Images in Rheumatology

A Recurrent Central Band Keratopathy in a Child

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For the first time, to our knowledge, we present the case of a child with simultaneous occurrence of a C3-glomerulopathy and an anterior uveitis complicated with severe band keratopathy (BK). In addition, repeated evaluations of uveitis disclosed important differences between slit lamp and laser flare photometry (LFP) results due to BK, making treatment choice difficult.

A 6-year-old boy presented with a red, painful left eye as well as face edema with macroscopic hematuria. He had also proteinuria in a nephrotic range, moderate hypertension, and mild decrease of glomerular filtration. Diagnoses of simultaneous unilateral anterior nongranulomatous uveitis and biopsy-proven C3G were made.

Etiologic investigations of uveitis included infectious (tuberculosis, syphilis, lyme, bartonellosis, toxocarosis, toxoplasmosis,

leptospirosis, HIV, hepatitis B virus, hepatitis C virus, herpes simplex virus, cytomegalovirus, Epstein-Barr virus: all negative; varicella zoster virus former infection IgG-positive), and inflammatory [no evidence of sarcoidosis, arthritis, systemic lupus erythematosus or other connectivitis, inflammatory bowel disease, vasculitis, and hepatic transaminases, angiotensin-converting enzyme, serum protein electrophoresis normal; HLA-B27-negative, antinuclear antibody (ANA)- and rheumatoid factor (RF)-negative, antisaccharomyces cerevisiae antibody- and antineutrophil cytoplasmic autoantibody-negative; pulmonary radiographs normal] markers. Etiologic investigations of C3G included no familial history of glomerulonephritis, CH50 and C4 normal, C3 chronic hypocomplementemia, comprehensive evaluation of alternate complement

Figure 1. Left eye, recurrence of central band keratopathy, 3 months after the second surgical procedure.

pathway [factor H and I antigens normal, no anti-factor H antibody, monocyte chemotactic protein normal, low factor B antigen and positive antifactor B antibody, positive C3 nephritic factor (C3Nef)]; ANA, RF, and cryoglobulinemia were negative. In summary, etiologic investigations of C3G showed autoimmunity against regulators of the complement alternate pathway with positive C3Nef and antifactor B, and no additional cause for uveitis. C3G healed with steroids in 2 years, but uveitis became chronic despite topical steroids and was complicated with BK and cataract, 3 years after disease onset¹. He was 9 years old when he was referred to us. Visual acuity (VA) of the left eye was 5/10. Full control of uveitis was achieved with methotrexate (MTX), reaching 6/10 in VA. At the age of 13, ocular inflammation relapsed, then BK gradually covered the visual axis, leading to major visual loss in 1 year. Subsequently, he had cataract extraction and 2 mechanical scrappings with EDTA because of early recurrence of central BK. Three months after the second surgery, at 15 years of age, VA worsened to 1/20. Central BK had recurred (Figure 1) and macular edema appeared for the first time. BK relapses suggested that inflammation was not correctly controlled. Low/absent cellular Tyndall with slit lamp contrasted with high LFP measures evaluated the same day during the last 2 years². Treatment was intensified with adalimumab added to MTX³; this combination put a quick end to macular edema. Six months later, BK was again removed by surgery. VA improved up to 9/10, and the patient remained stable until the present time, 4 years since the surgery; he is now 20 years old.

To our knowledge, the simultaneous occurrence of anterior uveitis with biopsy-proven C3-glomerulonephritis (C3G) has not been reported before, but this case suggests the possible involvement of uncontrolled alternate complement pathway in the pathogenesis of both diseases. In addition, this case shows the importance of suppressing ocular inflammation to avoid extension and relapses of BK in the setting of uveitis-dependent BK, sometimes requiring biologics. In this situation, LFP may help detect the presence of hidden inflammation behind BK.

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