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Ocular adnexal lymphoma presenting as incidental tarsal follicles

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ABSTRACT

Purpose: To report a case of ocular adnexal lymphoma presenting as asymptomatic follicles discovered incidentally on routine examination.

Observations: A 43-year-old woman presented for routine annual examination and was incidentally found to have unilateral giant follicles in the left eye inferior fornix. She denied any ocular or systemic symptoms. The remainder of the examination was unremarkable, and the patient was otherwise healthy. A conjunctival biopsy revealed a diagnosis of mucosa-associated lymphoid tissue (MALT) lymphoma. She underwent external beam radiation therapy, resulting in complete resolution of the follicles.

Conclusions and Importance: Awareness of atypical presentations of conjunctival lymphoma and thorough slit lamp examinations, even on routine exams, may help expedite diagnosis and treatment.

1. Case report

A 43-year-old healthy woman with no medical or ocular history presented for her annual routine eye examination and was incidentally found to have giant follicles in the left eye inferior fornix extending onto the nasal palpebral conjunctiva (Fig. 1). She denied any ocular redness, swelling, discharge, irritation, pain, foreign body sensation, ptosis, or changes in vision, and had not noticed any ocular lesions. She had not experienced any fevers, chills, malaise, or unintentional weight loss. The examination was otherwise unremarkable. Lymphadenopathy was absent. The follicles did not resolve with topical antibiotics, topical steroids or oral doxycycline. Chlamydia, Herpes Simplex Virus and bacterial swabs were negative. Conjunctival biopsy was performed. A formalin-fixed sample stained with hematoxylin-eosin showed lymphomatous infiltration on light microscopy. Immunohistochemical staining of a fresh tissue specimen revealed infiltrating cells expressing CD20 and BCL2 with lambda light chain restriction; BCL6 and Cyclin D1 were negative. These findings confirmed a diagnosis of ocular adnexal mucosa-associated lymphoid tissue (MALT) lymphoma. The patient was referred to medical oncology for systemic evaluation, including computed tomography scans of the chest, abdomen and pelvis, total body positron emission tomography, brain magnetic resonance imaging, complete blood count, serum chemistry panel, and erythrocyte sedimentation rate, which revealed no abnormal findings. She underwent external beam radiation therapy (EBRT) to the inferior fornix (25.2 Gy) and nasal palpebral conjunctiva (29.2 Gy), resulting in complete resolution of the follicles. She remained recurrence-free over the ensuing twelve years.

2. Discussion

Ocular non-Hodgkin's lymphomas (NHL) are a group of heterogeneous malignancies that comprise approximately 8% of all extranodal NHLs.[1] The most common histologic subtype is MALT lymphoma, which constitutes approximately 80% of conjunctival B-cell NHLs.[1] The condition classically presents as a painless, “salmon-pink” patch on the conjunctiva.[2] Prompt detection is imperative, as nearly 20% of patients who present with localized conjunctival lymphoma will eventually develop disseminated disease.[3] Treatment with EBRT is typically effective in controlling disease—studies have reported conjunctival MALT lymphoma recurrence rates from 0 to 11% following radiation therapy.[4,5] Unfortunately, misdiagnosis and delayed treatment are common, as conjunctival lymphoma frequently mimics other benign entities, including lymphoid hyperplasia and chronic conjunctivitis.[2]

In published literature, patients are frequently described as presenting with nonspecific symptoms, such as swelling, irritation, redness, decreased visual acuity, or ptosis, and typically note a lesion of concern.[6] In a large multicenter study, Kirkegaard et al. reported that 90% of patients with conjunctival MALT lymphoma presented with the feeling of a “tumor” or swelling and 27% reported irritation or pain.[6] Furthermore, patients experienced these symptoms for a median duration of 6 months prior to presentation.[6] Here, we report a case of

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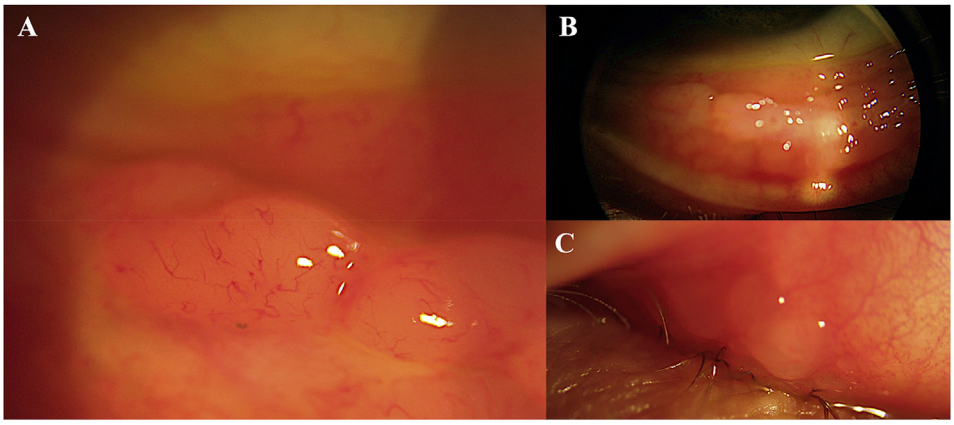

Fig. 1. Photographs showing large follicles of the left inferior tarsal conjunctiva (A,B) and inferior fornix (C).

incidental detection of conjunctival lymphoma on a routine examination. In contrast to prior literature, our patient was not aware of the large conjunctival follicles and denied any ocular or systemic symptoms. It is possible that she was examined at an earlier stage of disease prior to symptom manifestation, or perhaps this presentation was atypical in its lack of even mild symptoms despite the development of sizable conjunctival follicles.

3. Conclusions

This case is a reminder of the variation in presentations of conjunctival MALT lymphoma and the importance of a thorough slit lamp examination, including eyelid eversion and examination of the fornices, even in seemingly low-risk asymptomatic patients. Awareness of atypical presentations may help expedite diagnosis and avoid delayed treatment.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

CRediT authorship contribution statement

Aneesha Ahluwalia: Conceptualization, Writing - original draft, Writing - review & editing, Visualization. **Paula W. Feng:** Conceptualization, Writing - review & editing. **Seth W. Meskin:** Conceptualization, Writing - review & editing, Supervision.

Declaration of competing interest

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