Clinical Oncology: Case Reports

Case Report

A SCITECHNOL JOURNAL

Lacrimal Sac Diffuse Large B-Cell Lymphoma Presenting as Sudden Onset Binocular Diplopia: A Case Report

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Abstract

Introduction: Diffuse Large B Cell Lymphoma (DLBCL) is a prevalent subtype of Non-Hodgkin Lymphoma (NHL) affecting predominantly elderly individuals. Lacrimal gland DLBCL is rare, posing diagnostic challenges due to its atypical presentation.

Case Presentation: A 68-year-old man with a history of hypertension, hyperlipidemia, and a small pituitary gland tumor presented with sudden-onset binocular diplopia and right eye blurry vision. MRI of the brain revealed enhancing soft tissue in right superolateral orbit inseperable from the lacrimal gland and extending medially to the right superior rectus muscle and soft tissue. Further scanning showed widespread metastasis to bilateral retroperitoneal lymph nodes, adrenal gland, spine, and lymph nodes in the neck. Biopsy of the lacrimal gland confirmed DLBCL. The patient was started on chemotherapy consisting of methotrexate and rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone.

Conclusion: Primary lacrimal gland DLBCL is a rare and diagnostically challenging entity. The delayed diagnosis often stems from the resemblance of its clinical manifestations to more benign conditions such as dacryocystitis, dacryostenosis, or mucocele. Timely recognition and accurate diagnosis are essential for initiating appropriate treatment and improving patient outcomes.

Keywords: Diffuse large B cell lymphoma; Ocular dnexal lymphomas; Diplopia

Introduction

In the United States, the annual incidence of Non-Hodgkin Lymphoma (NHL) is reported to be 7 cases per 100,000 individuals, with Diffuse Large B Cell Lymphoma (DLBCL) comprising approximately 30%-40% of all NHL cases across diverse geographic regions. DLBCL exhibits a predilection for the elderly population, particularly those in the 6th to 7th decade of life, with a notable male predominance. The gastrointestinal tract is identified as the most prevalent primary site for DLBCL [1].

Clinical manifestations of DLBCL commonly include B symptoms, such as weight loss, fever, and night sweats, along with organ-specific symptoms corresponding to the involvement of specific anatomical regions, such as abdominal pain in gastrointestinal tract cases or headaches in central nervous system presentations. Due to the nonspecific nature of these symptoms, DLBCL is often diagnosed at advanced stages, with approximately 50% of patients presenting at stage III or IV. Subsequent classification as concordant or discordant is determined through bone marrow biopsy, with concordant involvement predicting a less favorable overall survival [2].

Ocular adnexal lymphomas, encompassing lymphomas affecting the orbits, eyelids, conjunctiva, lacrimal gland, and lacrimal sac, constitute 2% of all extranodal lymphomas and stand as the most prevalent malignant tumors within the orbit. Notably, lacrimal gland DLBCL, despite DLBCL's general male predominance, exhibits a female predominance. This rarity contributes to frequent misdiagnoses as benign or inflammatory pathologies, given its infrequent occurrence and atypical presentation [3].

Herein, we present a case involving a 68-year-old man with lacrimal gland diffuse large B-cell lymphoma, who presented with sudden-onset binocular diplopia. The CARE checklist has been completed by the authors for this case report attached as supplementary material.

Case Presentation

68-year-old man with past medical history of hypertension, hyperlipidemia, small pituitary gland tumor (0.8 cm discovered one year before admission), recent sinus polypectomy, presents to the Emergency Department due to Ophthalmology referral for sudden same day onset of binocular diplopia on distance vision associated with 10 days of right eye blurry vision and cranial nerve III palsy.

In the ED, patient arrived hypertensive, with otherwise stable vitals. General appearance revealed a well-developed, not ill-appearing elderly male, with unremarkable physical exam aside from cranial nerve III palsy evidenced due to supraduction limitation of the right eye and V deviation. Patient was taken to CT head, CTA head and neck and MRI orbits and brain to rule out stroke vs aneurysm vs intracranial process. CTA head and neck showed no significant stenosis, aneurysms, vascular malformation, or large vessel occlusion. CT head revealed no evidence of acute intracranial hemorrhage, midline shift or mass effect. MRI of the brain showed enhancing soft tissue in right superolateral orbit inseparable from the lacrimal gland and extending medially to the right superior rectus muscle and soft tissue abutting and possibly eroding posterior dorsal seller with possible invasion of posterior left cavernous sinus concerning for lymphoma vs. idiopathic orbital pseudo tumor (Figure 1).

