Case Report

DOI: 10.7860/JCDR/2021/47763.15339

Primary Non Hodgkin's Lymphoma of the Lacrimal Sac-A Noteworthy Watery Eye

Pathology Section

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ABSTRACT

Lacrimal sac tumours are rare tumours in ophthalmology but may have serious complications when misdiagnosed. Lacrimal sac tumours can be either epithelial or non epithelial tumours. Epithelial tumours account for 60-94% of tumours. Pseudostratified ciliated columnar epithelium lines the lacrimal sac. Squamous cell carcinoma, transitional cell carcinoma, mucoepidermoid carcinoma, oncocytic adenocarcinoma, adenoid cystic carcinoma and metastasis are the commonly occurring epithelial tumours. Inverted papilloma is a locally aggressive tumour. Non epithelial tumours (25%) are rarer than the epithelial tumours. Lymphoproliferative, melanocytic, and mesenchymal tumours are the non epithelial tumours, among which the lymphoproliferative tumours constitute 2-8%. Very rarely Non Hodgkin's lymphoma can occur as a primary in the lacrimal sac. Melanoma can occur as a primary from the melanocytes of the lining in the lacrimal sac or as a secondary from the conjuctival melanoma. Kaposi sarcoma and rhabdomyosarcoma are the mesenchymal tumours. Here is a case of Non Hodgkin's Lymphoma (NHL) occurring in the lacrimal sac in an 80-year-old male. The patient came with the chief complaint of swelling around the right eye since two months. On radiological examination, Computed Tomography (CT) revealed a homogenous solid mass in the right medial canthus, causing bony erosions of the orbit and extending into the nasolacrimal canal. An incisional biopsy of the mass was consistent with the diagnosis of Non Hodgkin's lymphoma which was further confirmed by immunohistochemistry. This unusual presentation and histopathological features are described.

Keywords: Immunohistochemistry, Lymphoproliferative lesion, Non epithelial tumour, Orbit

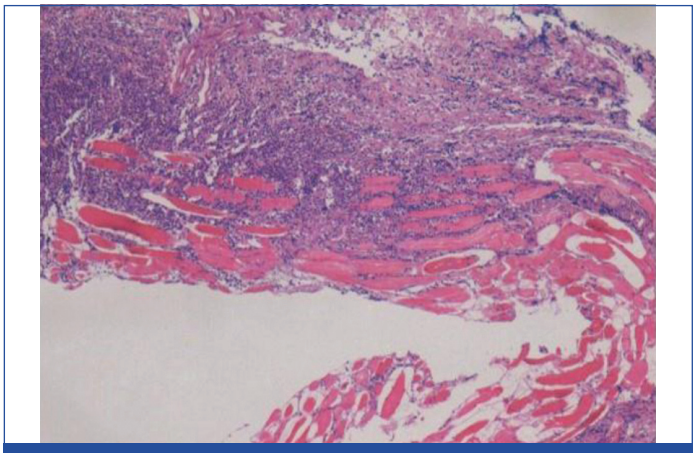
CASE REPORT

An 80-year-old male patient presented originally with the history of swelling around the right eye since two months. The swelling was insidious in onset, and gradually progressive in nature. There were no aggravating or relieving factors and it was not associated with pain. History of increased watering of the eyes was present. There was no history of dryness of eyes, fever with chills or rigors. Patient had a history of coronary artery disease and was on medications. Previous surgical history of fistulectomy and cholecystectomy was present. Patient was a smoker. There was no history of diabetes and hypertension.

On examination, a swelling was palpated in the subcutaneous area of the right medial canthus. It was a diffuse swelling, 2×2 cm in size and firm in consistency, extending upto the lateral wall of the nose. Skin over the swelling was normal. No warmth or tenderness was noted. A clinical diagnosis of lacrimal duct stenosis with dacryocystitis was made. The blood tests showed a deranged renal function test and patient was treated accordingly. CT of the orbit revealed an ill-defined solid mass in the right medial canthus causing bony erosion and extension into the nasolacrimal canal. The mass showed homogeneous and isointense patterns on T1WI and T2WI. Radiologically a diagnosis of lacrimal sac malignant lesion was given. A differential diagnosis of lacrimal sac tumour and orbital metastasis was given.

Under strict monitoring of the cardiac and renal parameters, incisional biopsy via external approach was performed under local anaesthesia. Gross examination showed three grey-white soft tissue bits altogether measuring 0.5×0.3×0.3 cms which was embedded in-toto.

