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A Case of Rapid Progression and Vision Loss in a Patient with Primary Intraocular Lymphoma

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Abstract

Purpose—To describe an atypical case of primary intraocular lymphoma (PIOL) with rapid progression leading to vision loss and death.

Methods—A 71-year-old woman with a history of PIOL presented with a sudden decline in vision. MRI, CSF cytology, and ocular examination 2 months prior were unremarkable.

Results—Repeat MRI of the brain and orbits at this time revealed a 0.8 × 2.4-cm lesion of the left choroid plexus. LP revealed rare atypical B cells. The patient rapidly deteriorated and died a month later.

Conclusion—Recognition of the potential for rapid progression and mortality in patients with PIOL is essential.

Keywords

Primary intraocular lymphoma; progression; sudden; rapid

Primary intraocular lymphoma (PIOL) is a rare type of malignancy whose progression can be variable. PIOL mostly presents as a diffuse, large B cell lymphoma and frequently occurs in conjunction with primary central nervous system lymphoma (PCNSL).[1] A recent rise in the incidence of PCNSL, likely attributable to increased awareness and detection, has brought about a related increase in PIOL.2 Clinical presentations of PIOL include a wide spectrum of ocular inflammatory processes that may make diagnosis difficult to discover.3 Cases such as this, with a sudden onset and accelerated progression, emphasize the urgency of uncovering an accurate diagnosis.

A 71-year-old Caucasian female presented to NEI with bilateral vitritis. MRI from 2 months prior was negative for central nervous system (CNS) lymphoma. Additionally, normal lumbar puncture (LP) with negative cerebral spinal fluid (CSF) cytology and flow at that time confirmed this finding. She was diagnosed with PIOL OU upon vitrectomy OU, which revealed large atypical lymphoid cells, and was treated with 5 cycles of high-dose systemic methotrexate followed by 10 cycles of methotrexate, procarbazine, and vincristine upon new brain lesions on MRI (negative LP) in about a year. Repeat MRI showed mild age-related

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changes without CNS lymphoma and LP, anterior chamber (AC) tap (prompted by new keratitic precipitates OS) were negative upon completion of therapy. One month later, she developed unexplained fever, weakness, ataxia, headache, and wavy vision. Cytologic analysis from repeat LP done as part of fever workup outside showed no abnormal lymphocytes. Two weeks later, visual acuity OS dropped significantly, from 20/32 to CF (count fingers) at 6 in., with no change OD. Dilated examination OS revealed a severely swollen optic nerve (Figure 1A). A neuro-oncology consult followed by brain MRI (Figure 1B, C), 2 months from the last one, revealed a 0.8 × 2.4-cm lesion of the left choroid plexus with optic chiasm enlargement. PET scan showed increased uptake in the left temporal and right frontal lobes. LP revealed rare atypical B cells. The patient then deteriorated, both visually and physically, and died a month later despite initiation of radiotherapy and decadron.

Due to its rarity, PIOL progression is seldom mentioned in the literature. One review[4] noted median disease-free survival (DFS) at 24 months to be 65%, with overall survival at 85% in treated patients. Another study reported DFS and overall survival at 2 years to be 20 and 40%, respectively, demonstrating the variability of PIOL progression.[1] A report by Gill et al. 3 chronicles a similar course of an 83-year-old male presenting with vitritis and death 4 months after diagnosis of PIOL despite radiation therapy. However, our case appeared to have initially responded to treatment with a subsequent sudden, unexpected decline. Studies to determine predictors of prognosis in PCNSL, summarized by Chan and Gonzales,5 found age greater than 65 and performance status, among others, to be indicative of poor outcome. However, utility of PIOL prognosticators may not align with those of PCNSL. Thus, awareness of the potential for rapid progression and abrupt, subsequent mortality is vital. If suspected, the ophthalmologist should seek experienced radiological and pathological consultation.

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REFERENCES

1. Jahnke K, Korfel A, Komm J, et al. Intraocular lymphoma 2000–2005: results of a retrospective multicentre trial. Graefes Arch Clin Exp Ophthalmol 2006;244:663–669. [PubMed: 16228920]
2. Eby NL, Grufferman S, Flannelly CM, et al. Increasing incidence of primary brain lymphoma in the U.S. Cancer 1988;62:2641–2665.
3. Gill MK, Jampol LM. Variations in the presentation of primary intraocular lymphoma: case reports and a review. Surv Ophthalmol 2001;45:463–471. [PubMed: 11425352]
4. Grimm SA, Pulido JS, Jahnke K, et al. Primary intraocular lymphoma: an International Primary Central Nervous System Lymphoma Collaborative Group Report. Ann Oncol 2007;18:1851–1855. [PubMed: 17804469]
5. Chan, CC.; Gonzales, JA. Primary Intraocular Lymphoma. Hackensack, NJ: World Scientific; 2007. Prognosis in PIOL; p. 197-205.

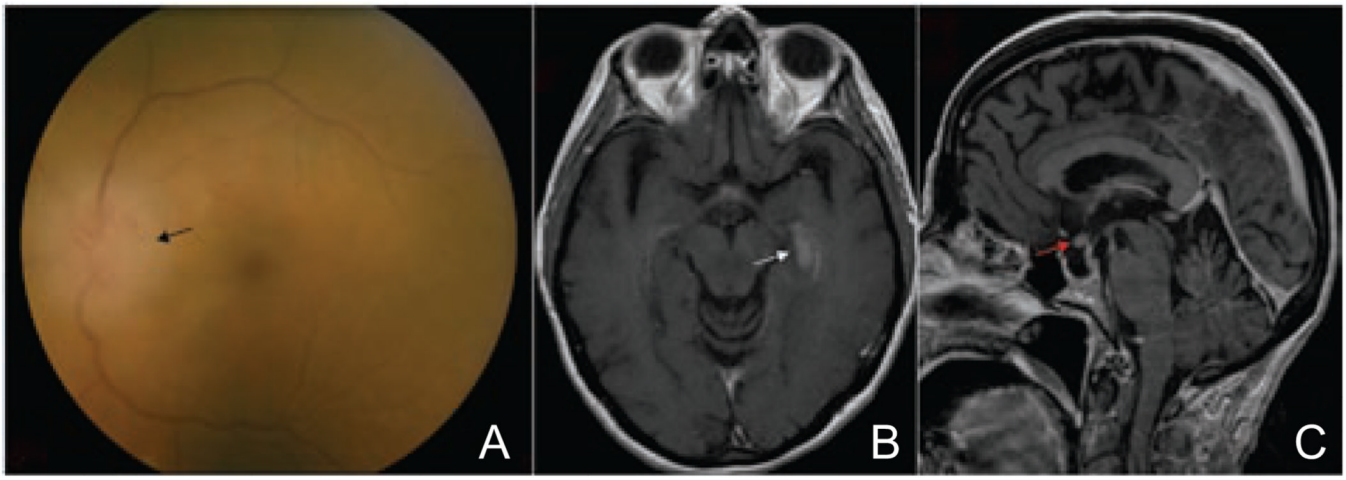

FIGURE 1.

Fundus photograph and MRI scans at time of sudden vision loss in the left eye. (A) Fundus photograph OS demonstrating severe optic disc edema (black arrow). (B) Axial FLAIR-weighted MRI of the brain demonstrating new tubular-shaped lesion (white arrow). (C) Mid-sagittal post-contrast MRI of the brain demonstrating new abnormal enhancement of left optic chiasm (red arrow).