Due to past history significant for small pituitary gland tumor that was diagnosed one year before presentation and did not require surgery at the time, MRI of the pituitary gland was ordered showing soft tissue mass involving sella, upper cloves and posterior dorsum sellae with extension into posterior left cavernous sinus concerning for pituitary adenoma, meningioma or metastatic disease.

Endocrinology was consulted for para sellar mass and stated unlikely pituitary origin given small size of adenoma last year and rapid, vast extension of mass and obtained pituitary function exams showed on (Table 1). Neurology was consulted where notable left cranial nerve 6 palsy and right cranial nerve 4 palsy was evidenced, and LP was done showing increased WBC in CSF (Table 2). Lacrimal gland biopsy was done by Ophthalmology which revealed diffuse large B cell lymphoma with nodular proliferation of large B cells of the right lacrimal gland.

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Received: March 14, 2024; Manuscript No: COCR-24-129453 **Editor Assigned:** March 17, 2024; PreQC Id: COCR-24-129453 (PQ) **Reviewed:** March 25, 2024; QC No: COCR-24-129453 (Q) **Revised:** April 05, 2024; Manuscript No: COCR-24-129453 (R) **Published:** April 10, 2024; DOI: 10.4173/cocr.7(3).339

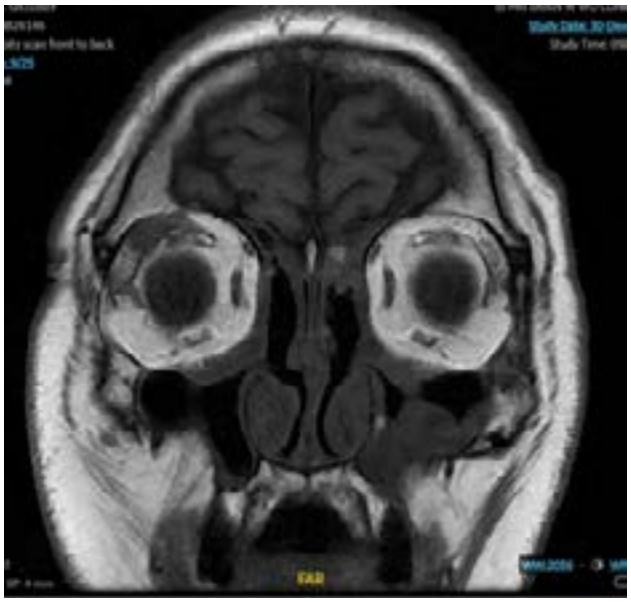

Figure 1. MRI orbits showing enhancing soft tissue in right superolateral orbit inseparable from the lacrimal gland and extending medially to the right superior rectus muscle

Laboratory	Patient results	Normal range
8 AM Cortisol	5.5 mcg/dl	6.7 mcg/dl -22.6 mcg/dl
ACTH	18.5 pg/dl	7.2 pg/ml -63.3 pg/ml
TSH	3.65 uIU/ml	0.4 uIU/ml -4.20 uIU/ml
Free T4	0.73 ng/dl	0.8 ng/dl -1.5 ng/dl
LH	2.44 mIU/ml	1.2 mIU/ml -8.6 mIU/ml
Testosterone	66 ng/dl	77 ng/dl -357 ng/dl
Prolactin	15.4 ng/ml	1.60 ng/ml -18.80 ng/ml
IGF-1	180 ng/ml	59 ng/ml -230 ng/ml
LDH	318 U/L	140 U/L -280 U/L

Table 1: Lab Exams.

	CSF
Nucleated cells	100 cells
Differential	70% lymphocyte predominance
RBC	6010 RBC
Glucose	75 mg/dl

Table 2: CSF study.