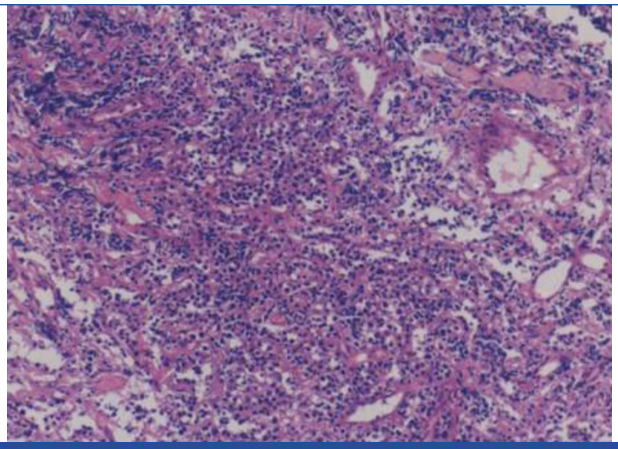
Microscopy showed a Small Round Blue Cell Tumour (SRBCT) invading into the underlying muscle bundles [Table/Fig-1]. The tumour was composed of diffuse monomorphic sheets of atypical cells [Table/Fig-2]. Individual cells had round to oval nuclei, dense chromatin with increased nuclear-cytoplasmic ratio and scant cytoplasm [Table/Fig-3]. Morphologically, the cells resembled a SRBCT and hence a provisional diagnosis of the same was made. The differential diagnosis were lymphoma, amelanotic melanoma and metastasis. Amelanotic melanoma was considered since the tumour was composed of sheets of atypical cells, with absence of melanin pigment, with few cells showing prominent nucleoli. Immunohistochemical (IHC) examination revealed that the tumour cells were diffusely positive for CD45 [Table/Fig-4] and CD20 [Table/Fig-5]. The background lymphocytes were positive for CD3 [Table/Fig-6]. The tumour cells were negative for HMB45 [Table/Fig-7], ruling out an amelanotic melanoma. With the light microscopy and immunohistochemical features, a diagnosis of Non-Hodgkin's lymphoma-B cell type was given. Chemotherapy with (R-CHOP) regimen consisiting of Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisone along with radiotherapy was administered. The patient responded well to the treatment and was followed-up for a period of two years.



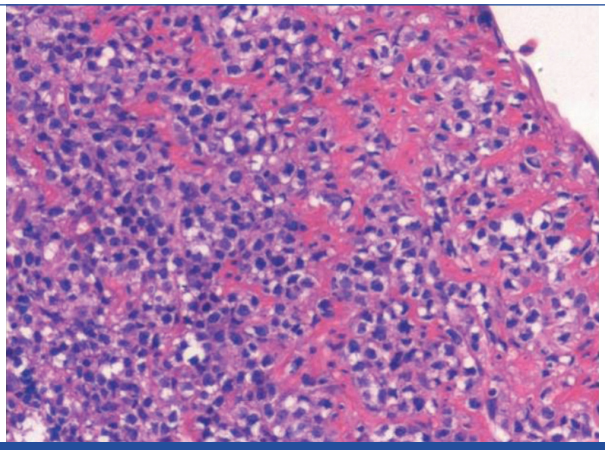
[Table/Fig-1]: Histopathology Examination (HPE) of the biopsied lacrimal sac lesion-photomicrograph reveals tumour invading into the underlying muscle bundles. H&E: X40.

DISCUSSION

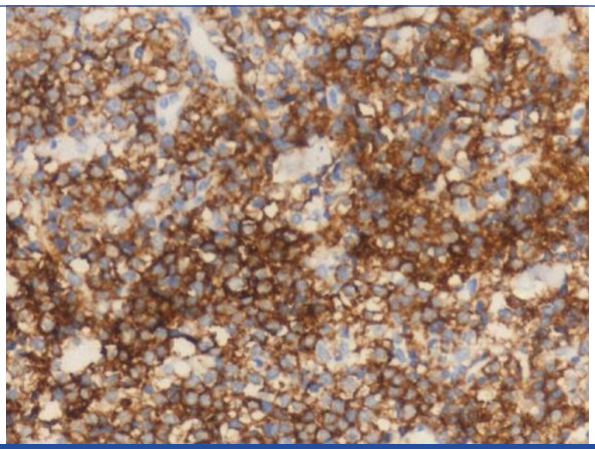
Primary malignancy of the eye is a rare entity. The presence of lymphoma as a primary tumour in the eye is even rare at the rate of 0.3% per 100,000 persons [1]. Extra-nodal lymphoma of the eye can occur in the orbit, eyelids, conjunctiva, lacrimal glands, lacrimal sac and contribute to about 2% of all the extra-nodal lymphomas [2]. In



