Case Report

Ophthalmic Horizon
Volume : 16, Issue : I, March 2021; 59-63

Diffuse Large Cell Lymphoma of Lacrimal Sac May Mimic as Acute Dacryocystitis – A Case Report

Dr. Soma Rani Roy[1], Dr. Fahmida Hoque[2], Dr. Hardeep S Mudhar[3], Dr. Murtuza Nuruddin[4]

[1] Resident Surgeon, Chittagong Eye Infirmary & Training Complex (CEITC), Chattogram, Bangladesh
[2] Assistant Surgeon, Chittagong Eye Infirmary & Training Complex (CEITC), Chattogram, Bangladesh
[3] Consultant Eye Pathologist, National Specialist Ophthalmic Pathology Service (NSOPS), Royal Hallamshire Hospital, England, UK
[4] Director, Academics and Consultant Ophthalmologist, Chevron Eye Hospital and Research Centre, Chattogram, Bangladesh

ABSTRACT

Purpose: To report a case of diffuse large cell lymphoma of lacrimal sac in a young male patient.

Case report: A 36 years old male patient presented with recurrent episodes of painful swelling in right medial canthal region at Oculoplasty department of Chittagong Eye Infirmary and treated with systemic antibiotic considering the case as acute dacryocystitis. On examination his visual acuity was 20/20 in both eyes and anterior and posterior segment findings were unremarkable. The only ocular finding was firm swelling in right lacrimal sac region. Computed tomography showed soft tissue mass in lacrimal sac region. Due to diagnostic dilemma, surgery was deferred. Incisional biopsy confirmed the diffuse large cell lymphoma of non Hodgkin's type of right lacrimal sac. Thorough systemic evaluation was done and no other site was involved with lymphoma. Patient was treated with 6 cycles of Chemotherapy combination drug of Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisolone (CHOP) therapy and the lesion was resolved completely. Subsequently patient underwent DCR surgery with intubation. After removal of tube no epiphora or recurrence of lesion was noted in three year of follow up.

Conclusion: Primary lacrimal sac lymphoma is a rare entity. High suspicion and prompt decision making in atypical cases can save life of aggressive diffuse large cell lymphoma patient.

Keywords : Diffuse large cell lymphoma, acute dacryocyctitis, CHOP therapy.

Introduction

Though infection is the major ailment of lacrimal drainage system, neoplasm involving the same cannot be ignored. Tumors of the lacrimal sac, however relatively rare, it may be primary, secondary or metastatic. The majority of neoplasms are epithelial in origin which accounts for 73% and most of them (75%) are malignant[1]. Squamous cell carcinoma is the commonest one[1]. Lymphomas of the lacrimal sac are very rare[2,3,4]. The vast majority of nasolacrimal lymphoma is either secondary or metastatic[5]. Histopathologically, non-Hodgkin's B- cell lymphomas are commonest type of lymphoma in lacrimal drainage system[1]. Sjo et al have showed Diffuse large B-cell lymphoma (DLBCL) and extra nodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) are the most common histological types of lacrimal sac lymphoma in the UK[6]. But in Japan most reported cases of primary lymphoma in lacrimal sac are Diffuse large B-cell lymphoma[7]. In Bangladesh, we do not have any published data on lacrimal sac neoplasm.

Here we are reporting a case of diffuse large B - cell lymphoma (DLBCL) of lacrimal sac in a 36 year old male who presented as recurrent acute dacryocyctitis.

Manuscript Received : 03.02.2021
Revision Accepted : 28.02.2021

Correspondence to:
Dr. Soma Rani Roy; DCO, FCPS
Ocular Oncologist and Oculoplastic Surgeon
Resident Surgeon
Chittagong Eye Infirmary & Training Complex
E-mail: dr.somaroy2020@gmail.com

Ophthalmic Horizon
Volume : 16, Issue : I, March 2021; 59-63

Case Report

A 36 years old male patient presented with recurrent episodes of painful swelling in right medial canthal region and treated with systemic antibiotic considering the case as acute dacryocystitis. On examination his visual acuity was 20/20 in both eyes and anterior and posterior segment findings were unremarkable. The only ocular finding was firm swelling in right lacrimal sac region. Computed tomography of orbit showed soft tissue mass in lacrimal sac region without any bony erosion. Though patient was planned for dacryocystorhinostomy (DCR) surgery elsewhere, due to diagnostic dilemma lacrimal surgery was deferred and incisional biopsy from lesion was taken. Subsequent histopathology and diagnosis was confirmed as diffuse large cell lymphoma of non Hodgkin's type of right lacrimal sac. Thorough systemic evaluation was done and no other site was involved with lymphoma. Patient was treated with 6 cycles of CHOP therapy (Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisolone) and the lesion was resolved completely. At this stage patient underwent DCR surgery with intubation. Tube was removed after 3 months. No epiphora or recurrence of lesion was noted on further three year follow up.

