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CASE REPORT

Two Cases Presenting with Unilateral Ptosis at an Advanced Age: Primary Lymphoma of the Lacrimal Gland

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Abstract

Lesions of the lacrimal gland constitute one of the subgroups of orbital masses and can have benign or malignant characteristics. They are evaluated in two main histopathological groups as epithelial and non-epithelial, with lymphomas belonging to the non-epithelial group. Lacrimal gland lymphomas have been analyzed under the orbital lymphomas header in most of the literature studies and there are only a few articles evaluating these lesions separately. The aim of this study was to present the clinical, radiological, and histopathological evaluation and related findings of two cases with a confirmed biopsy diagnosis of primary lacrimal gland lymphoma that presented with unilateral ptosis at an advanced age. In conclusion, primary lacrimal gland lymphomas are limited to the orbit and can appear without any systemic findings. The possibility of lacrimal gland lymphoma should be considered especially in patients at an advanced age presenting with unilateral ptosis. Organic pathologies that could cause ptosis must be ruled out before planning ptosis surgery.

Keywords: Lacrimal gland lesions; lymphoma; ptosis; orbital tumours.

Orbital masses can originate from different tissues in different localizations in the orbit[1-16]. Lacrimal gland masses are also one of the subgroups of orbital lesions, and can be benign or malignant[5-16]. Histopathologically, they are evaluated in two main groups as epithelial and non-epithelial lesions, and lymphomas are examined in the group of non-epithelial lesions[5-11]. Lymphomas are malignant and have heterogeneous features both histopathologically and clinically. Lymphoid infiltration of the eye is often seen in the orbit; involvement of eyelids, conjunctiva, lacrimal gland, optic nerve and uveal tissue can also be observed rarely[1-16].

In studies on orbital lymphoma in the literature, subgroups were mostly analyzed together, and there are very few publications evaluating lacrimal gland lymphomas only. Therefore, we aimed to present the clinical, radiological and histopathological findings of two elderly patients who presented with unilateral ptosis and whose primary lacrimal gland lymphoma diagnosis was confirmed by biopsy.

Case Report

Case 1 – A 71-year-old male patient was admitted to our outpatient clinic because of ptosis in the right eye that developed six months ago. He had no known history of ocular

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Figure 1. (a) Upper eyelid fullness with ptosis in the patient's right eye. **(b)** Coronal section. **(c)** Axial section. **(d)** Image of intraoperative lacrimal mass. **(e)** Atypical lymphoid infiltrates of 1.5-2 times the size of a mature lymphocyte, with large hyperchromatic nuclei, small nucleoli in between, most of them showing monotonous atypia, some of them in centroblastic morphology (H&E, X200).

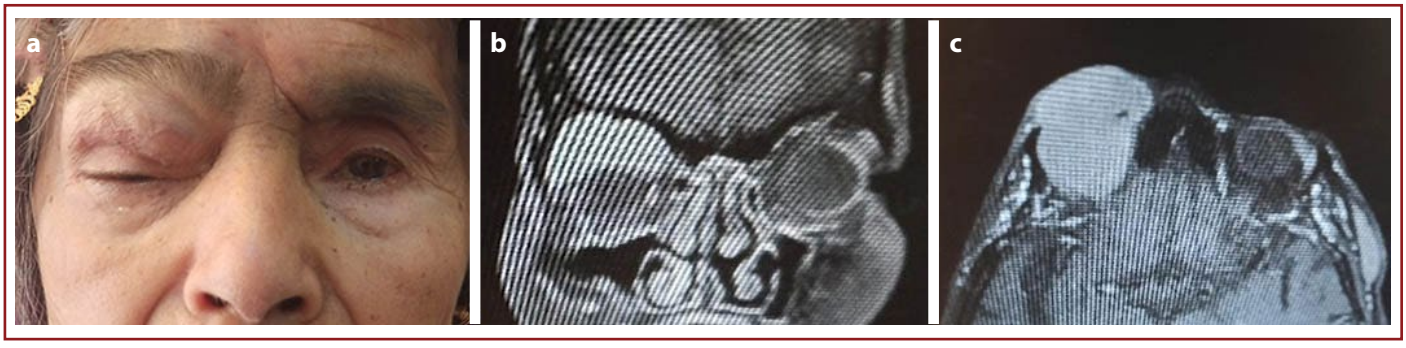

Figure 2. (a) Image of a mass that completely fills the upper eyelid with severe ptosis in the right eye of the patient. **(b)** Coronal section. **(c)** Axial section.

