



Case report

Cystic mucosa-associated lymphoid tissue lymphoma of lacrimal gland associated with vision loss: A case report



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ABSTRACT

Purpose: To report an atypical case of cystic extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) of lacrimal gland associated with vision loss.

Observations: An 89-year-old woman was presented with a rapidly progressing proptosis, lagophthalmos, and vision loss. Endophthalmitis was also present. Computed tomography scan images showed a hyperdense mass with hypodense cystic areas occupying the superolateral orbit, which displaced the globe antero-inferiorly with optic nerve compression and stretching. An erosion to the adjacent superior and lateral orbital walls was also demonstrated. Complete tumor excision was performed via upper transconjunctival orbitotomy concurrently with enucleation. The immunohistopathological diagnosis was MALT lymphoma.

Conclusions and importance: This case emphasizes the importance of considering lymphoma in the differential diagnosis of a cystic superolateral orbital mass.

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1. Introduction

Malignant lymphoma is a neoplasm derived from clonal proliferation of lymphocytes which leads to formation of solid tumors.¹ It is the most common malignant tumor of the orbit in patients older than 60 years.¹ Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) is a low grade lymphoma and it is the most frequently observed subtype in the orbit.² It often arises from the lacrimal gland, which usually presents as indolent, slow-growing, solid mass without a cystic lesion, or bone erosion or destruction.^{3,4} This study reports a case of lacrimal gland MALT lymphoma with atypical presentation, which showed a rapid enlargement of cystic lesion, associated with vision loss.

1.1. Case report

An 89-year-old woman presented with a right upper eyelid palpable mass for 6 months, which was rapidly enlarging in a month. The patient had severe dementia.

At the first examination, her visual acuity was no light perception and direct pupillary light reflex was lost in the right eye. The mass was firmly elastic with a smooth surface. There was proptosis and inferior displacement of the globe. Lagophthalmos was present. Ophthalmologic examination showed chemosis, corneal ulcer in the interpalpebral zone and hypopyon in the right eye (Fig. 1). Contrast-enhanced computed tomography (CT) showed a hyperdense, well-defined mass with a hypodense cystic area with enhanced wall and septa measuring 35 × 40 × 45mm in the superolateral orbit (Fig. 2). The mass followed the globe contour, but there was globe displacement with optic nerve compression and stretching. An erosion to the adjacent superior and lateral orbital walls was also demonstrated. Pre-operative differential diagnosis was schwannoma or adenoid cystic carcinoma, with associated endophthalmitis secondary to exposure keratopathy and subsequent corneal ulceration.

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Fig. 1. Pre-operative photograph. This shows a mass in the upper eyelid with proptosis, inferior globe displacement, chemosis and hypopyon of the right eye.

She underwent tumor excision via superior transconjunctival orbitotomy. A pale, pinkish, multi-lobulated mass was encountered. Rupture of the mass expressed non-viscous fluid. Although intra-operative frozen section demonstrated the diagnosis of malignant lymphoma, the tumor was removed completely (Fig. 3) to eliminate necessity of additional management for the orbital lesion. Enucleation was also performed after tumor excision.

Post-operative histopathological examination revealed many lymphocytes and plasma cells infiltrating the lacrimal gland, with few germinal centers. The cyst consisted of dilated ducts with limpho-epithelial lesion, containing fluids with lymphocytes, histiocytes and degenerated substances. Tumor cells were positive for CD20, CD79a, and bcl-2, but negative for CD3, CD5, CD10, bcl-6,

cyclin-D1. In situ hybridization showed kappa was greater than lambda (Fig. 4A–C). Immunohistochemical findings supported the diagnosis of MALT lymphoma. Fluid in cysts composed of lymphocytes, histiocytes, and degenerated substances (Fig. 4D). Infiltration of neutrophils in the cornea, anterior chamber, iris and vitreous were present and no involvement of lymphoma.

At the time of last examination, 2 months after surgery, no evidence of recurrence of MALT lymphoma in the orbit was observed. The patient's family refused review of other systemic lesions.

2. Discussion

We presented an atypical case of cystic MALT lymphoma of lacrimal gland associated with vision loss. Lacrimal gland lymphoma is usually identified as diffuse enlargement and axial elongation of the gland, which follows the contour of the globe,⁵ and without producing bone erosion or destruction.⁴ In this case, lymphoma was not considered pre-operatively because of its cystic appearance, aggressive course and bone involvement, although it followed the globe contour. Cystic appearance on CT scan rather raises the differential diagnosis of schwannoma and adenoid cystic carcinoma.⁶ Adenoid cystic carcinoma especially shows aggressive course and bone involvement.⁷

MALT lymphoma rarely appears as cyst in the orbit. A cyst is defined as a closed sac with a distinct membrane compared to the nearby tissue. Whereas, this tumor arises from non-encapsulated clusters of lymphocytes in mucosal tissues and does not form a membrane. The mechanism of cyst-like formation in this case was predominantly due to lacrimal duct obstruction by the tumor, which impeded the outflow and caused accumulation of fluids containing lymphoma cells leading to the atypically rapid enlargement of the tumor.



