Review Article

Journal of Optometry and Ophthalmology Research

Lacrimal Sac Lymphomas: Clinical Presentation, Diagnostic Challenges, and Management – A Focused Review with Institutional Experience

Dalal Fatani

King Khaled Eye Specialist Hospital, Oculoplastics & Orbit Division, Saudi Arabia

Corresponding author

Dalal Fatani, King Khaled Eye Specialist Hospital, Oculoplastics & Orbit Division, Saudi Arabia.

Received: February 25, 2026; **Accepted:** March 04, 2026; **Published:** March 11, 2026

ABSTRACT

Purpose: To provide an updated focused review of lacrimal sac lymphomas, highlighting epidemiology, clinical presentation, diagnostic workup, histopathological subtypes, management strategies, and outcomes, supplemented by institutional experience.

Methods: Narrative literature review of lacrimal sac lymphomas with contextual reference to cases identified at a tertiary eye hospital.

Results: Lacrimal sac lymphomas are rare lesions of the lacrimal drainage system, most commonly diffuse large B-cell lymphoma (DLBCL) and mucosa-associated lymphoid tissue (MALT) lymphoma. Presentation frequently mimics primary acquired nasolacrimal duct obstruction, leading to diagnostic delay. Cross-sectional imaging may show a solid lacrimal sac mass with possible extension to adjacent structures. Radiotherapy is commonly used for localized disease, while systemic therapy is employed for high-grade histologies or disseminated involvement.

Conclusions: Lacrimal sac lymphomas require a high index of suspicion in atypical, recurrent, or unilateral lacrimal outflow obstruction. Timely biopsy with appropriate immunophenotyping and multidisciplinary care are essential for optimal outcomes.

Keywords: Lacrimal Sac Lymphoma, Lacrimal Drainage System, Nasolacrimal Duct Obstruction; DLBCL, MALT Lymphoma, Ocular Adnexal Lymphoma

Introduction

Primary and secondary lymphomas of the lacrimal drainage system are rare non-epithelial malignancies that may arise from lymphoid tissue surrounding the lacrimal sac mucosa. The lacrimal sac mucosa contains lymphoid tissue and is lined by non-keratinized stratified columnar epithelium, providing a substrate for lymphoproliferative disease [1]. Lacrimal sac tumors are uncommon overall, and lymphomas comprise a minority of malignant lacrimal sac lesions in published series [2-6,14-16]. A key clinical issue is that lacrimal sac lymphomas frequently present with symptoms typical of benign nasolacrimal duct obstruction, resulting in misdiagnosis and delayed definitive workup [2,14,16,26].

Epidemiology and Pathogenesis

Reported lacrimal sac lymphomas occur most often in older adults, although pediatric and young adult cases have been described [11,16-23]. Across reports, B-cell lymphomas predominate, particularly DLBCL and MALT lymphoma [10,14-16]. Rare histologies have also been reported in recent literature, including mantle cell lymphoma and Burkitt lymphoma involving the lacrimal drainage system [27,28]. Chronic antigenic stimulation of mucosa-associated lymphoid tissue is a proposed mechanism for extranodal marginal zone (MALT) lymphoma within ocular adnexal structures [14,29].

Clinical Presentation and Red Flags

Patients typically present with epiphora, medial canthal swelling, or recurrent dacryocystitis—features that overlap with primary acquired nasolacrimal duct obstruction [2,9,10,14]. Hemolacria is classically considered concerning but is not consistently present [9,11]. Less common features such as globe dystopia, proptosis, diplopia, or hypoesthesia may suggest aggressive disease or extension beyond the sac [12,13,26,30]. Clinical clues that should prompt imaging and biopsy include: firm noncompressible mass, rapid progression, recurrent/

Citation: Dalal Fatani. Lacrimal Sac Lymphomas: Clinical Presentation, Diagnostic Challenges, and Management – A Focused Review with Institutional Experience. J Opto Opht Res. 2026. 2(1): 1-3. DOI: doi.org/10.61440/JOOR.2026.v2.06

unilateral dacryocystitis, bloody discharge/hemolacria, palpable lymphadenopathy, or atypical intraoperative sac appearance during dacryocystorhinostomy (DCR) [7,14,16].

Imaging and Diagnostic Workup

CT is frequently used to confirm a lacrimal sac mass and assess bony changes; MRI provides superior characterization of soft tissue and extension into orbit or sinonasal spaces [2]. Dacryocystography may demonstrate filling defects but generally does not replace cross-sectional imaging when a mass is suspected [2]. Because imaging may not reliably distinguish inflammatory lesions from lymphoma, tissue diagnosis is required. Biopsy should be planned to obtain adequate specimen for histology, immunohistochemistry, and, when needed, flow cytometry [14,16]. Routine lacrimal sac biopsy at DCR remains debated, but targeted biopsy is strongly supported in suspicious or atypical cases [7].