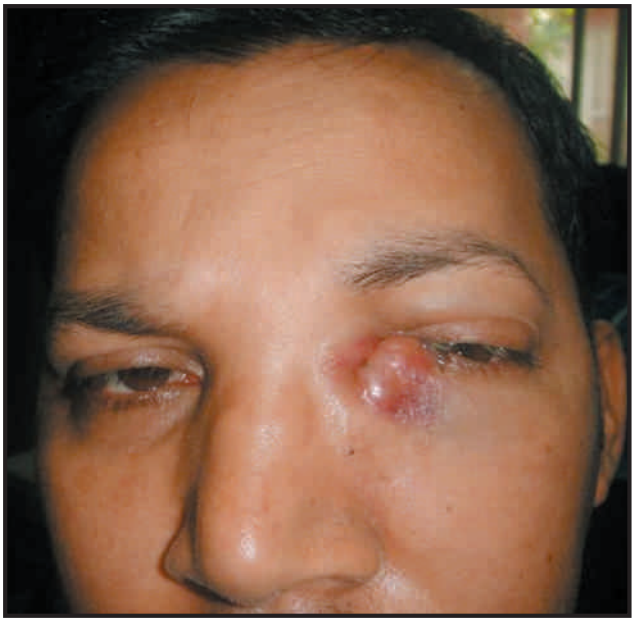

Figure-01: Mimicking as acute dacryocystitis at presentation.

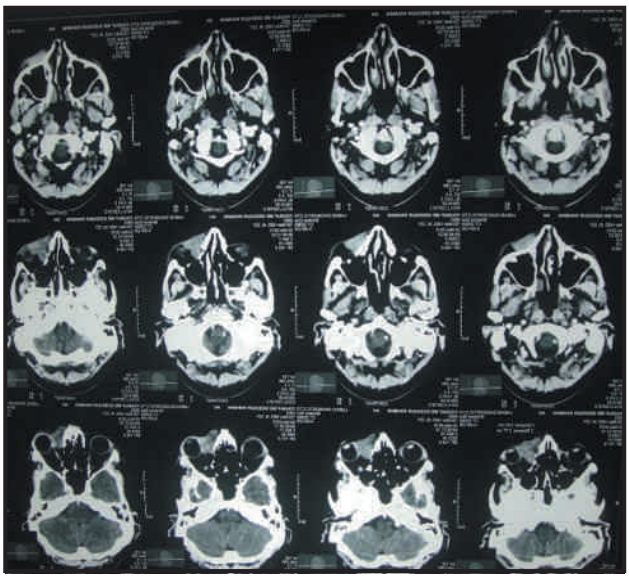

Figure-02: CT scan showing diffuse mass in lacrimal region

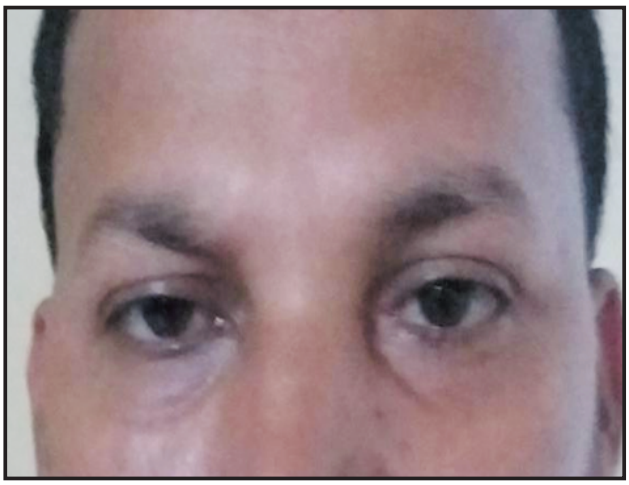

Figure-03: After receiving CHOP therapy and DCR operation

Discussion

Acute dacryocystitis is the acute inflammation or infection of the lacrimal sac characterized by redness, tenderness and swelling over the sac area along with epiphora. Our patient had recurrent episodes of similar features. He was treated with antibiotic and anti-inflammatory drugs on each episode and responded well with relief of pain and tenderness though the swelling persisted. The presentation of our patient was mimicking as acute dacryocystitis, which might be due to associated inflammation or concomitant infection

