymphomas, and non-kill-cell lymphomas.[1] Ocular adnexal lymphoma (OAL) is a form of lymphoma that localizes to the orbit, lacrimal gland, eyelids, and conjunctiva.[2] Most OAL are extranodal NHLs.[3]

Lymphoma is the most common malignancy of the ocular adnexa, but only comprises 5%–10% of all extranodal lymphomas and 1%–2% of all NHLs.[4,5] The frequency of involvement in periorbital sites has been studied to be approximately 46%–74% in the orbit, 20%–33% in the conjunctiva, 5%–10% in the lacrimal gland, and 5%–20% in the eyelid.[6,7] The diagnosis is confirmed through biopsy of the lesion.[7] OAL usually presents in the seventh decade of life and has a female predominance.[8] Many cases are asymptomatic, but the most common presenting signs and symptoms are painless orbital mass with a "salmon-patch" appearance (67%), proptosis (54%), restriction of extraocular motility (39%), conjunctival swelling (22%), and irritation, tearing, headache, itchiness, diplopia, facial numbness, and pressure sensation. B symptoms, such as fever, night sweats, or weight loss, may also be reported.[4,7]

Given the slow-growing nature of OAL, coupled with its asymptomatic nature, lymphoma can easily be misdiagnosed, leading to delays in diagnosis. This series emphasizes the clinical course of identification and workup of four patients with periocular lymphomas and includes cases of primary, isolated, recurrent, and metastatic periorbital lymphoma. This information may aid plastic surgeons in developing a higher degree of suspicion to promptly perform comprehensive examinations, imaging studies, specialist referrals, and biopsies to establish a diagnosis. This report is Health Insurance Portability and Accountability Act compliant, and written consents were obtained.

## CASE PRESENTATIONS

### Patient 1

A 63-year-old woman presented with complaints of a bump on the left upper eyelid for 2.5 years with associated occasional itchiness and pressure worsening for 2 months. An optometrist saw her 8 months prior and advised her to perform warm compresses, with little relief. On examination, there was a mobile left superior orbital mass at the superior fornix causing mild ptosis (Fig. 1). She had a computed tomography (CT) of the orbits showing enlargement of the left lacrimal gland and a slight inferior displacement of the left orbit. The patient underwent an orbitotomy with excision and biopsy, and pathology demonstrated a low-grade B-cell lymphoma. She had a positron-emission tomography-CT (PET-CT) that was unremarkable with no residual abnormalities. The patient

*From the *Florida State University College of Medicine, Tallahassee, Fla.; and †Center for Sight, Sarasota, Fla.*

*Received for publication February 8, 2024; accepted May 1, 2024.*

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DOI: 10.1097/GOX.0000000000005913

Disclosure statements are at the end of this article, following the correspondence information.

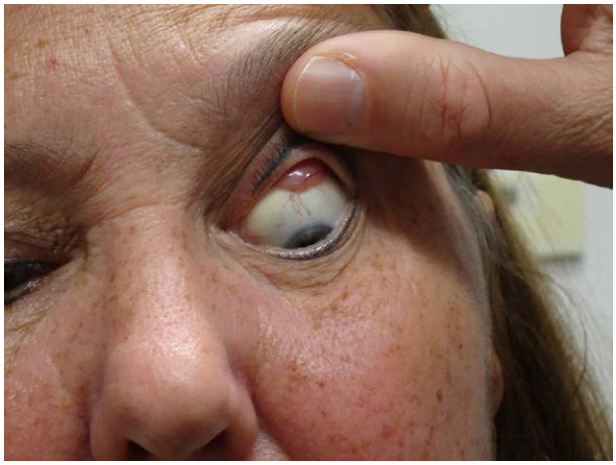

**Fig. 1.** A 63-year-old woman with a lesion in the superior fornix seen on left upper eyelid elevation.

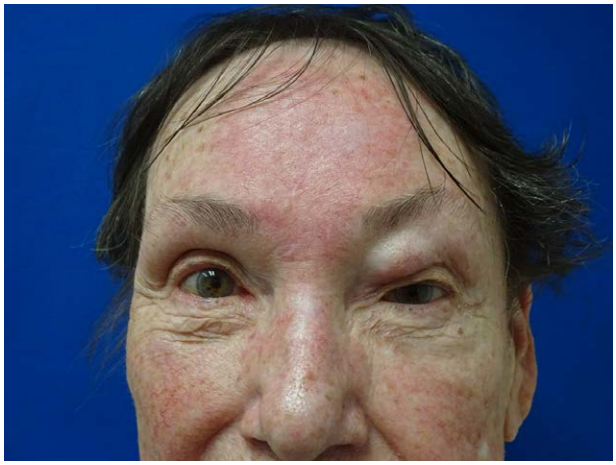

**Fig. 2.** A 78-year-old woman with left upper eyelid mass for 1–2 months without reported pain or visual disturbances.

completed radiation treatment to the left orbit and achieved remission.

**Patient 2**

A 78-year-old woman presented for a bump on the left upper eyelid for 2 months with no discomfort. Physical examination revealed a firm, mobile left upper eyelid mass with no skin breakdown (Fig. 2). She underwent a CT of the orbits revealing a well-circumscribed, soft-tissue mass superior to the equator of the left globe measuring 1×2 × 1.4 cm concerning for neoplasm. The patient underwent a left upper eyelid medial anterior orbitotomy with lesion removal, and pathology was consistent with follicular lymphoma. The PET-CT scan showed diffuse cervical, submandibular, and axillary lymphadenopathy. She completed a course of radiation therapy to the left upper eyelid region. Ten months later, the patient presented with a mass behind the right ear. On examination, she had numerous palpable anterior cervical lymph nodes and a 2.5×2 cm firm postauricular mass. She followed up with her oncologist for further management for metastatic disease.

