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CORRESPONDENCE

Vitreoretinal lymphoma: an alternative view

Eye (2022) 36:1845; https://doi.org/10.1038/s41433-021-01841-2

TO THE EDITOR:

I commend Hearne et al on their effort to raise awareness of vitreoretinal lymphoma (VRL) [1]. However, I respectfully disagree with the authors' interpretation of the term 'primary', which implies that the disease originates intraocularly. This may not be the case, even if systemic investigations are negative. I had a patient with VRL with sheathing and occlusion of the inferotemporal retinal arterioles in the left eye; she subsequently developed lymphomatous deposits throughout the fundus, except in the area of the vascular occlusion. This case suggests a systemic source of the lymphoma cells, which percolate through the retinal pigment epithelium to accumulate at Bruch's membrane [2].

At the University of California, San Francisco, we hypothesised that if VRL is a systemic disease, then systemic treatment is more likely to prolong life than ocular treatment, which we considered to be only symptomatic or palliative. We achieved encouraging ocular and systemic outcomes by treating VRL with systemic chemotherapy, which (importantly) was followed by long-term maintenance immunotherapy to suppress any minimal residual disease [3]. Vitreous infiltrates were more resistant to systemic treatment, whose efficacy increased following therapeutic vitrectomy [4].

Hearne et al. use the term 'primary intraocular lymphoma (PIOL)' synonymously with VRL; however, PIOL includes uveal lymphoma, which is quite different, consisting of low-grade extranodal marginal-zone lymphoma, with an excellent survival probability. PIOL is therefore an umbrella term and should be recognised as such.

Hearne et al. cite an article on metagenomic deep sequencing by Gonzales et al. but missed the opportunity to emphasise the great diagnostic potential of this test, which detects MYD88 mutation, a well-recognised lymphoma biomarker, and which also identifies the organisms causing any infectious uveitis or, indeed, lymphoma [5].

I have seen several patients whose VRL diagnosis was delayed by many months of futile uveitis therapy so that any opportunities for conserving vision and life were missed. These tragic outcomes could be avoided if more ophthalmologists learn to recognise the VRL signs on fundus autofluorescence imaging and optical coherence tomography and if they send these images to an ocular oncologist for a second opinion at the slightest suspicion of this highly lethal disease.

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COMPETING INTERESTS

The author declares no competing interests.

ADDITIONAL INFORMATION

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Received: 23 July 2021 Revised: 9 October 2021 Accepted: 2 November 2021
Published online: 18 November 2021