

## Chapter 1

# Definition of Primary Intraocular Lymphoma

Primary intraocular lymphoma (PIOL) is a neoplasm, most frequently of B-cell, and rarely, T-cell, origin arising from or initially presenting in the sub-retinal pigment epithelium (RPE), retina and vitreous.<sup>1-6</sup> PIOL of the B-cell type is a non-Hodgkin's lymphomatous process of the diffuse large B-cell histologic type and its distinction lies in the fact that it is a subtype of primary central nervous system lymphoma (PCNSL). The neurosensory retina is, in fact, an extension of the CNS. Indeed, the neurosensory retina and retinal pigment epithelium are derived from the same neuroectoderm that forms the CNS during embryogenesis. And, like the retina, the CNS is an immunologically privileged organ as well.

PIOL is distinct from the other lymphomatous processes inside the eye much more commonly including metastatic systemic lymphoma to the eye, which typically affects the uveal tract and ocular adnexa via hematogenous routes; and the extremely rare extranodal marginal zone B-cell lymphoma (mucosa-associated lymphoid tissue lymphoma) that affects the choroid.<sup>7-9</sup> In addition, although a subtype of PCNSL, PIOL may arise *de novo* in the neurosensory retina rather than metastasizing from a primary non-ocular CNS structure.

As an immunologically privileged organ, the internal tissues of the eye are normally protected from inflammatory processes mediated by the immune system's B- and T-cells. Thus, the induction of inflammatory conditions within the eye is intriguing, as inflammatory B- and T-cells are not typically found there. Equally intriguing is the development of lymphomatous processes involving these same cells from which the eye is normally protected.

What is most curious about PCNSL and PIOL is the recent increase in prevalence in these malignancies. Certainly the rising prevalence of immunodeficiency states can explain part of this phenomenon,<sup>10</sup> but concurrent rises in the prevalence rate in immunocompetent patients warrant an in-depth look into this fascinating disease process. To that end, this book serves to be a guide into the history, diagnosis, management and treatment of PIOL, as well as providing insights into the future progress yet to be made in this most interesting and fatal malignancy.

### References

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