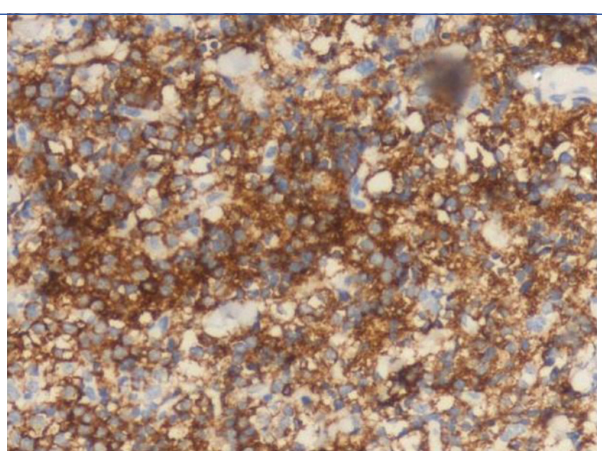
[Table/Fig-2]: The tumour was composed of diffuse monomorphic sheets of neoplastic lymphoid cells. H&E: X100.



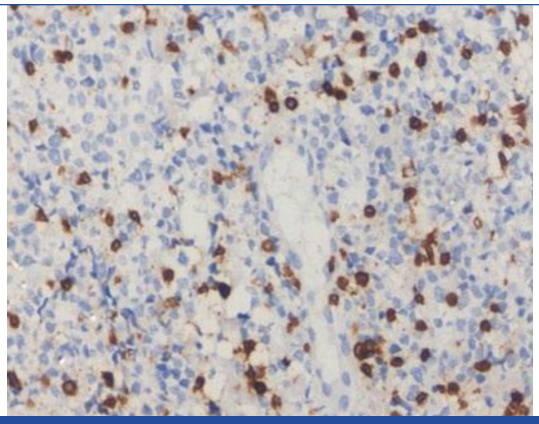
[Table/Fig-3]: Individual cells had round to oval nuclei, dense chromatin with increased nuclear-cytoplasmic ratio and scant cytoplasm. H&E: X400.



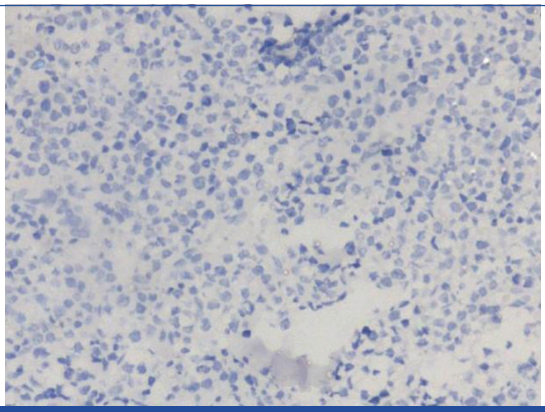
[Table/Fig-4]: IHC stains of the lacrimal sac lesion-lymphoid cells stained strongly positive for CD45, IHC: X400.



[Table/Fig-5]: The lymphoid cells were strongly positive for CD20, IHC: X400.



[Table/Fig-6]: IHC was also positive for the background lymphocytes with CD3 X400.



[Table/Fig-7]: The tumour cells were negative for HMB45 x400.

a study of 95 patients, by Lim SH et al., localisation of the lymphoma was found in the conjunctiva in 62 (65.2%) patients, orbital soft tissue in 15 (15.8%) patients, eyelid in 13 (13.75%) patients and lacrimal gland in 5 (5.2%) patients [3].

The signs and symptoms may be variable and include: swelling, irritation/pain, ptosis, epiphora, decreased visual accuracy, dry eye, lacrimal gland mass, proptosis/displacement, oedema, retrobulbar resistance and chemosis [4]. In this case, the patient had symptoms of painless swelling and epiphora. Nasolacrimal malignancy is predominantly epithelial accounting to 90% of the malignancies and nasolacrimal lymphomas are predominantly secondary to metastasis. The pathological diagnosis can be made on biopsy. Biopsy is done through a transfacial approach directly at the internal canthus. Lymphomas are neoplasms derived from clonal proliferations of lymphocytes. Among the primary lacrimal sac tumours, primary Diffuse Large B-cell Lymphoma (DLBCL) is the most frequently occurring lymphoma (43%). Other subtypes include Mucosa Associated Lymphoid Tissue Lymphoma (MALToma) (24%) unclassified B-cell lymphoma (21%), lymphoid hyperplasia (5%), small lymphocytic lymphoma (3%) and natural killer (NK)/T-cell lymphoma (3%) [5]. Large cell lymphoma, well differentiated NHL, and immunoblastic lymphoma was the diagnosis in few cases. In [Table/Fig-8] [6-15], it can be observed that out of the 10 reference cases nine were males and eight were females which reflects no specific gender predilection. The age group ranged between 12 years and 78 years whereas the present patient was 80-years-old. The majority had B phenotype. Histologically 14 cases of DLBCL, one case of B-cell lymphoma, one case of low-grade NHL were observed. Older age, male sex, no radiation therapy and DLBCL histology were associated with poor prognosis [16].