of the lacrimal sac as the passage was blocked. Presence of a relatively firm swelling was suspicious of neoplasm within the lacrimal sac. Lacrimal sac neoplasms are rare but life threatening. Tumours that arise from lacrimal drainage system may be epithelial or non-epithelial. It may be benign or malignant and may be primary, secondary or metastatic. Among these neoplasm 73% are epithelial origin of which 75% are malignant[1]. The different epithelial tumours that arise from lacrimal sac are papillomas, squamous cell carcinomas, transitional cell carcinomas, adenocarcinomas and mucoepidermoid carcinomas[8,9]. Squamous cell carcinoma is the commonest. Non-epithelial tumors can be further divided into lymph-oproliferative, melanocytic, and mesenchymal tumors. Lymphoproliferative tumors account for approximately 2-8% of lacrimal sac tumors. These tumors may be primary tumors of the lacrimal sac, but more commonly arise secondary to systemic spread in a patient with leukemia or lymphoma[5]. Primary lymphoma of lacrimal sac is extremely rare[2, 3, 4].

Systemically, DLBCL represents the most common lymphoma and it is responsible for about 30% of all lymphoid malignancy[10]. It is also one of most high grade lymphoma. DLBCL has clinical, histopathological, genetic and immunohisto-chemical heterogenicity[10,11,12]. That's why DLBCL is unlikely to represent a single disease process and has a variable prognosis[10]. Lymphomas related to eye are either intraocular or extraocular (orbital and adnexal). Intraocular lymphoma may be primary or secondary and are rare. On the other hand, orbital lymphoma comprises the commonest form of malignancy in the orbit . It represents 55% cases in adult[13] and 10% cases in older patients[14,15]. Non Hodgkin lymphoma (NHL) is the most common type of ocular lymphoma. Extra nodal marginal zone lymphoma of MALT is the most common lymphoma of ocular adnexa and accounting for 50-70% of cases according to different case series[16,17]. Orbital and periorbital DLBCL is an uncommon entity, representing 7–21% of primary ocular adnexal lymphoma in several case series[11,18,19], but 33% of secondary orbital lymphoma[20].

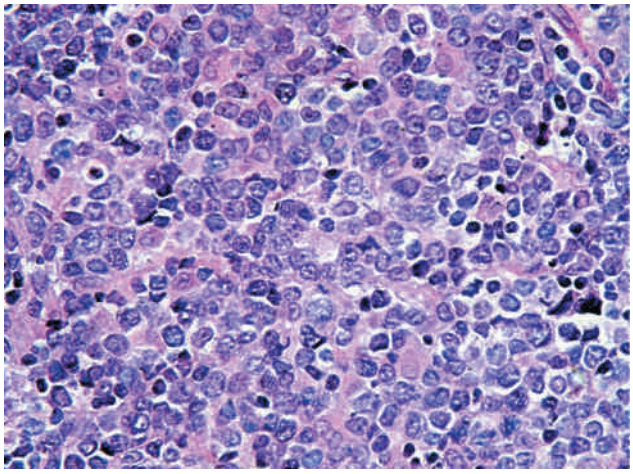

Figure-04: H&E stain: Diffuse infiltrate of atypical centroblast-type lymphoid cells

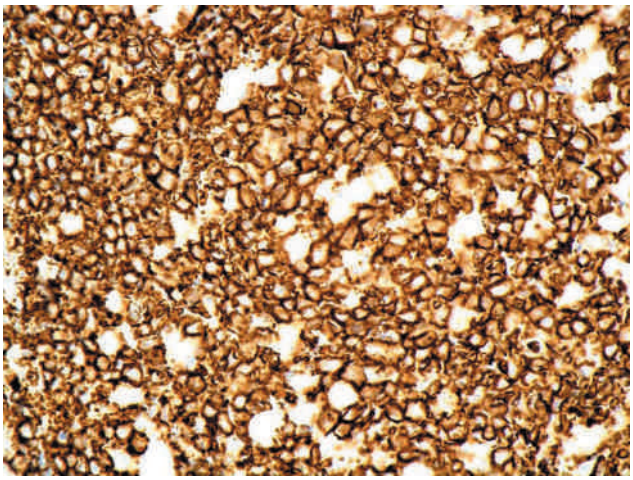

Figure-05: The atypical centroblast-type lymphoid cells are positive for B-cell marker CD20. Brown is positive membranous staining

