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Pars Planitis: A Syndrome of Unknown Etiology or a Clinical Picture of Multiple Etiologies?

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For the first time, In the 1950s, Schepens described what is now known as pars planitis as "peripheral uveitis"; however, the term "pars planitis" was first used by Welch in 1960 to describe a syndrome characterized by peripheral posterior segment inflammation, vitreous opacities and edema of the posterior pole. The major clinical feature in the described disease was the accumulation of yellow-gray inflammatory exudates on the pars plana and ora serrata. Other terms, including chronic cyclitis, vitritis, and peripheral uveoretinitis have also been used to describe this syndrome.

In 1987 the International Uveitis Study Group introduced the term intermediate uveitis (IU) to denote inflammation in this anatomic area, in an attempt to bring order out of the naming chaos. In 2005, the SUN (Standardization of Uveitis Nomenclature) Working Group defined intermediate uveitis anatomically and used standardized grading for vitreous inflammation (vitreous haze), which is the key clinical feature. Since then, the term IU has been used to identify any inflammation which is primarily in the region of vitreous, pars plana and peripheral retina. The term pars planitis is reserved for idiopathic IU in which the principal clinical finding is inflammatory exudation over the pars plana, without an identifiable underlying systemic or ocular disease. The cause of pars planitis, whether endogenous or exogenous, has not been identified, although an immune-mediated mechanism has been proposed. Histopathologic studies have been performed in only a few patients, and no relevant animal model of pars planitis exists.

In the majority of patients with IU, the condition is an isolated ocular syndrome (pars planitis); however, a number of systemic diseases have been associated with IU, including multiple sclerosis (MS), sarcoidosis, Adamantiades-Behçet disease (ABD), Lyme disease, seronegative spondyloarthropathies associated with HLA-B27, tuberculosis, and brucellosis, all of which may

present with uveitis restricted to the intermediate segment of the eye. We have also reported cat-scratch disease as a cause of IU.

In our cohort of 33 patients diagnosed with pars planitis, 18 had or were suspected of having sarcoidosis, 7 had multiple sclerosis, and 5 had miscellaneous immunologic diseases. Patients with systemic disease usually presented with granulomatous uveitis, whereas those with pars planitis usually presented with non-granulomatous uveitis ($p = 0.003$). Posterior synechiae formation was the only other finding with a statistically significant difference between the two groups ($p = 0.024$). The clinical course and final outcomes of pars planitis were comparable between the groups.

In conclusion, we found similarities in the clinical findings, natural course, and outcomes of noninfectious intermediate uveitis (IU) and pars planitis between patients with no evidence of underlying systemic disease and those with proven or suspected systemic disease of immunologic origin. These similarities may represent indirect evidence of a common pathogenetic mechanism and shared therapeutic needs.

References

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