Histopathology and Immunophenotype

DLBCL and MALT lymphoma are the most frequently reported subtypes of lacrimal sac lymphoma [10,14-16]. DLBCL is an aggressive lymphoma that typically warrants systemic staging and systemic therapy; MALT lymphoma is often indolent and may be localized at presentation [14,16,29]. Accurate classification depends on immunophenotyping and correlation with systemic evaluation. Distinguishing lymphoma from lymphoid hyperplasia is essential, as benign lymphoid hyperplasia may also occur in the lacrimal sac [4,14].

Management

Management is guided by histologic subtype and disease stage. For localized indolent disease, radiotherapy is commonly employed and can achieve durable local control [14,16]. High-grade lymphomas (e.g., DLBCL) and disseminated disease generally require systemic chemoimmunotherapy; selected cases may use combined modality therapy [10,14,16]. Surgery is primarily diagnostic (biopsy) or occasionally used for debulking/relief of obstruction; definitive oncologic surgery is not standard for lymphoma [14,16]. Importantly, DCR performed before diagnosis may create a conduit to the nasal cavity; therefore, suspected sac tumors should undergo imaging and biopsy prior to DCR whenever possible [16].

Recent Literature Updates

Recent case reports and reviews continue to emphasize frequent initial misdiagnosis and the importance of early biopsy. A 2024 report described lacrimal sac DLBCL presenting with sudden-onset binocular diplopia, underscoring that atypical neuro-ophthalmic symptoms may occur and should prompt evaluation for lacrimal sac malignancy [26]. Additional recent case reports describe unusual patterns of spread (e.g., infraorbital nerve involvement and sinonasal extension) and rare histologies involving the lacrimal drainage system [27,28,30]. A 2025 histotype-driven review of lacrimal sac tumors provides updated context on diagnostic delay and multidisciplinary management across histologies [31].

Conclusion

Lacrimal sac lymphomas are rare but clinically important causes of lacrimal outflow obstruction. Because symptoms often mimic benign disease, clinicians should maintain suspicion in atypical or recurrent unilateral presentations and obtain timely imaging and biopsy. Accurate histopathologic classification, systemic staging, and multidisciplinary management are central to optimizing outcomes.

Table 1: Reported pediatric cases of lacrimal sac lymphoma in the literature

First author (Ref)	Age (years)	Subtype	Management
Carlin et al [20]	10	Poorly differentiated lymphoma	External beam radiotherapy
Schefler et al [17]	10	MALT lymphoma	Surgical excision + chemotherapy
Köksal et al [19]	9	DLBCL	Systemic chemotherapy
Parikh et al [21]	13	DLBCL	Systemic chemotherapy
Meng et al [16]	13	MALT lymphoma	Radiotherapy
Ucgul et al [22]	13	MALT lymphoma	Surgical excision + radiotherapy + chemotherapy
Ucgul et al [22]	12	MALT lymphoma	Radiotherapy

Abbreviations: DLBCL, diffuse large B-cell lymphoma; MALT, mucosa-associated lymphoid tissue lymphoma.

References

1. Ali MJ, Paulsen F. Human Lacrimal Drainage System Reconstruction, Recanalization, and Regeneration. Curr Eye Res. 2020. 45: 241-252.
2. Krishna Y, Coupland SE. Lacrimal Sac Tumors--A Review. Asia Pac J Ophthalmol (Phila). 2017. 6: 173-178.
3. Heindl LM, Jünemann AG, Kruse FE, Holbach LM. Tumors of the lacrimal drainage system. Orbit. 2010. 29: 298-306.
4. Pe'er JJ, Stefanyszyn M, Hidayat AA. Nonepithelial tumors of the lacrimal sac. Am J Ophthalmol. 1994. 118: 650-658.
5. Stefanyszyn MA, Hidayat AA, Pe'er JJ, Flanagan JC. Lacrimal sac tumors. Ophthalmic Plast Reconstr Surg. 1994. 10: 169-184.
6. Ryan SJ, Font RL. Primary epithelial neoplasms of the lacrimal sac. Am J Ophthalmol. 1973. 76:73-88.
7. Koturović Z, Knežević M, Rašić DM. Clinical significance of routine lacrimal sac biopsy during dacryocystorhinostomy: A comprehensive review of literature. Bosn J Basic Med Sci. 2017. 17: 1-8.
8. Ramberg I, Toft PB, Heegaard S. Carcinomas of the lacrimal drainage system. Surv Ophthalmol. 2020. 65: 691-707.
9. Singh S, Ali MJ. Primary Malignant Epithelial Tumors of the Lacrimal Drainage System: A Major Review. Orbit. 2021. 40: 179-192.
10. Kadir SMU, Rashid R, Sultana S, Nuruddin M, Nessa MS, et al. Lacrimal Sac Tumors: A Case Series. Ocul Oncol Pathol. 2022. 8: 42-51.
11. Wakasaki T, Yasumatsu R, Tanabe M, Yoshikawa H, Jiromaru R, et al. Lacrimal Sac Tumors: A Single-institution

