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LETTER TO THE EDITOR

Chronic Anterior Uveitis in Common Variable Immunodeficiency

Erica Z. Oltra, MD[1], Cody Morris, MD[1], Andrea D. Birnbaum, MD, PhD[2], Howard H. Tessler, MD[1], and Debra A. Goldstein, MD[1]

[1]*Illinois Eye and Ear Infirmary, Department of Ophthalmology and Visual Sciences, University of Illinois at Chicago, Chicago, Illinois, USA, and* [2]*Feinberg School of Medicine, Northwestern University, Department of Ophthalmology, Chicago, Illinois, USA*

ABSTRACT

Purpose: To describe patients with uveitis and common variable immunodeficiency (CVID).

Design: Retrospective observational case series.

Methods: Retrospective review of patients with uveitis and CVID, specifically focusing on clinical presentation and treatment.

Results: Three patients with CVID and uveitis were identified. All patients had chronic anterior uveitis. Two required systemic immunosuppression to control uveitis, with one of these patients relapsing after discontinuation of immunoglobulin therapy. One improved on topical steroid therapy; however, follow-up on this patient did not extend beyond 4 months.

Conclusions: CVID can be associated with chronic anterior uveitis. Patients presenting with uveitis should be questioned about frequent infections and autoimmune disorders as part of the uveitis evaluation.

Keywords: common variable immunodeficiency, iridocyclitis, iritis, sarcoidosis, uveitis

Common variable immunodeficiency (CVID) is the most common primary immunodeficiency. It is characterized by recurrent pyogenic sinopulmonary infections as a result of defective antibody formation and hypogammaglobulinemia.[1] Autoimmune disorders have been reported in up to 20% of patients with CVID and are frequently the first manifestation.[2]

A sarcoid-like variant of CVID has been described, in which patients develop noncaseating granulomas in liver, lungs, spleen, lymph nodes, or eyes, and serum ACE may be elevated.[2,3] Conjunctival granulomas, granulomatous anterior and posterior uveitis, multifocal choroiditis, disc swelling, retinal vasculitis, and retinal neovascularization have been reported.[4-8] We describe the first series of patients with CVID and anterior uveitis without posterior involvement.

MATERIALS AND METHODS

Retrospective review of patients with uveitis and CVID seen at the uveitis service of the University of Illinois Chicago and the private office of one of the authors from 2005 to 2010. Approval for this study was obtained from the University of Illinois–Chicago Institutional Review Board.

RESULTS

Three patients were identified with CVID and uveitis. All three presented with a known history of CVID and were receiving monthly intravenous immunoglobulin (IVIG) therapy.

Patient 1

Patient 1 was a 53-year-old Caucasian female with blurred vision and floaters in both eyes (OU) that began 2 years before when she was diagnosed with iritis. At presentation to our referral center, best-corrected visual acuity (BCVA) was 20/50 right eye (OD) and 20/60 left eye (OS). Examination revealed

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Correspondence: Debra Goldstein, 1905 West Taylor St. Chicago, IL 60612. Email: Debrgold@yahoo.com

Koeppe nodules, 2+ flare and 3+ cells in the anterior chamber (AC) and anterior vitreous cells OU. Cystoid macular edema was noted bilaterally and confirmed with optical coherence tomography. Investigation revealed an elevated ACE level of 114 U/L (normal 9–67 U/L), elevated lysozyme of 16.4 μg/mL (normal 4–10.3 μg/mL), negative QuantiFERON Gold test, and negative FTA-ABS. Chest radiography (CXR) revealed mild perihilar scarring. Vision improved to 20/30 OU within 6 weeks of initiating difluprednate drops, but she was lost to follow-up after 4 months.

Patient 2

Patient 2 was a 53-year-old Caucasian female who presented with floaters and decreased vision OS. In addition to CVID, she had a history of multiorgan granulomas. BCVA was 20/25 OU. Examination OD revealed 1+ AC cells, while OS had granulomatous KP, 1 + AC flare, 3+ AC cells, and rare anterior vitreous cells. She had a negative purified protein derivative (PPD) test, though ACE and lysozyme results were not available. Despite therapy with topical corticosteroids and systemic infliximab, she continued to have chronic iritis and systemic granulomas throughout 5 years of follow-up.

Patient 3

Patient 3 was an 11-year-old Caucasian female diagnosed with juvenile idiopathic arthritis (JIA) at the age of 17 months and CVID at age 5. Routine ocular examination at age 18 months revealed nongranulomatous iritis OU. Uveitis and systemic disease were controlled with systemic infliximab; however, in her fourth year of follow-up she developed a recurrence of uveitis five months after IVIG was discontinued.

DISCUSSION

There are few cases of uveitis associated with CVID reported in the literature.[4,5,7,8] All 3 patients in this series had chronic anterior uveitis with no posterior involvement. The 2 adults had symptomatic granulomatous disease and the child had asymptomatic nongranulomatous uveitis, diagnosed on screening because of JIA. It is possible that the child's uveitis was related to the diagnosis of JIA, although the recurrence of uveitis after stopping IVIG argues for a contribution from CVID. Two patients required systemic immunosuppressive therapy. One improved on topical corticosteroid therapy, but follow-up on this patient was limited.

The first patient had an elevated serum ACE and perihilar scarring on CXR and the second patient had systemic granulomas, which raises the possibility of sarcoid as a co-diagnosis. This phenomenon has been described in CVID patients as both concurrent sarcoidosis[9,10] as well as a sarcoid-like syndrome.[3,7] Fasano describes 8 patients with sarcoidosis and CVID and identifies 22 others in a literature review.[10]

Whether this represents sarcoid or a sarcoid-variant is unclear. Recent studies on the pathogenesis of CVID suggest that the general immaturity of B cells leads to a lack of capacity for isotype switching, which results in a reduction in switched memory B cells. This lack of capacity for isotype switching can cause the retention of autoimmune clones as well as other abnormal immunological phenotypes, leading to inappropriate macrophage and epithelioid activation, resulting in granuloma formation.[3]

CVID has a rare association with uveitis, but ophthalmologists should consider the diagnosis of CVID in uveitis patients with a history of recurrent infections. Similarly, physicians caring for patients with CVID should be aware of the association with uveitis.

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