Diffuse Bilateral Subacute Neuroretinitis

First Patient With Documented Nematodes in Both Eyes

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Objective: To describe the first patient with documented evidence of diffuse unilateral subacute neuroretinitis (DUSN) in both eyes.

Methods: A 10-year-old healthy Brazilian girl was first seen with signs of late-stage DUSN in both eyes. A careful search for a nematode was performed in each eye.

Results: A motile 550- to 660-µm nematode was found in the inferotemporal retina of the left eye. A similarly sized motile nematode was found in the superotemporal retina of the right eye. Both nematodes were treated with argon green laser applications with bilateral improvement of visual function.

Conclusion: Although most patients with DUSN do not develop the disease in the fellow eye, this case demonstrates that DUSN can occasionally affect both eyes.


Classically, diffuse unilateral subacute neuroretinitis (DUSN) is a clinical syndrome that affects only 1 eye. Early-stage DUSN is characterized by visual complaints, vitritis, papillitis, and often recurrent crops of evanescent gray-white outer retinal lesions; late-stage DUSN is characterized by progressive visual loss, optic atrophy, retinal vessel narrowing, and focal and diffuse retinal pigment epithelial degenerative changes in otherwise healthy patients. Diffuse unilateral subacute neuroretinitis is caused by a motile, white, often glistening nematode that can wander for months or years in the subretinal space. It can be found at any stage of the disease, and should be sought, even in patients with optic atrophy and advanced degenerative changes in the pigment epithelium and retina. Two sizes of unidentified nematodes cause DUSN: most commonly, a small nematode—400 to 700 µm long—in endemic areas, including the southeastern United States, the Caribbean, and South America (Brazil). Less frequently, a larger nematode—1500 to 2000 µm long—has occurred in patients with DUSN in the northern midwestern United States, Germany, and, recently, Brazil. Early signs of DUSN are often mistaken for multifocal choroiditis, acute posterior multifocal placoid pigment epitheliopathy, multiple evanescent white dot syndrome, or nonspecific optic neuritis and papillitis. Late signs of DUSN include multifocal chorioretinal scars, diffuse degenerative changes in the pigment epithelium, narrowing of the retinal arteries, and optic atrophy, which are sufficiently characteristic that the correct diagnosis is often considered when the patient is first encountered. Herein, we describe such a situation in a young Brazilian patient with bilateral late-stage DUSN and documented living small nematodes in both eyes.

Report of a Case

A 10-year-old healthy white girl was first seen with a 3-year history of progressive visual loss in her right eye. She had previously developed poor vision associated with strabismus in her left eye. There was no history of ocular inflammation or pain. The patient was the product of a normal pregnancy and delivery, weighing 2900 g at birth. There was no history of cutaneous larval migrans. She had contact with dogs during her childhood in a rural community in São Paulo, Brazil. Treatment for her eye disorder with corticosteroids, antibiotics, and anthelmintic drugs on several occasions resulted in no improvement.

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Visual acuity corrected with a pinhole was 20/400 OD and light perception only in the left eye. There was a relative afferent pupillary defect in both eyes and an exotropia in the left eye. Intraocular press-
sure was 8 mm Hg OU. A few cells were present in the anterior vitreous of both eyes, which were otherwise clear. The left fundus had severe optic atrophy, narrowing and whitening of the retinal arteries, and focal and widespread retinal pigment epithelial degeneration (Figure 1). Similar but less severe changes were present in the right eye. Retinal pigment epithelial degeneration was more localized in the right eye, especially in the nasal middle and temporal peripheral fundus (Figure 2). Diffuse unilateral subacute neuroretinitis was suspected in both eyes. A motile 550- to 660-µm nematode was found in the left eye inferotemporally (Figure 3). There were several neighboring focal chorioretinal scars but no definite active outer retinal lesions. Multiple fundus examinations of the right eye were required before finding a similar-sized nematode in the superotemporal retina near a cluster of more peripheral outer retinal active white lesions (Figure 4, left and right). Both nematodes were treated with continuous 150- to 160-mW, 150-µm spot size argon green laser applications.

Results of laboratory investigations revealed normal erythrocyte sedimentation rates and blood cell counts. Serum enzyme-linked immunosorbent assay titer for Toxocara canis was strongly positive (1/2948 [compared with the laboratory cutoff value of 1/500 to avoid cross-reaction with other ascaris agents]). Stool examination results were positive for Ascaris lumbricoides.

Four weeks after laser treatment, her visual acuity improved to 20/100 OD and the active outer retinal lesions were no longer present. Six months after laser treatment, visual acuity was 20/80 OD and visual field by Goldmann perimetry had improved markedly (Figure 5, left and right). She had hand motion vision in the left eye.

To our knowledge, this case represents the first patient with documented evidence of DUSN in both eyes. One of us (J.D.M.G.) has seen several patients with sus-
expected bilateral DUSN in the early and late phases of the disease, but in no patient was the diagnosis confirmed by discovery of the subretinal nematode. In the literature, only 1 suspected bilateral case in the late phase was briefly reported in 1978. Although evidence suggests that most patients with DUSN will not develop it in the fellow eye, such a diagnosis must be considered in patients with signs of DUSN occurring in both eyes. Early detection of the nematode and its destruction with photocoagulation can prevent severe visual loss. Use of anthelmintic agents is ineffective, except in those few patients with moderately severe vitritis.

Neither the small nor the larger nematode responsible for DUSN has been identified. The association of cutaneous larval migrans in some patients with DUSN suggests that Ancylostoma caninum may be the small nematode that causes the syndrome. Contact with raccoons in 2 patients was cited as circumstantial evidence that Baylisascaris procyonis may be the large nematode that causes DUSN. This nematode tends to involve the central nervous system, yet none of the reported cases of a large nematode in patients with DUSN have shown evidence of such involvement. Most patients with DUSN have no history of exposure to raccoons. Although *T. canis* was implicated morphologically after examination of a partly decomposed or poorly fixated nematode removed by vitrectomy from a patient with DUSN, there is considerable evidence that most cases of DUSN are unrelated to this organism. The clinical picture of intracocular toxocariasis, its worldwide distribution, and its strong association with serologic evidence of systemic infection contrast with that occurring in DUSN. The strongly positive serologic test result for toxocariasis in our patient is of interest, yet in a child with evidence of other intestinal parasites, might be nothing more than coincidental to the eye findings.

Awareness of the multiple early and late ophthalmoscopic and biomicroscopic signs of DUSN, and a careful search for the nematode with a fundus noncontact aspheric lens (78-diopter lens), is important to prevent severe visual loss. Even in advanced cases, in the experience of 1 of us (E.C.S.), some improvement in vision and visual field may occur after laser treatment of the worm. Our patient demonstrates that DUSN can occasionally affect the fellow eye. A more appropriate term for this ocular condition might be diffuse subacute neuroretinitis.

Accepted for publication June 30, 1999.

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REFERENCES