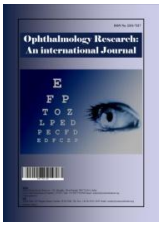

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Primary Orbital Lymphoma – A Rare Challenging Diagnostic Entity

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Authors' contributions

This work was carried out in collaboration among all authors. Author NM designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors ND and MS managed the analyses of the study. Author SS managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

The diagnosis of lymphoproliferative disorder of orbit is quite challenging as both Pseudolymphomas and Non-Hodgkin's lymphomas can occur in orbit. Primary orbital lymphoma is a rare entity comprising of 1-2% of Non-Hodgkin's lymphoma, majority of them are B-cell type. It is a slow growing tumor. It presents in the age group of 50–70 years, with a slight female preponderance. Proptosis is the most common presentation. We, herein, report a case of 48 years old male presenting with right non-axial proptosis, watering of eye, blurring of vision and redness for the last 9 months. After thorough clinical and radiological evaluation, a biopsy was taken for histopathological examination. A primary diagnosis of Lymphoproliferative Disorder was made. Following this, a panel of immunohistochemical markers was applied and a final diagnosis of Non-Hodgkin Lymphoma, B-cell type was given.

Keywords: Primary; non-hodgkin's lymphoma; B cell type; orbit.

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1. INTRODUCTION

Lymphoid proliferations of the ocular adnexa pose a diagnostic challenge [1]. Normal lymphoid tissue in the ocular region remains confined to the eyelid, conjunctiva, and lacrimal gland [2]. Lymphoid proliferations of the ocular adnexa constituted 10-24.7% of primary ocular adnexal tumors [3]. Lymphoid proliferations have variety of lesions ranging from benign lymphoid hyperplasia (LH) to malignant lymphomas [4]. Rarely, NHL may occur simultaneous with LH, hence, long-term follow-up is required [2].

Primary lymphoma of the orbit is an uncommon type of NHL, accounting for 1% of all NHL and 8-10% of extranodal NHL. [5] They are indolent, slow-growing B cell lymphomas, most common, being mucosa associated lymphoid tissue (MALT) type (57%) followed by follicular lymphomas (19%), diffuse large B-cell lymphomas (DLCL) and mantle cell lymphomas [5,6]. Orbital lymphoma have also been documented to be associated with Helicobacter pylori, Chlamydia psittaci, Hepatitis-C, human Herpes virus, human T-cell lymphotrophic virus type-1 (HTLV-1) and Epstein-Barr virus (EBV) [7]. There is a multimodality treatment approach for Orbital lymphoma depending on the stage and extent of disease. [8]

2. PRESENTATION OF CASE

A case of 48 years old male reported to eye OPD with complaints of right non-axial proptosis since 9 months associated with intermittent watering, blurring of vision, diplopia, redness and decreased visual acuity. There was no past medical history. On ophthalmological examination, visual acuity of right eye was 6/24 along with circumcorneal congestion, relative afferent papillary defect and restriction of ocular movement in upward gaze of right eye. Left eye was apparently normal. Optic nerve and fundus examination were within normal limits. Following this, MRI scan was done which showed homogenously enhancing lesion in right orbit. (Fig. 1A) After thorough clinical and radiological evaluation, a biopsy was taken for histopathological examination. Microscopic examination revealed nodular aggregates of monomorphic lymphoid cells. (Fig. 1B,C) Following this, a panel of immunohistochemical markers including CD3, CD20, CD5, CD23, CD10, BCL2, BCL6, Cyclin D1, TdT, Kappa, Lambda and Ki-67 were applied which showed positivity of CD20, BCL2 and Kappa with Ki-67 index of 4-5%. (Fig.1D,E,F) A final diagnosis of Non-Hodgkin Lymphoma, B-cell type was given.