Histopathologically, primary lymphoma of the lacrimal drainage system is usually a non-Hodgkin's B-cell lesion. Different study shows different histological features of primary lacrimal sac lymphoma. Sjo et al, done a case series of fifteen patients and five patients had diffuse large cell lymphoma, five had extra nodal marginal zone B cell lymphoma of mucosa associated lymphoid tissue (MALT lymphoma), three had "transitional MALT lymphoma" and two had unclassified B cell lymphomas[6]. Satoshi et al reported, in Japan the most common primary lacrimal sac lymphomas are Diffuse large B-cell lymphoma[7]. In our case the patient also had diffuse large cell lymphoma. The age of patient in

our case was 36 years. A large case seriese of 775 cases of lacrimal sac tumours from 1930's to till date by Krishna et al showed that benign tumours tend to present in younger patients and malignant tumours present more commonly in fifth decade[21]. Sjo et al, showed the median age at the time of diagnosis was 71 years (range 45–95 years)[6]. Therefore our patient had this lesion at an earlier age than expected. The common signs and symptoms associated with lacrimal sac neoplasms include epiphora, recurrent dacryocystitis, and lacrimal sac mass[22]. As epiphora is the commonest symptom,in most of cases the diagnosis is also difficult[23,24]. Another study showed the most frequent presenting symptoms were epiphora (85%), swelling in the region of the lacrimal sac (79%), and dacryocystitis (21%)[6]. In our case report patient presented with recurrent episides of acute dacryocystitis. He underwent incisional biopsy and histopathological report revealed high grade aggressive B cell lymphoma of non- Hodgkin's type which was positive for CD20. According to WHO classification it was diffuse large cell lymphoma.

According to literature, orbital and adnexal lymphoma is associated with systemic lymphoma in 30-35% of cases. So, all patients with ocular lymphoma should have a complete workup to rule out systemic lymphoma[25]. After appropriate staging of disease, treatment for the condition continues to involve chemotherapy, typically an anthracycline-based regimen such as CHOP (Cyclophosphamide, Adriamycin, Vincristine, and Prednisolone) and/or radiotherapy, although newer immunotherapies such as anti-CD20 antibodies may revolutionize management of the condition and improve the overall prognosis[10,26,27]. Thorough systemic evaluation was done in our case by oncologist and no other site was involved with lymphoma. Patient was treated with 6 cycles of CHOP therapy (Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisolone) and the lesion was resolved completely. Diffuse large cell lymphoma is typically high grade lymphoma but it is potentially curable[6]. After completion of chemotherapy and cure of lesion our patient underwent DCR surgery with intubation. Tube was removed after 3 months. No epiphora or recurrence of lesion was noted on further three year follow up. Survival rate of patient depends on lesion either it is localized or not. One cohort study representing the largest series of 37 patients with orbital DLBCL in the literature found that localized disease provided a high 5-year survival rate of 90.9%. An advanced disease does not provide such an optimistic survival rate.

References

1. Myron Yanoff MD, Joseph W. Sassani MD MHA, in Ocular Pathology (Eighth Edition).
2. Gao HW, Lee HS, Lin YS, Sheu LF: Primary lymphoma of nasolacrimal drainage system: a case report and literature review. Am J Otolaryngol 2005; 26: 356–359.
3. Litschel R, Siano M,Tasman AJ, Cogliatti S: Nasolacrimal duct obstruction caused by lymphoproliferative infiltration in the course of chronic lymphocytic leukemia. Allergy Rhinol Providence 2015; 6: 191– 194.
4. Yip CC, Bartley GB, Habermann TM, Garrity JA: Involvement of the lacrimal drainage system by leukemia or lymphoma. Ophthal Plast Reconstr Surg 2002; 18: 242–246.
5. M. A. Stefanyszyn, A. A. Hidayat, J. J. Pe'er, and J. C. Flanagan,"Lacrimal sac tumors," Ophthalmic Plastic and Reconstructive Surgery, vol. 10, no. 3, pp. 169–184, 1994.
6. Sjö LD, Ralfkiaer E, Juhl BR, Prause JU, Kivelä T, Auw-Haedrich C, et al. European Organization for Research and Treatment of Cancer Primary lymphoma of the lacrimal sac: an EORTC ophthalmic oncology task force study. Br J Ophthalmol. 2006 Aug;90((8)):1004–9
7. Satoshi Kakutani, Yasuhiro Takahashi,* Ma. Regina Paula Valencia, and Hirohiko Kakizaki , Case Rep Ophthalmol. 2018 Sep-Dec; 9(3): 516–519.
8. Bi YW, Chen RJ, Li XP. Clinical and pathological analysis of primary lacrimal sac tumors. Zhonghua Yan Ke Za Zhi.2007; 43: 499-504.
9. Flanagan JC, Stokes DP. Lacrimal sac tumors. Ophthalmology.1978; 85: 1282-1287.
10. Hunt KE, Reichard KK. Diffuse large B-cell lymphoma. Arch Pathol Lab Med 2007; 132: 118 -124.
11. Ahmed S, Shahid RK, Sison CP, Fuchs A, Mehrotra B. Orbital lymphomas: a clinicopathologic study of a rare disease. Am J Med Sci 2006; 331(2): 79–83.