or systemic disease. In the examination, there was ptosis in the right eye and fullness in the upper eyelid compared to the left eye (Fig. 1a). Since there was a palpable mass in the upper temporal region on palpation, radiological imaging was requested. In orbital imaging, a 27*12 mm lesion with soft tissue density was detected in the superolateral neighborhood of the globe in the right eye (Fig. 1b,1c). The patient was informed and orbitotomy was planned. The mass was excised subtotally by lateral orbitotomy, eyelid crease incision (Fig. 1d). A diagnosis of marginal zone B-cell lymphoma was made as a result of histopathological examination (Fig. 1e). No metastases were detected in the systemic screening. Local external radiotherapy was additionally applied, and no recurrence was observed in the last control examination.

Case 2 – A 92-year-old female patient was admitted to our outpatient clinic due to a feeling of drooping and fullness in the upper eyelid that developed one year ago in the right eye. There was no known history of ocular or systemic disease other than previous cataract surgery. In the examination of the patient, there was a mass that completely filled the upper eyelid with severe ptosis in the right eye (Fig. 2a). In orbital imaging, a lesion measuring 40*32 mm completely filling the superior part of the globe was observed in the right eye (Fig. 2b, 2c). The orbitotomy was planned by informing the patient and her relatives. The mass was excised with a superior orbitotomy, eyelid crease incision (Fig. 3a-c). Diffuse large B-cell lymphoma was diagnosed as a result of histopathological examination (Fig. 3d). Since metastasis was detected in the systemic screening,

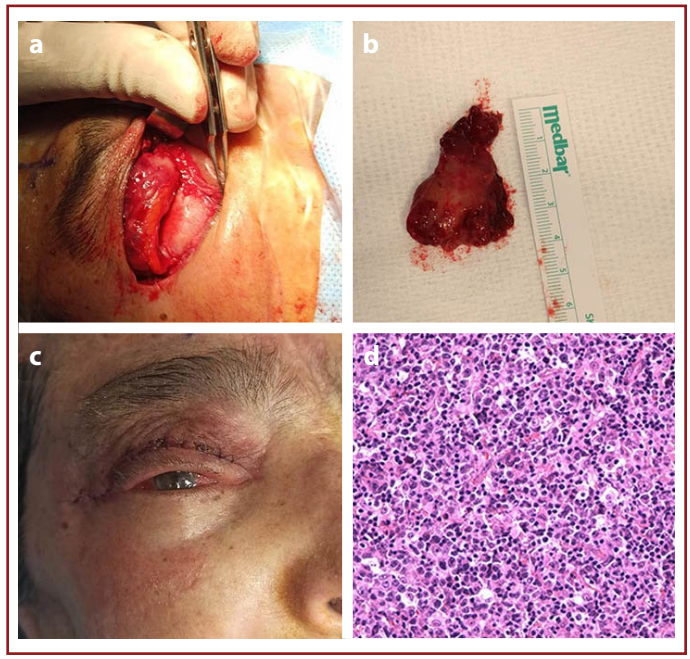

Figure 3. (a) Image of intraoperative lacrimal mass. **(b)** Image of the excised mass. **(c)** View of completed surgery. **(d)** Pleomorphic atypical lymphoid infiltrates of 2.5-3 times the size of a mature lymphocyte, with large hyperchromatic nuclei, prominent nucleoli, and irregular nucleolemmas (H&E, X200).

an additional 6 cycles of chemotherapy was applied to the patient and no recurrence was observed in the last control examination.

Discussion

Orbital lymphomas can originates in the conjunctiva, lacrimal gland, eyelid, soft connective tissue and extraocular muscles[1-11]. In previous studies, orbital lymphoma cases were generally evaluated together, and studies evaluating only lacrimal gland lymphomas are very few. The most important reason for this is the low number of cases in a single center. For this reason, researchers recently published an international multicenter study of 260 patients on lacrimal gland lymphomas[6]. In this study, the mean age of the patients was 58 years, and 258 cases (99%) had Non-Hodgkin B-cell lymphoma. The most common subtype was reported as marginal zone B-cell lymphoma (68%), which is in the low-grade lymphoma class, and radiotherapy is preferred for its treatment, as in one of our cases. In the same study, the frequency of diffuse large B-cell lymphoma was reported as 10%, which is in the high-grade lymphoma class, and chemotherapy is generally preferred for its treatment, as in one of our cases. It has also been combined with rituximab and/or radiotherapy in cases with inadequate response to chemotherapy[6]. In this study, it was emphasized that the histopathological subtype of lacrimal gland lymphoma was the most important factor in determining survival, and it was reported that marginal zone B-cell lymphoma had the best prognosis and diffuse large B-cell lymphoma had the worst prognosis[6]. Karadeniz Uğurlu and Garrity, in their study consisting of 45 patients with lacrimal gland involvement, reported that lymphoma responds very well to treatment in terms of ocular symptoms and signs, but it is a lymphoma group that needs long-term follow-up due to the risk of developing systemic lymphoma[7].

