



SECONDARY INTRAOCULAR LYMPHOMA: CASE REPORT

Ophthalmology

Dr Sajad Bashir Khanday	Associate Professor, Department of Ophthalmology, Government Medical College Srinagar, India.
Dr Rabia Saif*	Postgraduate scholar, Department of Ophthalmology, Government Medical College Srinagar, India. *Corresponding Author
Dr Suraya	Consultant, Department of Ophthalmology, Government Medical College Srinagar, India.
Dr Syed Tabinda	Senior Resident, Department of Ophthalmology, Government Medical College Srinagar, India.

ABSTRACT

We report a case of secondary intraocular lymphoma in a 60 year old female who is a known case of Non-Hodgkin lymphoma who came to us with complaint of gradual diminution of vision in both eyes for one month. On fundus examination there were hypopigmented lesions with infiltrates and membranes. B scan showed vitreous opacities suggestive of vitritis.

KEYWORDS

Intraocular lymphoma, Central Nervous System, Vitritis, Vision.

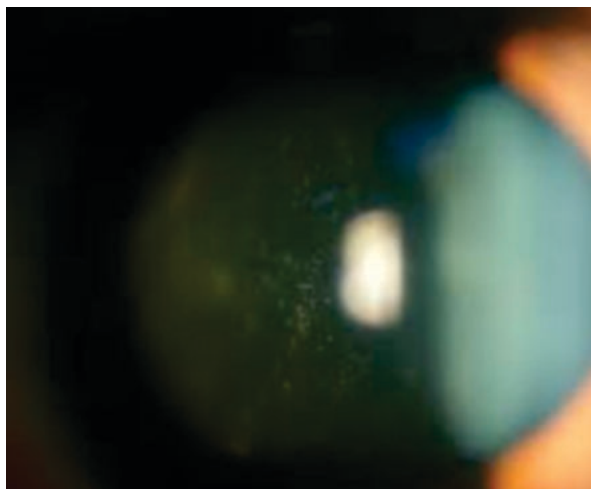
INTRODUCTION

Vitreoretinal Lymphoma currently known as primary intraocular lymphoma is the most common intraocular lympho proliferative disease¹. It is considered a variant of primary central nervous system lymphomas. Rarely, a vitreoretinal lymphoma can be classified as secondary when it arises due to metastasis from a systemic lymphoma^(2,3). Vitreoretinal lymphomas represent <1% of all intraocular tumours¹, 1-2 % of all extra nodal Non-Hodgkin's lymphomas⁵. Women are more commonly affected than men⁴ and patients generally present in 4th to 6th decade. 80-90% patients will have bilateral disease, although initial presentation may be unilateral or asymmetric⁵.

Case Presentation

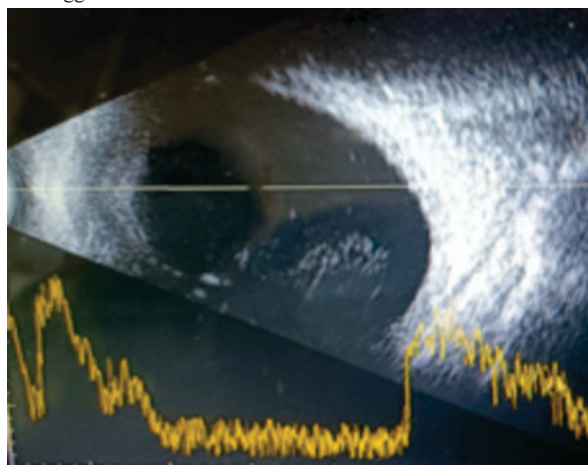
A 60 year old female who is a known case of Non-Hodgkin Lymphoma for which she has received 6 cycles of chemotherapy, presented to outpatient department of our hospital with complaints of diminution of vision of both eyes, left more than right, which was gradual in onset and progressive over 2 month period. There was no history of diabetes mellitus, hypertension or any other comorbidity. There was no history of ocular trauma.

On examination Best corrected visual acuity was 6/36 in the right eye and 6/60 in the left eye, bilateral pupillary reactions were normal. There was right patchy iris atrophy, rest anterior segment examination was normal and IOP was within normal limits in both eyes. On slit lamp, vitritis was seen. On Fundus examination, media was hazy, revealed hypopigmented lesions with infiltrates and membranes in both eyes.