On further imaging to stage lymphoma, CTAP showed bulky bilateral retroperitoneal lymphadenopathy measuring 2.6 cm, 1.7 cm right adrenal lesion, and further metastases were found in the spine (T1, T11, L1) and lymphadenopathy in right internal jugular chain and right submandibular triangle (Figure 2). Patient was then started on methotrexate and rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone.

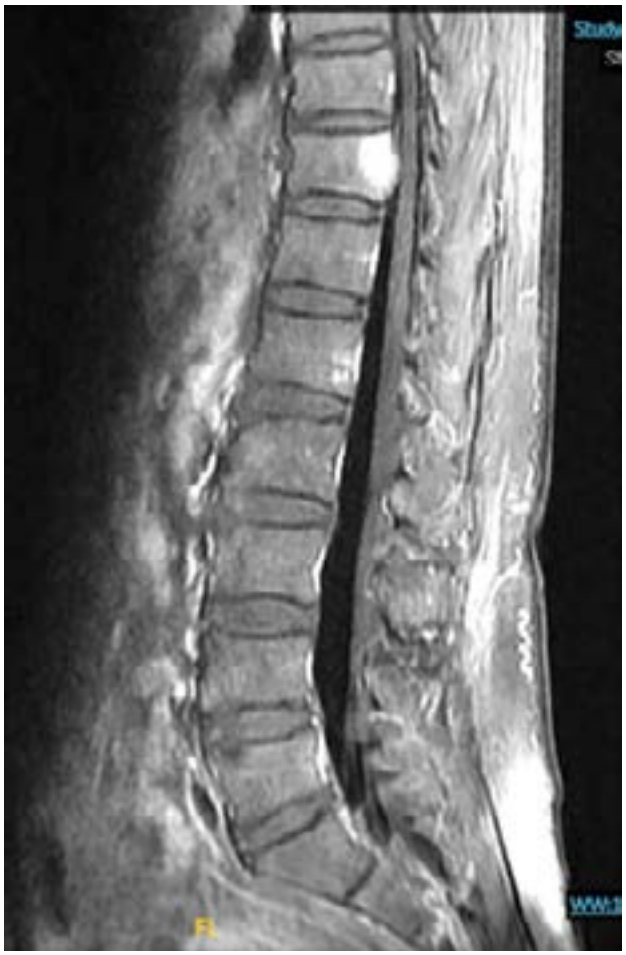

Figure 2. Graph of laboratory values throughout hospital admission

Discussion

Extranodal involvement is a frequently observed phenomenon in Diffuse Large B Cell Lymphoma (DLBCL), with predominant affliction sites typically encompassing the gastrointestinal tract and the head and neck region. However, lacrimal sac tumors present a distinctive rarity within the spectrum of DLBCL, exhibiting uncommon clinical presentations [4]. Notably, lacrimal sac lymphomas manifest predominantly in the elderly, with an average age of onset around 71 years. Furthermore, a discernible gender predilection is evident, with a higher incidence observed among women. This specific demographic pattern characterizes lacrimal sac lymphomas as an atypical subset within the broader spectrum of DLBCL.

The clinical presentation of lacrimal sac large B-cell lymphoma is often characterized by nonspecific symptoms, posing a considerable diagnostic challenge. Initial misdiagnoses commonly involve confusion with conditions such as dacryocystitis or dacryostenosis, while alternative misidentifications as mucocele have also been reported [5]. A prompt diagnosis is achieved in less than 15% of cases due to the elusive nature of the presenting symptoms. The diagnostic approach for lacrimal sac large B-cell lymphoma involves a comprehensive strategy, beginning with a biopsy incorporating flow cytometry, molecular studies, and immunophenotyping. Subsequent categorization and staging of the tumor are accomplished through the utilization of advanced imaging modalities such as Computed Tomography (CT) and Magnetic Resonance (MR) imaging, as demonstrated in the assessment of our presented patient.