12. Bardenstein DS. Ocular adnexal lymphoma: classification, clinical disease and molecular biology. Ophthalmol Clin N Am 2005; 18: 187–197.
13. Valvassori GE, Sabnis SS, Mafee RF, Brown MS, Putterman A. Imaging of orbital lymphoproliferative disorders. Radiol Clin North Am. 1999 Jan. 37(1):135-50, x-xi.
14. Shields JA, Shields CL, Scartozzi R. Survey of 1264 patients with orbital tumors and simulating lesions: The 2002 Montgomery Lecture, part 1. Ophthalmology. 2004 May. 111(5):997-1008.
15. Bonavolontà G, Strianese D, Grassi P, Comune C, Tranfa F, Uccello G, et al. An analysis of 2,480 space- occupying lesions of the orbit from 1976 to 2011. Ophthal Plast Reconstr Surg. 2013 Mar-Apr. 29(2):79 -86.
16. Coupland SE, Krause L, Delecluse HJ, Anagnostopoulos I, Foss HD, et al. (1998) Lymphoproliferative lesions of the ocular adnexa. Analysis of 112 cases. Ophthalmology 105: 1430-1441.
17. Coupland SE, Hellmich M, Auw-Haedrich C, Lee WR, Stein H (2004) Prognostic value of cell-cycle markers in ocular adnexal lymphoma: an assessment of 230 cases. Graefes Arch Clin Exp Ophthalmol 242: 130-145.
18. Sullivan TJ, Whitehead K, Williamson R, Grimes D, Schlect D, Brown I et al. Lymphoproliferative disease of the ocular adnexa: a clinical and pathologic study with statistical anaylsis of 69 patients. Ophthal Plast Reconstr Surg 2005; 21(3): 177–188.
19. Henderson JW, Campbell RJ, Farrow GM, Garrity JA. Orbital Tumors, 3rd ed. Raven Press: New York, 1994.
20. Esmaeli B, Ahmadi MA, Manning J, McLaughlin PW, Ginsberg L. Clinical presentation and treatment of secondary orbital lymphoma. Ophthal Plast Reconstr Surg 2002; 18(4): 247–253.
21. Krishna Y, Coupland SE. Lacrimal Sac Tumors--A Review. Asia Pac J Ophthalmol (Phila). 2017; 6(2):173–178. doi:10.22608/APO.201713
22. Stefanyszyn MA, Hidayat AA, Pe'er JJ, Flanagan JC. Lacrimal sac tumors. Ophthal Plast Reconstr Surg. 1994; 10: 169–84.
23. Parmar DN, Rose GE. Management of lacrimal sac tumours. Eye (Lond) 2003; 17: 599–606.
24. Ryan SJ, Font RL. Primary epithelial neoplasms of the lacrimal sac. Am J Ophthalmol. 1973; 76:73–88.
25. Özkan MC, Palamar M, Tombuloğlu M, Hekimgil M, Özsan N, Saydam G, et al. Ocular Adnexal Lymphomas: Single-Center Experience. Clin Lymphoma Myeloma Leuk. 2015 Jun. 15 Suppl:S 158-60.
26. Sullivan TJ, Grimes D, Bunce I. Monoclonal antibody treatment of orbital lymphoma. Ophthal Plast Reconstr Surg 2004; 20(2): 103–106.
27. Cvetkovic RS, Perry CM. Rituximab. A review of its use in non-Hodgkin's lymphoma and chronic lymphocytic leukaemia. Drugs 2006; 66(6): 791–820.
28. S. N. Madge, A. Mccormick, I. Patel et al., "Ocular adnexal diffuse large B-cell lymphoma: local disease correlates with better outcomes," Eye, vol. 24, no. 6, pp. 954–961, 2010.