In conclusion, primary lacrimal gland lymphomas are limited to the orbit and may occur without any systemic findings. The possibility of lacrimal gland lymphoma should be considered, especially in elderly patients who present with the complaint of unilateral drooping of the eyelid. Before planning ptosis surgery, organic pathologies that may cause droopy eyelid must be ruled out.

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References

1. Gündüz K, Esmaeli B. Diagnosis and management of malignant tumors of the eyelids, conjunctiva and orbit. Expert Review Ophthalmol 2008;3:63–75. [CrossRef]
2. Soysal HG, Aköz A, Ardıç F. Lymphomas of the orbit and ocular adnexa. Turk J Ophthalmol 2008;38:69–73.
3. Pazarlı H, Yolar M, Yigitsubay U, Oguz V, Ferhanoglu B. Lymphomas of the orbita and ocular adnexa. Turk J Ophthalmol 2003;33:435–40.
4. Tunç M. Current diagnostic and management methods in orbital tumors. Turk J Ophthalmol 2014;44:1–7. [CrossRef]
5. Dürük K, Günalp İ, Şimşek T. Gözyaşı bezi malign tümörleri. T Klin Oftalmoloji 1997;6:89–93.
6. Vest SD, Mikkelsen LH, Holm F, Rasmussen PK, Hindso TG, Knudsen MKH, et al. Lymphoma of the lacrimal gland - an ınternational multicenter retrospective study. Am J Ophthalmol 2020;219:107–20. [CrossRef]
7. Karadeniz Uğurlu S, Garrity JA. Ocular adnexal lymphoma with lacrimal gland involvement. T Klin Oftalmoloji 2000;9:250–5.
8. Çalış F, Gündüz K, Kuzu I, Erden E. Clinical findings and treatment results in ocular adnexal lymphomas. Turk J Ophthalmol 2014;44:374–8. [CrossRef]
9. Çoban Karataş M, Altan Yaycıoğlu R, Aydın Akova Y. Intraorbital non-hodgkin lymphoma: A case report. Turk J Ophthalmol 2011;41:118–20. [CrossRef]
10. İlhan HD, Türkoğlu EB, Bilgin AB, Akkaya B, Ünal M. Marginal zone B cell lymphoma of lacrimal gland secondary to sjogren's syndrome. Turk J Ophthalmol 2014;44:322–4. [CrossRef]
11. Yalçın Tök Ö, Tök L, Akbaş Kocaoğlu F, Yalnız Z, Örnek F. Involvement of bilateral lacrimal gland in systemic Hodgkin disease. Dicle Med J 2010;37:67–70.
12. Gündüz K, Shields CL, Günalp I, Shields JA. Magnetic resonance imaging of unilateral lacrimal gland lesions. Graefes Arch Clin Exp Ophthalmol 2003;241:907–13. [CrossRef]
13. Buttanrı İB, Serin D, Akbaba M, Karslıoğlu Ş, Özkara S. Biopsy results in patients ınitially diagnosed as ıdiopathic orbital ınflammatory syndrome. Turk J Ophthalmol 2014;44:301–5. [CrossRef]
14. Kiratli H, Bilgiç S. An unusual clinical course of adenoid cystic carcinoma of the lacrimal gland. Orbit 1999;18:197–201. [CrossRef]
15. Acar Aydemir E, Karadeniz Uğurlu Ş, Yurdakul NS, Maden A, Rezanko T. Orbita dışı yayılım gösteren idiopatik orbital inflamatuar hastalık. T Oft Gaz [Article in Turkish] 2005;35:359–64.
16. Zor RK, Küçük E, Tutaş Günaydın N, Önder AF. Bilateral primary lacrimal gland lymphoma. Med Bull Haseki 2016;54:246–8.