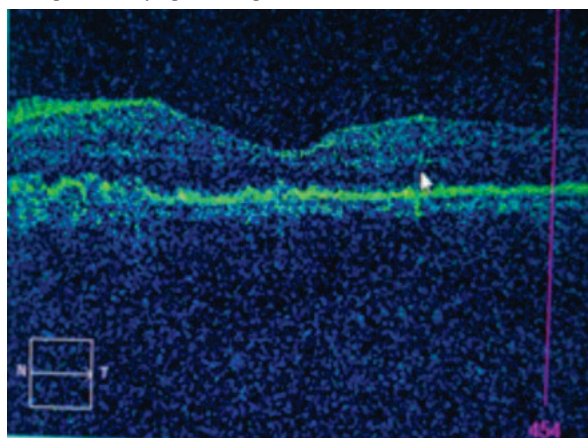


Investigations

1. Vitreous tap was taken and microscopic examination showed dispersed sheets of lymphoid cells and occasional neutrophils in a proteinaceous background.
2. KOH test was negative.
3. B-scan showed hyper reflective vitreous opacities in both eyes suggestive of vitritis.

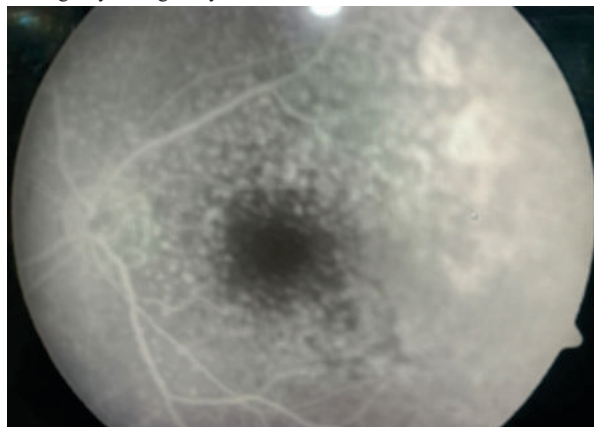


4. OCT of the left eye showed hyperreflective lesions in the retinal pigment epithelial layer corresponding to sub-retinal pigment epithelium lymphoma deposits



OCT of the right eye was normal.

5. Fundus fluorescein angiography(FFA) showed hyperfluorescent round spots which did not increase in area and intensity. FFA of the right eye was grossly normal.



6. PET-CT report showed normal FDG distribution in the head. Non metabolically active cervical, bilateral axillary and mediastinal lymph nodes.

DISCUSSION

Secondary intraocular lymphoma arises from known source of systemic Lymphoma and metastasizes to the eye hematogenously. It disseminates predominantly to the uvea which has rich blood supply or to the vitreous or the retina less commonly⁷. It is typically diffuse large B cell lymphoma type. The major subtype of Ophthalmic Lymphoma is extranodal marginal zone lymphoma of mucosa associated lymphoma tissue (MALT⁷). The lymphoma cells in the ocular central nervous system form of the disease appear to arise primarily in the vitreous and in the sub RPE space and only secondarily extend to the retina and choroid. Patients show a wide variety of pictures that may simulate many ocular disorders. They are usually in the sixth to seventh decade of life^{8,9}.

Young adults and children are occasionally affected¹⁰. The ocular CNS form of large cell lymphoma most frequently masquerades as posterior uveitis in patients complaining of floaters caused by the vitreous infiltration with lymphomatous and inflammatory cells.

Most of the patients develop multiple fundus lesions, which initially may be relatively flat and appear similar to multifocal choroiditis or multiple evanescent white dot syndrome, but typically enlarge to form solitary, sharply defined, blister like yellowish sub-RPE tumours that are usually sufficiently characteristic to allow an accurate diagnosis^{8,9,11}. The fine speckling of pigment on the surface of the amelanotic subretinal mounds is the biomicroscopic clue to the sub-RPE locations of the tumours, which may be confluent and massive in size. Other descriptions include white lesion tumours that may simulate acute retinitis¹², branch artery occlusion by infiltration of the major retinal artery¹³, haemorrhagic infarction of the retina simulating that seen in acute retinal necrosis¹⁴, lymphomatous infiltration of the retrobulbar portion of the optic nerve simulating retrobulbar neuritis or infiltration of the optic nerve head simulating papillitis¹⁵⁻¹⁸, iridocyclitis and secondary glaucoma⁹.