Fig. 2. Pre-operative (A) axial, (B) sagittal, and (C, D) coronal contrast-enhanced computed tomographic images. A superolateral hyperdense mass with a hypodense cystic area, which follows the globe contour and erodes the adjacent superior and lateral orbital walls (arrows).

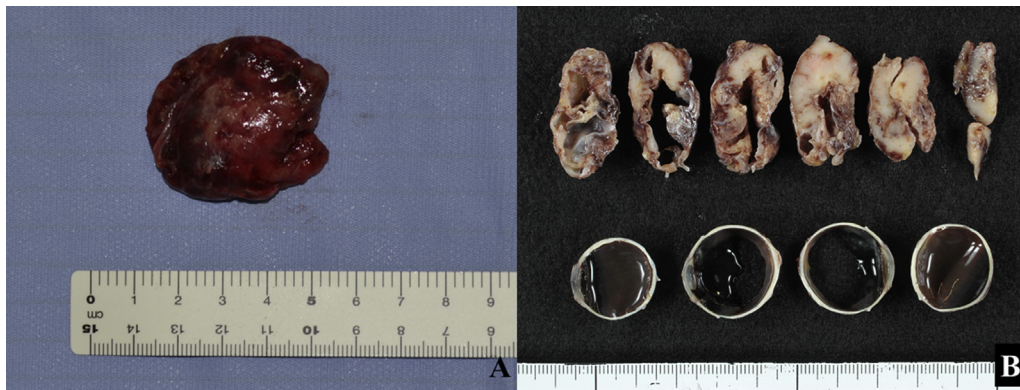


Fig. 3. Gross appearance of the excised mass and globe. (A) Pinkish mass measuring 35 × 40 × 45mm. (B) Pathological specimen shows multiple cysts within the mass.

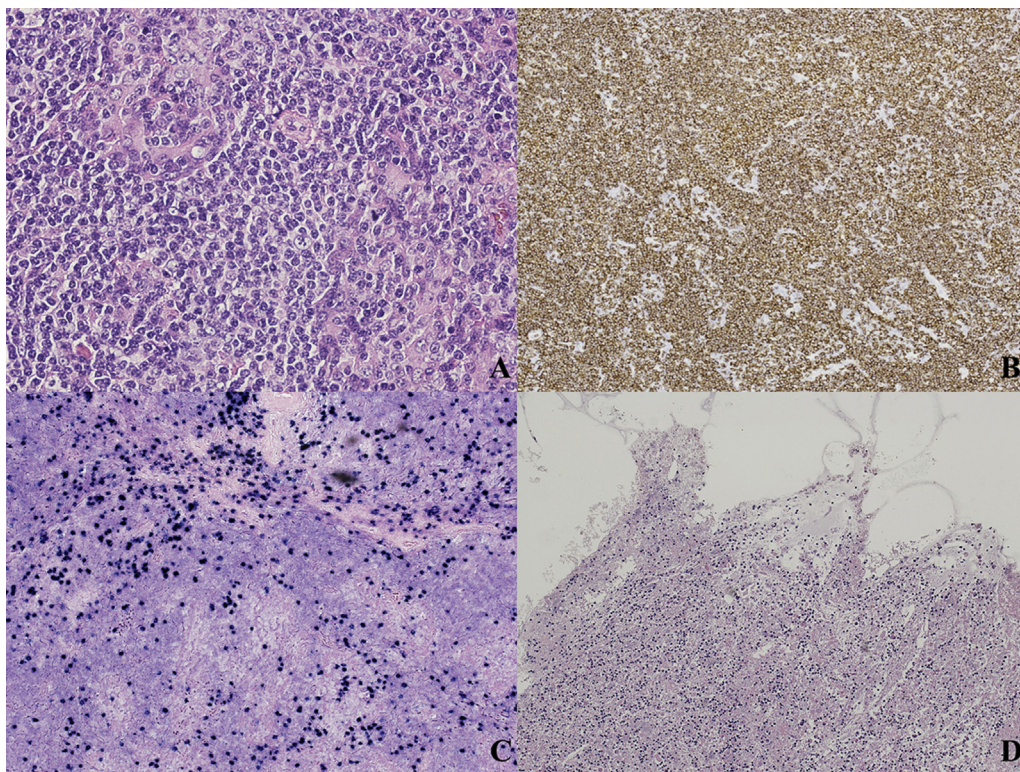


Fig. 4. Pathologic findings. (A) Aggregated lymphoma cells and plasma cells infiltrating the lacrimal gland (hematoxylin and eosin × 400) (B) Lymphocytes with positive immunoreactivity to monoclonal antibody anti-CD20 (×100). (C) Kappa light chain expression (×400). (D) Fluid in cysts composed of lymphocytes, histiocytes, and degenerated substances (hematoxylin and eosin × 100).

