Bilateral Neuroretinitis in a 6-Year-Old Boy With Acquired Toxoplasmosis

Toxoplasmosis is a widespread infection in nature that affects both humans and animals and is caused by the intracellular parasite, Toxoplasma gondii. The disease can be congenital or acquired.

Ocular toxoplasmosis can represent a recurrence of congenital disease or can be secondary to postnatal infection. The hallmark of the disease includes focal necroizing retinochoroiditis that ultimately results in a characteristic adjacent or nearby retinochoroidal atrophic scar. However, a variety of less common, “atypical” presentations may occur, e.g., punctate outer retinal toxoplasmosis, retinal vasculitis, retinal vascular occlusions, rhegmatogenous and serous retinal detachments, unilateral pigmentary retinopathy mimicking retinitis pigmentosa, neuroretinitis and other forms of optic neuropathy, and scleritis. Although in the past most cases of ocular toxoplasmosis were considered to be the result of the reactivation of congenital infections, it has been ascertained that many postnatally acquired infections are also responsible for the ocular involvement.

Herein, we report, what is to our knowledge, the first case of bilateral neuroretinitis associated with acquired toxoplasmosis that resulted in an optic nerve atrophy in an immunocompetent child.

Report of a Case. A previously healthy 6-year-old white boy was referred because of a 1-week history of painless loss of visual acuity and intermittent headaches. His visual acuity was 20/400 OU and a bilateral marked optic disc edema was found. No abnormalities were seen on a computed tomographic scan of his head. His medical history was only remarkable for a sideropenic anemia, diagnosed 18 months before the onset of his ocular symptoms, that was treated with iron supplementation. To resolve the anemia, his mother also fed him undercooked meat at least 3 times weekly. At the time of consultation the patient was afebrile with no adenopathy, skin lesions, or unusual findings on external ophthalmic examination. A magnetic resonance imaging (MRI) scan revealed a bilateral enlargement of the optic nerve (Figure 1). The patient was otherwise in good health with no history of focal neurologic deficit or other symptoms suggestive of demyelinating disease. He had no ocular or systemic diseases such as diabetes mellitus or hypertension that might account for disc edema. A further ophthalmologic examination performed a week later showed a very slight Tyndall effect and cells in the anterior chamber, vitreous inflammation, and bilateral optic disc edema with a macular star (Figure 2). Values from the following laboratory tests were normal: complete blood cell, erythrocyte sedimentation rate, antinuclear and anti-DNA antibodies, rheumatoid factor, fluorescent treponemal antibody absorption test, and tuberculin skin test; serologic test results were negative for Rickettsia typhi, Epstein-Barr virus, Leptospira interrogans, Toxocara canis, Bartonella henselae, Borrelia burgdorferi; and findings from a chest radiograph were normal.

The patient had serologic evidence of exposure to T gondii, which was verified by enzyme-linked immunosorbent assay (ELISA) and an indirect fluorescent antibody (IFA) test. Results of the serum ELISA for anti-Toxoplasma IgG were strongly positive at 552 IU, while results for anti-Toxoplasma IgM and IgA were negative. These results were confirmed by IFA (IgG titer, 1:256). A rise in the IgG antibody titer was found 1 month later.

Findings from previous serologic testing for toxoplasmosis performed on the patient during a febrile episode 2 years prior to our observation were negative. The mother was negative for anti-Toxoplasma antibodies during her pregnancy with this child and also at the time of our observation. No laboratory evidence of human immunodeficiency virus or other known causes of immunodeficiency were found in the child. The serum immunoglobulins and lymphocyte subset values were nor-
Based on these results, a presumptive diagnosis of ocular acquired toxoplasmosis in an immunocompetent child was made. An 8-week treatment with sulfadiazine, pyrimethamine, folinic acid supplementation, and prednisone was started. Since pyrimethamine may cause bone marrow depression, a weekly complete blood cell count was performed. No side effects were reported. Ophthalmologic follow-up after treatment showed a recovery from the optic disc edema and macular star, with a residual bilateral optic nerve atrophy (Figure 3). The MRI scan showed a normal optic nerve (Figure 4). The visual acuity was 20/50 OU and remained unchanged throughout a 15-month follow-up.

Figure 1. Magnetic resonance imaging scan (T1-weighted image) on admission. Note the enlargement of both optic nerves.

Figure 2. Funduscopic examination on admission. Optic disc edema and macular star are seen in both the right (A) and left (B) eyes.
Comment. As early as 1980, Gilbert reported 2 cases of ocular isolated lesion without systemic clinical manifestations of toxoplasmosis: the first was with retinal and optic nerve neovascularization, whereas the second had optic nerve edema associated with a macular star. In 1992, Moreno and coworkers reported a case of neuroretinitis as an unusual presentation of ocular toxoplasmosis. In 1993, Fish et al described 5 patients with unilateral neuroretinitis due to toxoplasmosis who developed a sudden decrease in visual acuity.

Figure 3. Funduscopic examination after therapy. Optic nerve atrophy is seen in both the right (A) and left (B) eyes.

Figure 4. Magnetic resonance imaging scan (T1-weighted image) after therapy shows normal optic nerves.
ity with optic nerve edema, vitreous inflammation, and macular star formation. In all of these patients the ocular involvement was always unilateral. In the case we report herein neuroretinitis was bilateral and associated with an elevated ELISA IgG Toxoplasma titer. The patient's favorable response to treatment also confirmed the diagnosis.

Neuroretinitis may be secondary to a panoply of specific infections, eg, syphilis, tuberculosis, leptospirosis, herpes, mumps, bacterial sepsis, toxoplasmosis, Lyme disease, or cat-scratch disease. Autoimmune or neoplastic diseases, hypertensive retinopathy, diabetic optic neuropathy, and melanocytoma must also be included in the differential diagnosis, but most cases remain idiopathic. Most cases of neuroretinitis are self-limited without a tendency toward recurrence and the patients involved have a good to excellent return to their preillness visual acuity.

The diagnosis in our patient had been debated because the patient showed only bilateral optic disc edema and he had no immunosuppressive factors. His clinical picture was not easily recognized as ocular toxoplasmosis, and only later on did he develop a macular star. The presence of a macular star in a patient with optic disc edema helps the diagnosis and prognosis because it militates strongly against cerebral neoplasm or the subsequent development of multiple sclerosis. The absence of preexisting scars or anterior chamber inflammation in our patient could lead to a misdiagnosis of idiopathic neuroretinitis, a benign, self-limited condition requiring no treatment.

It is unclear why a bilateral aggressive ocular toxoplasmosis, typically seen in patients who are frankly immunosuppressed, developed in our patient. Several hypotheses may be considered, such as infection at an early age, long-term antigen exposure, the role of reinfection, and more virulent strains of T gondii, as well as genetic differences between hosts in the immune response against the parasite. Since the patient showed no symptoms or signs of immunodeficiency, we hypothesize the involvement of factors linked to a parasite. Clinicians should be aware that bilateral optic nerve involvement may be a manifestation of toxoplasmosis responsible for a severe visual handicap despite treatment efforts.

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