Our patient presented with hypopigmented lesions in the fundus of both eyes. OCT depicted hyper-reflective lesions in the retinal pigment epithelial layer corresponding to subretinal and sub-retinal pigment epithelium deposits. Moreover exfoliative cytology report of the vitreous sample showed dispersed sheets of lymphoid cells and occasional neutrophils in a proteinaceous background. She is currently on chemotherapy for systemic Non-Hodgkin's lymphoma.

Conflict of Interest: Nil

Funding: Nil

REFERENCES

1. Coupland SE, Damato B. Understanding intraocular lymphomas. Clin Experiment Ophthalmol. 2008;36:564-578.
2. Biswas J, Majumdar PD. Uveitis: An Update. GotoH. Intraocular lymphoma. 2016. 93-100.
3. Salomão DR, Pulido JS, Johnston PB, Canal-Fontcuberta I, Feldman AL. Vitreoretinal presentation of secondary large B-cell lymphoma in patients with systemic lymphoma. JAMA Ophthalmol. 2013;131(9):1151-1158.
4. Bardenstein DS. Intraocular lymphoma. Cancer Control. 1998;5:317-325.

5. Freeman LN, Schachat AP, Knox DL, et al. Clinical features laboratory investigations, and survival in ocular reticulum sarcoma. Ophthalmology 1987;94: 1631-1639.
6. Sobolewska B, Chee SP, Zaguia F, Goldstein DA, Smith JR, Fend F, Mochizuki M, Zierhut M. Vitreoretinal lymphoma. Cancers (Basel). 2021;13:3921. 3910.3390/cancers13163921.
7. Gao X, Li B, You Q, Peng X. Primary extranodal marginal zone B-cell lymphoma with diffuse uveal involvement and focal infiltration of the trabecular meshwork: a case report and review of literature. BMC Ophthalmol. 2015;15:48. <https://doi.org/10.1186/s12886-12015-10038-12887>.
8. Gass JD. Stereoscopic atlas of macular diseases: diagnosis and treatment. 4th ed. St. Louis, MO: CV Mosby; 1997.
9. Wender A, Adar A, Maor E, Yassar Y. Primary B-cell lymphoma of the eyes and brain a 3-year-old boy. Arch Ophthalmol. 1994;112(4):450.
10. Givner I. Malignant lymphoma with ocular involvement; a clinico-pathologic report. Am J Ophthalmol. 1990;39(1):29-32.
11. De Smet MD, Nussenblatt RB, Davis JL, Palestine AG. Large cell lymphoma masquerading as a viral retinitis. Int Ophthalmol. 1990;14(5-6):413-7.
12. Gass JDM, Trattler HL. Retinal artery obstruction and atheromas associated with non-Hodgkin's large cell lymphoma (reticulum cell sarcoma). Arch Ophthalmol. 1991;109(8):1134-9.
13. Goder G, Klein S, Konigsdorfer E. [Clinical and pathologic characteristics of malignant lymphoma of the retina] Klin Monatsbl Augenheilkd. 1990; 197(6):514-8. German.
14. Ridley ME, McDonald HR, Sternberg P Jr, Blumenkranz MS, Zarbin MA, Schachat AP. Retinal manifestations of ocular lymphoma (reticulum cell sarcoma). Ophthalmology. 1992;99(7):1153-60; discussion 1160-1.
15. Gray RS, Abrahams JJ, Hufnagel TJ, Kim JH, Lesser RL, Spencer DD. Ghost-cell tumor of the optic chiasm. Primary CNS lymphoma. J Clin Neuro-ophthalmol. 1989;9(2):98-104.
16. Guyer DR, Green WR, Schachat AP, Bastacky S, Miller NR. Bilateral ischemic optic neuropathy and retinal vascular occlusions associated with lymphoma and sepsis. Clinicopathologic correlation. Ophthalmology. 1990; 97(7):882-8.
17. Kattah JC, Suski ET, Killen JY, Smith FP, Limaye SR. Optic neuritis and systemic lymphoma. Am J Ophthalmol. 1980;89(3):431-6.
18. Lang GK, Surer JL, Green WR, Finkelstein D, Michels RG, Maumenee AE. Ocular reticulum cell sarcoma. Clinicopathologic correlation of a case with multifocal lesions. Retina. 1985;5(2):79-86.