Meunier's study conducted a comprehensive analysis, revealing that factors such as age exceeding 59 years, elevated Lactate Dehydrogenase (LDH) levels, stage IV disease, high-grade histological subgroup, and the presence of B-symptoms and bone marrow metastasis exerted a negative influence on Overall Survival (OS) within the general population [6]. In our reported case, the patient exhibited several adverse prognostic indicators, including an age surpassing 59 years, elevated LDH levels at 318, and a diagnosis of stage IV disease. Notably, our patient had the absence of B-symptoms at the time of presentation. Furthermore, it is pertinent to acknowledge that the majority of lacrimal sac lymphomas belong to the B-cell type, which typically responds favorably to radiotherapy and chemotherapy. Consequently, the prognosis for such cases is generally optimistic, emphasizing the importance of subtype-specific considerations in predicting outcomes and guiding therapeutic decisions.

In the management of primary lacrimal sac lymphoma, chemotherapy and/or radiotherapy constitute the primary therapeutic modalities. CHOP regimens, comprising cyclophosphamide, doxorubicin, vincristine, and prednisolone, with adjunct immunotherapy such as Rituximab targeting the CD20 antigen, are commonly used, particularly in cases of diffuse large B-cell lymphomas. However, in the presented case, the therapeutic approach was augmented with the inclusion of methotrexate alongside the R-CHOP regimen. This modification was prompted by the identification of a pituitary gland tumor, raising concerns about the potential development of secondary Central Nervous System (CNS) lymphoma [7]. This tailored therapeutic strategy underscores the necessity of individualized treatment plans based on specific clinical presentations and associated findings. It has been found that patients diagnosed with systemic diffuse large B-cell lymphoma have a worse prognosis than those diagnosed with ocular adnexal diffuse large B-cell lymphoma [1].

Conclusion

In conclusion, primary lacrimal sac large B-cell lymphomas represent a rare and diagnostically challenging entity characterized by atypical presentations. The delayed diagnosis often stems from the resemblance of its clinical manifestations to more benign conditions such as dacryocystitis, dacryostenosis, or mucocele. Given the rarity of this lymphoma and the potential for misdiagnosis, it is imperative for clinicians to maintain a high index of suspicion and include this malignancy in the differential diagnosis when evaluating patients with related ocular or periocular conditions. Timely recognition and accurate diagnosis are pivotal for instituting prompt therapeutic interventions and improving patient outcomes

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding

This study was not supported by any sponsor or funder.

Author Contributions

AJY: Drafting of case report, write up, literature research, case collection
BD: Drafting of case report, write up, literature research, case collection
MP: Drafting of case report, write up, literature research, case collection
ML: Drafting of case report, write up, literature research, case collection

References

1. Li S, Young K.H, & Medeiros L J. (2018). Diffuse large B-cell lymphoma. Pathology, 50: 74-87.
2. Yao Z, Deng L, Xu-Monette ZY, Manyam GC, Jain P et al. (2018). Concordant bone marrow involvement of diffuse large B-cell lymphoma represents a distinct clinical and biological entity in the era of immunotherapy. Leukemia, 32: 353-363.
3. Rasmussen P, Ralfkiaer E, Prause J.U, Sjö L.D, et al. (2011). Malignant lymphoma of the lacrimal gland: a nation-based study. Arch Ophthalmol, 129: 1275-1280.
4. Soman S, Thambi S M, & Jacob P M. (2022). Lacrimal sac diffuse large B-cell lymphoma: A case report. Indian J Ophthalmol, 70: 2618-2620.
5. Tsao W S, Huang T L, Hsu Y H, Chen N, et al (2016). Primary diffuse large B cell lymphoma of the lacrimal sac. Taiwan J Ophthalmol, 6: 42-44.
6. Meunier J, Rouic L, L. L Vincent-Salomon, A Dendale, R Asselain et al. (2004). Ophthalmologic and intraocular non-Hodgkin's lymphoma: A large single centre study of initial characteristics, natural history, and prognostic factors. Hematol Oncol, 22: 143-158.
7. Masaki Y, Miki M, Sun Y, Nakamura T, Iwao H et al. (2011). High-dose methotrexate with R-CHOP therapy for the treatment of patients with primary central nervous system lymphoma. Int J Hematol, 93, 720-726.

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