Diagnosis of primary vitreoretinal lymphoma, a subtype of primary CNS lymphoma

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Primary vitreoretinal lymphoma (PVRL) is a rare extranodal non-Hodgkin’s lymphoma that originally invades intraocular tissues: the retina, vitreous, and/or optic nerve. PVRL is a subset of primary central nervous system lymphoma (PCNSL). Mostly PVRL belongs to a diffuse large B-cell lymphoma, infrequently of a T-cell type. PVRL, the most common lymphoma inside the eye, is a fatal ocular malignancy, as approximately 65-90% patients with PVRL will develop CNS lymphoma. PVRL often masquerades as chronic uveitis, making it difficult to diagnose. The diagnosis of PVRL requires the identification of lymphoma cells in the vitreous and/or retina by cytology and histopathology. Immunopathology and flow cytometry demonstrate monoclonality. Molecular pathology confirms immunoglobulin or T-cell receptor gene rearrangement in the PVRL cells. Since B-cell lymphoma produce large amount of interleukin-10, cytokine measurement in ocular fluids can be a helpful adjuvant for diagnosing PVRL. However, clinical manifestations and dilated fundus examinations including imaging tests (autofluorescent fundus photography, fluorescent angiography, retinal optical coherence tomography (OCT) imaging) are critical for providing data for the clinician to suspect PVRL and perform tissue biopsy. It often takes more than one invasive procedure to make the diagnosis of PVRL, and diagnostic vitrectomy has a high diagnostic yield compared to spinal tap and aqueous aspiration.

Biography

Chi-Chao Chan earned her M.D. from Johns Hopkins University and ophthalmology residency from Stanford University School of Medicine. She has completed two post-doctoral fellowships: ophthalmic pathology at Wilmer Institute, Johns Hopkins and clinical ocular immunology at National Eye Institute, National Institutes of Health. She is the Chief of Immunopathology Section, Laboratory of Immunology and Head of Histopathology Core, National Eye Institute, the federal government medical research institute in the US. She has published 582 papers in peer-reviewed journals, 51 book chapters, and one textbook. She also serves as an editorial board member for 16 medical journals.

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