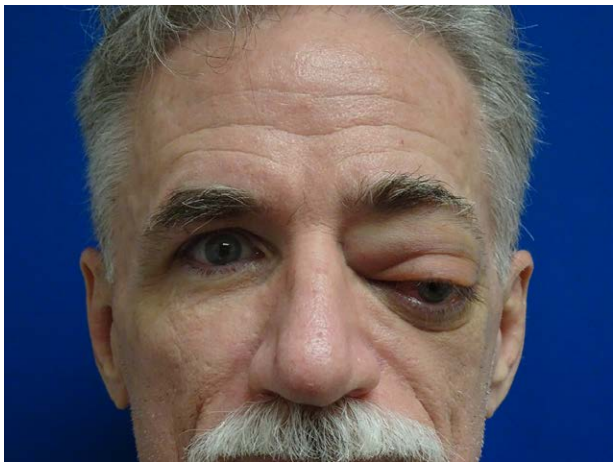

**Fig. 3.** A 62-year-old man with history of tearing and swelling of left eye for 7 months with proptosis, hypoglobus, and left medial orbital mass.

**Patient 3**

A 62-year-old man presented with left eye tearing and progressive swelling for 7 months. He had been prescribed Visine drops, warm compresses, and antibiotics with no relief. Examination of the left eye was significant for proptosis, hypoglobus, and fullness and induration of the medial superior orbit, associated with scleral corkscrew vessels (Fig. 3). Magnetic resonance imaging (MRI) revealed an orbital, extraconal mass superiorly measuring approximately 4.8×3.7×3.5 cm resulting in mass effect upon the left globe with severe stretching of the optic nerve. Findings were suggestive of orbital lymphoma with concerns for permanent vision loss secondary to optic nerve stretching. The patient underwent urgent left anterior orbitotomy with lesion biopsy, and pathology revealed a B-cell lymphoma. He completed 16 treatments of radiation to the left orbit with remission.

**Patient 4**

A 69-year-old man presented with painless eyelid swelling for 3 months associated with irritation, and "crusty" discharge in the left eye. Two years prior, he was diagnosed with mantle cell lymphoma involving the ileum and colon. On examination, he had left eye ptosis and a 5.5-mm firm, vascularized cyst in the left upper eyelid nasally, with scaling and crusting of the skin (Fig. 4). He underwent MRI of the brain and orbits showing a left orbital enhancing 3.2×2.5×2.8 cm soft-tissue mass involving both the preseptal and postseptal spaces. He had an excisional biopsy supporting the diagnosis of recurrent mantle cell lymphoma. He underwent radiation treatment to the left orbit.

## DISCUSSION

Periocular lymphomas often present with slowly progressive symptoms.[5] Obtaining a biopsy is essential, as

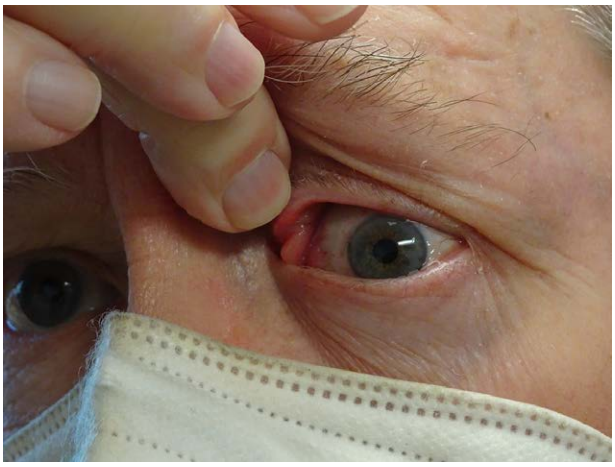

**Fig. 4.** Eversion of the medial left upper eyelid showing a pink, fleshy lesion in a 69-year-old man presenting with painless eyelid swelling.

histopathologic confirmation remains the gold standard for diagnosis. Staging of orbital lymphoma involves a full systemic workup by an oncologist, laboratory studies, PET-CT or MRI, and bone marrow biopsy.[5,7]

Prognosis is intricately linked to histological subtype and site of the lesion, with approximately 35% of orbital lymphoma developing into systemic lymphoma after 4 years.[9,10] Several treatment options include chemotherapy, surgical removal, and radiation, with radiation being the preferred treatment in most cases.[9]

The clinical presentations of periocular lymphomas can be diverse and nonspecific, which may result in delayed diagnosis.[9] Clinicians should maintain a high level of suspicion for orbital lymphomas in patients with orbital masses. Awareness of the varied presentations can lead to early detection, treatment, and improved overall prognosis.

***Yordan A. Urrutia, MS***
Florida State University College of Medicine
1115 W Call Street
Tallahassee, FL 32304
E-mail: yau13@med.fsu.edu

## DISCLOSURE

*The authors have no financial interest to declare in relation to the content of this article.*

## PATIENT CONSENT

*Patient provided written consent for the use of their images.*

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