In a study of 95 patients by Lim SH et al., the median duration of symptoms before diagnosis was 10.5 months. Since the lacrimal sac tumours are rare, information is limited and clinically it mimics primary obstruction of the nasolacrimal system, they are considered life-threatening [17]. Delay in diagnosis can be avoided only with high suspicion. The treatment is similar to other Non-Hodgkin's lymphoma

Case	Author Name, Year [6-15]	Age (years)/ Gender	Chief complaint/Site of lesion	Duration	Histopathological diagnosis	B/T phenotype
1	Köksal Y et al., 2005 [6]	10/Male	Epiphora, lacrimal sac mass	Several months	B-cell Lymphoma	B
		9/Female	Swelling, lacrimal sac mass	1 month	DLBCL	B
2	Coloma-González I et al., 2014 [7]	72/Female	Epiphora, lacrimal sac mass	5 months	DLBCL	B
		51/Female	Epiphora, mass in lacrimal sac and NLD	5 years	Low grade NHL	B
3	Thomas A et al., 2010 [8]	54/Male	Epiphora, nasal obstruction, nasal polyps, lacrimal sac mass	3 months	DLBCL	B
4	Kajita F et al., 2010 [9]	77/Male	Epiphora, lacrimal sac mass	3 years	DLBCL	B
		70/Female	Epiphora, lacrimal sac mass	3 months	DLBCL	B
		40/Female	Epiphora, discharge, lacrimal sac mass	Several months	DLBCL	B
5	Tan S et al., 2011 [10]	14/Male	Epiphora, pain, mass in NLD	2 months	B-cell lymphoma	B
6	Alkatan HM et al., 2011 [11]	32/Male	Epiphora, lacrimal sac mass	Several months	DLBCL	B
		65/Female	Proptosis, loss of weight, mass in orbit	3 months	DLBCL	B
7	Gokce G et al., 2015 [12]	12/Male	Epiphora, mass in NLD	Few weeks	DLBCL	B
8	Tsao WS et al., 2015 [13]	47/Male	Epiphora, lacrimal sac mass	4 months	DLBCL	B
9	Marunaka H et al., 2015[14]	78/Female	Epiphora, lacrimal sac mass	5 months	DLBCL	B
10	Sabundayo MS et al., 2018 [15]	48/Male	Pain, lacrimal sac mass	2 months	DLBCL	B
		61/Male	Lacrimal sac mass	4 months	DLBCL	B
		62/Female	Lacrimal sac mass	5 months	DLBCL	B
11	Present study, 2021	80/Male	Epiphora, lacrimal sac mass	2 months	NHL-B cell type	B

[Table/Fig-8]: Literature search of similar case reports [6-15].
*NLD: Nasolacrimal duct; DLBCL: Diffuse large B-cell lymphoma

like chemotherapy with Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisone (R-CHOP regimen), radiotherapy and immunotherapy. When the tumour is deep-seated or when surgery is contraindicated, radiotherapy is the main treatment of choice [18]. Cataract (12.1%), dry eye (8.5%), conjunctivitis (6.8%), and keratitis (4.9%) are the complications related to orbital radiotherapy [19]. Thus, early diagnosis and management of the patient in a multidisciplinary centre with the expertise of Ophthalmologists, Pathologists and Oncologists is necessary for visual and vital prognosis.

CONCLUSION(S)

In a case of epiphora, high index of suspicion of chronic dacryocystitis secondary to lymphoma with subsequent biopsy is suggested to rule out these potentially fatal tumours in order to provide timely assessment and early therapeutic intervention.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Nov 19, 2020
- Manual Googling: Apr 21, 2021
- iThenticate Software: Jul 02, 2021 (12%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Nov 18, 2020**
Date of Peer Review: **Jan 09, 2021**
Date of Acceptance: **Apr 23, 2021**
Date of Publishing: **Sep 01, 2021**