A Case of Primary Vitreo-retinal Lymphoma

Sir,

Primary vitreoretinal lymphoma (PVRL) is a rare extranodal lymphoma, with over 95% of cases being non-Hodgkin diffuse large B-cell lymphomas.[1] PVRL is highly malignant, and approximately 56-90% of patients eventually develop central nervous system (CNS) involvement.[2] Clinically, the ocular manifestations of PVRL lack specificity, often resembling inflammatory or chronic infectious uveitis and vascular diseases, resulting in a high rate of misdiagnosis and delayed diagnosis.[3] With the rising incidence of CNS lymphoma, an ageing population, and advancements in diagnostic techniques, the incidence of PVRL is expected to increase in the future.[4]

A 60-year female was admitted with a 1-week history of declined vision in the left eye. She reported no other ocular or systemic discomforts and denied any history of other systemic diseases, trauma, or surgery. On admission, the right eye visual acuity (VA) was 0.8, and the intraocular pressure (IOP) was 13 mmHg, with no obvious abnormalities in its structures. The left eye VA was 0.02, and the IOP was 15 mmHg. There were no obvious abnormalities in the anterior segment; the vitreous was not significantly turbid; the optic disc margin was clear; there was a bleeding focus in the superior temporal branch vein. Optos ultra-widefield fundus photography showed a bleeding focus in the superior temporal branch vein of the left eye (Figure 1A). Optical coherence tomography (OCT) indicated macular oedema (Figure 1E). Fluorescein fundus angiography (FFA) showed a star-shaped exudate in the macular area of the left eye, retinal oedema below, delayed filling of the superior temporal branch vein, with gradually increasing fluorescein leakage below (Figure 1D). The initial diagnosis was branch retinal vein occlusion with associated macular oedema in the left eye. The patient received intravitreal anti-VEGF (Conbercept) treatment in the left eye.

One month later, on re-examination, the left-eye VA was hand motion (HM) at 20 cm. Dust-like keratic precipitates (KPs) and mild vitreous turbidity was observed. Fundus photography showed yellow-white punctate and patchy lesions, retinal haemorrhage, exudation, and oedema (Figure 1B). Macular OCT showed reduced macular oedema compared with the previous examination (Figure 1F). Vitreous cavity fluid examination revealed IL-6 at 857.3 pg/mL, IL-10 at 17177.3 pg/mL, and an IL-10/IL-6 ratio of 20.0365. The diagnosis was PVRL in the left eye. Immediate intravitreal injection of methotrexate (MTX) was given: twice a week for 4 weeks, once a week for 2 months, and once a month for 9 months. After 8 doses of MTX chemotherapy, on re-examination, the VA of the left eye was still HM at 20 cm, with dust-like KPs and mild vitreous turbidity. Fundus photography showed fewer yellow-white punctate and patchy lesions, more retinal bleeding foci, and less yellow-white exudation and oedema than the previous examination (Figure 1C). Vitreous cavity fluid examination showed IL-6 at 349.5 pg/mL, IL-10 at 2 pg/mL, and an IL-10/IL-6 ratio of 0.0057. The data from the vitreous cavity fluid examination indicated a significant reduction in intraocular inflammation, thereby improving the ocular condition. The patient was remained in this treatment regimen.

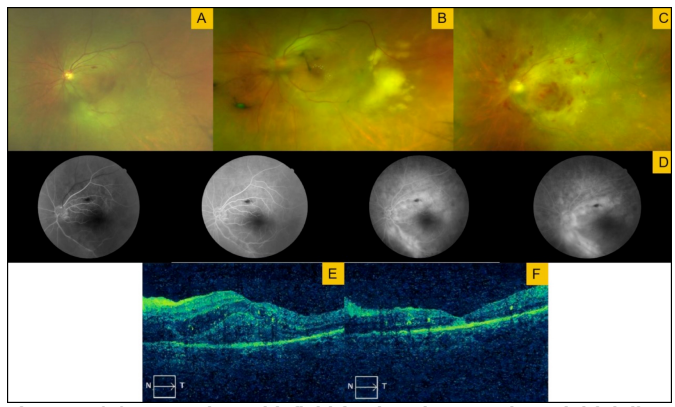

Figure 1: (A) Optos ultra-widefield fundus photography at initial diagnosis showed a bleeding focus in the superior temporal branch vein of the left eye; (B) Optos ultra-widefield fundus photography after anti-VEGF treatment showed yellow-white punctate and patchy lesions in the retina and vitreous turbidity of the left eye; (C) Optos ultra-widefield fundus photography after MTX chemotherapy showed fewer yellow-white punctate and patchy lesions, more retinal bleeding foci, and less exudation and oedema than before; (D) FFA at initial diagnosis showed delayed filling of the superior temporal branch vein of the left eye, with gradually increasing fluorescein leakage below, a star-shaped exudate in the macular area, and retinal oedema below; (E) Macular OCT at initial diagnosis showed macular oedema in the left eye; (F) Macular OCT after anti-VEGF treatment showed reduced macular oedema in the left eye.

Clinicians should enhance their understanding of PVRL and be more vigilant about this rare disease, especially when dealing with patients with atypical fundus lesions or poor responses to conventional treatments. A comprehensive analysis of clinical manifestations, imaging findings, and intraocular fluid specimen tests can improve diagnostic accuracy. Detection of cytokine levels in intraocular fluid (aqueous humor or vitreous) is an important auxiliary diagnostic method for PVRL; therefore, it should be emphasised. The diagnosis and treatment of PVRL require multidisciplinary collaboration among ophthalmology, pathology, haematology, neurology, and laboratory medicine. Through the cooperation of a multi-disciplinary team, diagnostic accuracy and treatment effectiveness can be improved, thereby providing more effective treatment for patients and improving their prognosis.

COMPETING INTEREST:
The authors declared no conflict of interest.

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CW: Drafting, revision, and editing of the manuscript.
SW: Data collection, analysis, and interpretation.
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