It is rare for a MALT lymphoma to present with vision loss. In this case, vision loss is mostly attributed to tumor compression of the optic nerve. Furthermore, exposure keratopathy that developed to corneal ulcer and subsequent endophthalmitis also contributed to visual impairment. Infiltration of the tumor to the globe, which was reported previously and caused vision loss,⁸ was, however, not evident in this case.

As radiotherapy and/or chemotherapy are effective for MALT lymphoma, a simple incisional biopsy is usually performed. However in this case, the tumor was totally removed. If the lymphoma is limited to ocular adnexa, the mode of therapy, either surgical excision, radiation, chemotherapy, or combination, achieves satisfactory results and without prognostic significance.^{9,10} However, elderly patients, particularly with dementia, are not cooperative during irradiation and chemotherapy, and they cannot tolerate

toxic effects of chemotherapy on occasion. In this case, tumor excision possibly benefited the elderly patient with dementia, from avoiding additional therapy. Yet, long term follow-up is mandatory.

Evisceration is the preferred surgical treatment for endophthalmitis with vision loss to reduce the risk of spreading the infection along the ophthalmic vein and provide better cosmesis.^{11,12} However in this case, enucleation was performed. Eucleation has the advantage of examining the entire globe for possible tumor extension. As well as, removal of the intraocular tumor is ensured if present.

3. Conclusions

It is essential that clinicians need to be aware of MALT lymphoma in the differential diagnosis for cystic lesion of lacrimal

gland. Early diagnosis and intervention are needed as this entity is sight-threatening.

3.1. Patient consent

The Institutional Review Board of the Ethics Committee of Aichi Medical University Hospital approved this retrospective case report, which adhered to the tenets of the 1964 Declaration of Helsinki. Written informed consent was also obtained from the patient's son, due to severe dementia in the patient, for the publication of this case report and any accompanying images.

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Conflict of interest

The authors have no competing interests to declare.

Authorship

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

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References

- Demirchi H, Shields CL, Shields JA, et al. Orbital tumors in the older adult population. *Ophthalmology*. 2002;109(2):243–248.
- Lee JL, Kim MK, Lee KH, et al. Extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue-type of the orbit and ocular adnexa. *Ann Hematol*. 2005;84(1):13–18.
- Rasmussen P, Ralfkiaer E, Prause JU, et al. Malignant lymphoma of the lacrimal gland: a nation-based study. *Arch Ophthalmol*. 2011;129(10):1275–1280.
- Lutz SC, Anderson SF, Wu CY, Townsend JC. Non-Hodgkin's orbital lymphoma. *Optom Vis Sci*. 2001;78(9):639–645.
- Weber AL, Jakobiec FA, Sabates NR. Lymphoproliferative disease of the orbit. *Neuroimaging Clin N Am*. 1996;6(1):93–111.
- Subramanian N, Rambhatia S, Mahesh L, et al. Cystic schwannoma of the orbit – a case series. *Orbit*. 2005;24(2):125–129.
- Qin W, Chong R, Huang X, Liu M, Yin ZQ. Adenoid cystic carcinoma of the lacrimal gland: CT and MRI findings. *Eur J Ophthalmol*. 2012;22(3):316–319.
- Sarraf D, Jain A, Dubovy S, et al. Mucosa-associated lymphoid tissue lymphoma with intraocular involvement. *Retina*. 2005;25(1):94–98.
- Nola M, Lukenda A, Bollmann M, et al. Outcome and prognostic factors in ocular adnexal lymphoma. *Croat Med J*. 2004;45(3):328–332.
- Auw-Haedrich, Coupland S, Kapp A, et al. Long term outcome of ocular adnexal lymphoma subtyped according to the REAL classification. Revised European and American lymphoma. *Br J Ophthalmol*. 2001;85(1):63–69.
- Dean WH, Sandford-Smith J. Evisceration, enucleation, and exenteration. In: Dean WH, Sandford-Smith J, eds. *Eye Surgery in Hot Climates*. fourth ed. London: JP Medical; 2015:307.
- Wong TY, Sng C, Lim L. Enucleation, evisceration & other orbital surgeries. In: Wong TY, Sng C, Lim L, eds. *The Ophthalmology Examinations Review*. second ed. New Jersey: World Scientific Publishing; 2011:386.