

Epstein Barr Virus Dacryoadenitis Resulting in Keratoconjunctivitis Sicca in a Child

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We evaluated a 10 year-old male with bilateral severe dry eye who was profoundly disabled by pain and photophobia despite aggressive conventional lubricant and tear conservation therapy. Serologic studies were initially unrevealing and misleading. A lacrimal gland biopsy, orbital portion, left side, was performed, and tissue was processed for light and immunohistochemical studies, which enabled us to establish the diagnosis.

The boy, six months prior to evaluation, had been swimming in a river in Arkansas with friends, and all developed "conjunctivitis" after this outing, but only the patient's ocular symptoms persisted, with bilateral lid swelling, conjunctival injection, and ocular discomfort. Viral conjunctivitis was diagnosed and treated with topical tobramycin and dexamethasone four times daily for two weeks without effect. Increasingly severe superficial punctate keratopathy developed, and Pred Forte every two hours and Decadron ointment at bedtime along with Ciloxan drops every four hours and lubricants were prescribed. Three weeks of this therapy was also not effective in reducing the signs and symptoms. Schirmer values were zero bilaterally, cornea sensitivity was normal bilaterally, and all cultures were negative. Serologic studies disclosed elevated alkaline phosphatase and aspartate aminotransferase, and serum angiotensin converting enzyme determination was elevated and gallium scanning disclosed abnormal uptake of gallium citrate in the parotid and lacrimal glands. All other studies were within normal limits, and HLA typing was not diagnostically helpful. The diagnosis of sarcoidosis was made, and the patient was treated with systemic Prednisone, 100 mg PO QD, without noticeable effect on the ocular signs and symptoms. The Prednisone was therefore tapered and the patient was referred to us.

Our examination disclosed an extremely photophobic patient who has been housebound and essentially socially disabled since the onset of his problem. He was nearly unexaminable in the clinic, although a detailed examination was accomplished under anesthesia. The best corrected acuities in the darkened clinic room were 20/100 OD and 20/70 OS. The examination disclosed 2+ conjunctival injection bilaterally, abundant mucus secretion, and 2+ superficial punctate epitheliopathy. The palpebral lobe of the lacrimal glands were enlarged bilaterally, and the rest of the examination was unremarkable.

Our serologic studies disclosed high antibody titers to EBV viral capsid antigen, nuclear antigen, and early restricted antigen.

The lacrimal gland biopsy disclosed no evidence of granulomatous inflammation, a significant amount of normal-appearing lacrimal gland, and focal dacryoadenitis with lymphocytic infiltrate and foci with neutrophil infiltration inside the tubules; small numbers of acini were scarred.

Immunohistochemical analysis disclosed almost no secretory IgA, indicating marked dysfunction of even the normal-appearing acini. EBV viral proteins EBNA-2 and VCA were identified by immunohistochemistry in the areas of lymphoproliferation, and EBV early restricted antigen was present in the tubular epithelial cells. No such staining was seen in control, normal lacrimal gland tissue.

We thought that the likelihood was relatively high that the patient had acute keratoconjunctivitis sicca most likely caused by EBV infection. Therapy with Acyclovir, 640 mg. IV four times daily for two weeks was employed, and then, in an effort to suppress what we presumed to be the immune/inflammatory host response producing lacrimal gland inflammation, we treated the patient with systemic Cyclosporin, 50 mg. PO BID. At the last evaluation, 12 months after the patient's initial visit with us, he was asymptomatic, with bilateral visual acuities of 20/20 OU, a large tear meniscus, and Schirmer values of 15 mm and 10 mm, right and left eye respectively. The EBV early restricted antigen titer had fallen from its abnormal level to within normal limits, and liver enzymes and the ACE level returned to normal.

We believe that this patient had Epstein-Barr virus induced severe keratoconjunctivitis sicca. Six months of immunomodulatory therapy was required, in addition to high dose intravenous Acyclovir therapy, to abort both the chronic infection and the infection-induced (presumed) autoimmune dacryoadenitis.