Experience, Including New Insights. In Vivo. 2023. 37: 1219-1225.
12. Anderson NG, Wojno TH, Grossniklaus HE. Clinicopathologic findings from lacrimal sac biopsy specimens obtained during dacryocystorhinostomy. Ophthalmic Plast Reconstr Surg. 2003. 19: 173-176.
13. Radnót M, Gáll J. Tumors of the lacrimal sac. Ophthalmologica. 1966. 151: 2-22.
14. Singh S, Ali MJ. Lymphoproliferative tumors involving the lacrimal drainage system: a major review. Orbit. 2020. 39: 276-284.
15. Kim J, Baek S. Diffuse Large B-Cell Lymphoma Arising in the Lacrimal Sac. J Craniofac Surg. 2022. 33: e19-e21.
16. Meng FX, Yue H, Yuan YQ, Zhang R, Yuan YF, et al. Lacrimal sac lymphoma: a case series and literature review. Int J Ophthalmol. 2022. 15: 1586-1590.
17. Schefler AC, Shields CL, Shields JA, Demirci H, Maus M, Eagle RC Jr. Lacrimal sac lymphoma in a child. Arch Ophthalmol. 2003. 121: 1330-1333.
18. Levitt AE, Choi CJ, Johnson TE. Bilateral Leukemic Infiltration of the Lacrimal Sac in a Child. Ophthalmic Plast Reconstr Surg. 2021. 37: e28-e30.
19. Köksal Y, Kiratli H, Varan A, Akçören Z, Büyükpamukçu M. Primary lacrimal sac non-Hodgkin's lymphoma in a child. Int J Pediatr Otorhinolaryngol. 2005. 69: 1551-1553.
20. Carlin R, Henderson JW. Malignant lymphoma of the nasolacrimal sac. Am J Ophthalmol. 1974. 78: 511-513.
21. Parikh D, Rodgers R, Kodsi S. Primary lacrimal sac diffuse large B-cell lymphoma in a child. J AAPOS. 2019. 23: 53-55.
22. Ucgul AY, Tarlan B, Gocun PU, Konuk O. Primary mucosa-associated lymphoid tissue (MALT) lymphoma of the lacrimal drainage system in two pediatric patients. Eur J Ophthalmol. 2020. 30: 18-23.
23. Gokce G, Ceylan OM, Uysal Y, Yildizoglu U, Atas E, et al. Epiphora as the presenting sign of relapsed non-Hodgkin lymphoma in a child with Bruton agammaglobulinemia. Eur J Ophthalmol. 2015. 25: 65-67.
24. Owens RM, Wax MK, Kostik D, Linberg JV, Hogg J. Malignant melanoma of the lacrimal sac. Otolaryngol Head Neck Surg. 1995. 113: 634-640.
25. Song X, Wang J, Wang S, Wang W, Wang S, et al. Clinical analysis of 90 cases of malignant lacrimal sac tumor. Graefes Arch Clin Exp Ophthalmol. 2018. 256: 1333-1338.
26. Jeri Yabar A, Vittini Hernandez L, Dev B, Patel M, Lin M. Lacrimal sac diffuse large B-cell lymphoma presenting as sudden-onset binocular diplopia. Eur J Case Rep Intern Med. 2024. 11.
27. Bothra N, Ali MJ. Mantle cell lymphoma of the lacrimal drainage system. Saudi J Ophthalmol. 2025. 39.
28. Yeoh SH, Khaliddin N, Talib N, Saravanamuthu K. Primary lacrimal sac Burkitt lymphoma: A case report and literature review. Am J Ophthalmol Case Rep. 2025. 37:102238.
29. Chung HU, Son JH. Ocular adnexal mucosa-associated lymphoid tissue lymphoma. Yeungnam Univ J Med. 2021. 38: 91-103.
30. Sadhar B, Slupek D, Steehler A, Denne C, Steehler K. Primary diffuse large B-cell lymphoma of the lacrimal sac with maxillary sinus and infraorbital nerve involvement. J Surg Case Rep. 2024. 2024: rjae453.
31. Locatello LG, Redolfi De Zan E, Marzolino R, Sowerby LJ, Tarantini A, et al. Lacrimal Sac Tumors: A Histotype-Driven Literature Review. Cancers. 2025. 17: 3718.