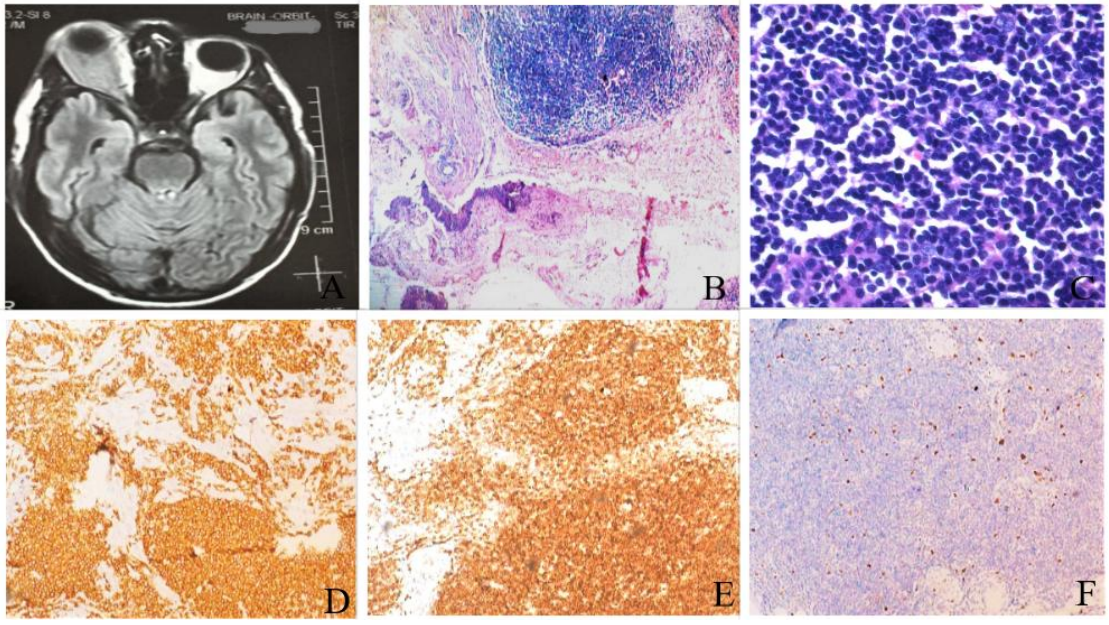

Fig. 1. A: MRI scan showed homogenous enhanced lesion in right orbit; B: Nodular aggregates of lymphoid cells (H&E, 100X); C: Monomorphous population of lymphoid cells (H&E, 400X); D: CD20 positivity (IHC, 400X); E: BCL2 positivity (IHC, 400X); F: Ki-67 index (IHC, 400X)

3. DISCUSSION

Primary orbital lymphoma is a rare entity, constituting about 1% of non‐hodgkin lymphomas [8]. The diagnosis of primary orbital lymphoma is considered when there is no history of prior lymphoma and systemic lymphoma. [6] Investigations viz., complete blood count, ESR, liver function tests, HIV serology, LDH levels and serum electrophoresis should be done to rule out systemic manifestations, while rendering diagnosis and also during follow up visits. [7]

Most of them are B-cell lymphomas, marginal zone lymphomas being the most common subtype. [6] Histologically, these extranodal lymphoid proliferations can be classified into chronic inflammatory lesions, benign pseudolymphomas and non-Hodgkin lymphomas (NHL). [1] A detailed study of clinical, histopatholgical and immunohistochemical techniques are required to identify and classify various orbital lymphoid infiltrates. [1] Majority of the orbital lymphomas are of low-grade variety (84%) and only 16% are high-grade. [5] It presents mostly in the age group of 50–70 years. The most common presentation is progressive proptosis, followed by decreased visual acuity, diplopia, and restricted ocular movement. The majority of lesions are unilateral, while, bilateral involvement is seen in 5% of the cases. [6]

Differential diagnosis includes orbital adnexal MALT lymphoma, idiopathic orbital inflammatory pseudotumour, lacrimal gland tumours, orbital melanoma and orbital metastases. Biopsy is mandatory to differentiate lymphoma from other lymphoproliferative disorders of ocular adenexa. [9] Orbital fat, lacrimal gland and extraocular muscles are frequently involved in orbital pseudotumor. [10] Superior lateral quadrant and superior rectus extraocular muscle is frequently involved in lymphoma while medial rectus, superior rectus, lateral rectus, followed by inferior rectus have predilection for pseudotumors. [6,11] Pseudotumors showed characterstic features of irregular borders of extraocular muscle and fat infiltration. [11]

There is a multimodality treatment approach for orbital lymphoma including radiotherapy, chemotherapy and surgery while surgery alone has been reserved for tissue biopsy. [8] The treatment depends on the histological subtype, extend of disease, visual function, and disease‐related prognostic factors. [8] Prognosis of orbital lymphoma is good with 5-year overall survival rate ranging between 50% and 94%. [12]

4. CONCLUSION

Primary lymphoma of the orbit is a rare aggressive entity with multimodality treatment approach. It is essential to differentiate it from other ocular adnexal lymphoproliferative disorder. Careful clinical, radiological and pathological examination should be performed to achieve the diagnosis of lymphoma.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

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All the authors have contributed to concept, literature search, data acquisition, data analysis, manuscript editing and review.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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