Idiopathic CD4⁺ Lymphocytopenia and Sjogren Syndrome

Idiopathic CD4⁺ lymphocytopenia (ICL) is a rare syndrome that is marked by a CD4⁺ count that is less than 300 cells/mm³ without human immunodeficiency virus infection. Its course differs from that of AIDS in that although patients with this disorder may develop opportunistic infections, the majority of them remain stable. No transmissible agent has been implicated in the pathogenesis of ICL. The ocular manifestations of ICL have only rarely been described, and there are no reports of ICL in ophthalmology literature. We report the case of a patient with ICL and Sjogren syndrome.

Report of a Case. A 52-year-old woman was referred to the ophthalmology department because of a several-year history of burning and stinging in both eyes. Her medical history was significant for ICL, with 5 CD4⁺ counts during 6 years ranging from 93 to 253, despite 3 negative human immunodeficiency virus test results. Additionally, assays for Epstein-Barr virus, cytomegalovirus, and human herpesviruses 6 and 8 were all negative. At the time, her visual acuity was 20/20 OD and 20/25 OS. The patient had marked superficial punctate keratitis and abundant mucus production in both eyes, and as a result, she began a course of applying artificial tears to both eyes every 2 hours with only minimal relief.

During the ensuing months, a bandage contact lens was placed over the patient’s left eye, but it failed to relieve her symptoms. Schirmer testing with topical anesthesia showed 6 mm of tearing in the right eye and 5.5 mm in the left. Subsequent bilateral inferior punctual plug placement provided some relief, but her symptoms and superficial punctate keratitis persisted. In addition to the aggressive use of artificial tears, other modalities (corticosteroid eye drops and systemic doxycycline administration) were employed, but the patient’s condition did not improve.

A diagnosis of Sjogren syndrome was confirmed after testing showed a Sjogren syndrome antigen antibody level of 13.1 (range, 0–4.9 U/mL). The patient then began a course of cevimeline hydrochloride (30 mg by mouth 3 times a day), and her symptoms improved considerably. Furthermore, her superficial punctate keratitis diminished appreciably. She remains stable and comfortable on this regimen with the use of artificial tears 4 times per day.

Comment. Idiopathic CD4⁺ lymphocytopenia is a rare disorder of CD4⁺ lymphocytopenia without human immunodeficiency virus infection. The ophthalmic sequelae of this syndrome have not yet been elucidated. In this report, we describe the characteristics and clinical courses of a patient with ICL and Sjogren syndrome.

The underlying pathophysiology of ICL results from apoptosis of CD4⁺ T-cells in idiopathic CD4⁺ lymphocytopenia. Autoimmune processes such as Sjogren syndrome may result from restriction of T-cell diversity, which may lead to a subsequent decrease in immune surveillance. This scenario would allow autoantibodies that may otherwise be cleared from systemic circulation to flourish. Kirtava et al. found an increased prevalence of ICL among patients with Sjogren syndrome.

In summary, both ophthalmologists and internists should be aware of the connection between Sjogren syndrome and patients with ICL. Further evaluation is necessary to determine other ocular manifestations of ICL.

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Advanced Keratomalacia With Descemetocele in an Infant With Cystic Fibrosis

Xerophthalmia refers to the spectrum of ocular manifestations of vitamin A deficiency. It represents the leading cause of childhood blindness worldwide but is uncommon in industrialized countries, where xerophthalmia is more often the result of malabsorption than malnutrition due to poverty. Cystic fibrosis (CF) is an autosomal recessive disease with hyperviscosity of mucus secretions causing chronic pulmonary changes and pancreatic insufficiency. Anderson was the first to note the association between xerophthalmia and CF, now thought to be due to fat malabsorption resulting in fat-soluble vitamin deficiency. Advanced xerophthalmia has been reported as an initial sign of CF. A recent review article summarized the ocular findings of CF to include xerophthalmia, tear film abnormalities, papilledema, and nystagmus. To our knowledge, this is the first clinicopathologic report of keratomalacia with a descemetocele requiring keratoplasty as the initial manifestation of CF.

Report of a Case. A 5-month-old girl from Juarez, Mexico, was admitted to a hospital in Las Cruces, NM, with...