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SHORT COMMUNICATION

Case report

Diffuse unilateral subacute neuroretinitis in Europe

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ABSTRACT: Background. *Diffuse unilateral subacute neuroretinitis is thought to be caused by a solitary helminth migrating within the subretinal space. Laser photocoagulation of the located worm is the preferred mode of therapy.*
Methods. *We describe the clinical and electrophysiological features of a case of a 15-year-old Caucasian male with a longstanding diffuse unilateral subacute neuroretinitis (DUSN), in whom two worms were seen in the fundus examination. Focal photocoagulation of the worms was applied as treatment.*
Results. *No signs of inflammation could be seen after treatment. Three months later, the patient was doing well. Follow-up examination 4 years later revealed an unchanged fundus appearance in the affected eye, with no evidence of progression of the syndrome.*
Conclusions. *If a worm is identified in DUSN, focal laser treatment of the located area is the treatment of choice, regardless of whether fundus changes suggest late stages of the disease. However, the eye of the patient should be thoroughly examined to rule out the presence of more than one worm that might cause the failure of therapy. (Eur J Ophthalmol 1999; 9: 58-62)*

KEY WORDS Helminth, Neuroretinitis, Retina, Worm

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INTRODUCTION

Diffuse unilateral subacute neuroretinitis (DUSN) was first described by Gass et al in 1978 (1). This ocular disease occurs in otherwise healthy patients and is characterized in its early stages by visual loss, vitreitis, papillitis and crops of multiple evanescent, gray-white outer retinal lesions. In the late stages of the disease, optic atrophy, retinal arterial narrowing, diffuse pigment epithelial degeneration and an abnormal electroretinogram (ERG) can be seen (1, 2). Although the cause of this condition remains uncertain, several species of helminths have been implicated as potential etiologic agents in DUSN, including *Toxocara canis, Baylisascaris procyonis, Ancylostoma caninum,* and *Alaria mesocercaria* (1-7). To date, evidence has suggested that DUSN is caused by a solitary helminth migrating within the subretinal space. At the present time, photocoagulation of the worm is the preferred mode of therapy using an argon laser.

This report represents an atypical case of DUSN in which two worms were found in the fundus examination, and in which laser photocoagulation had to be applied to the identified worms to prevent the syndrome from developing. Furthermore, DUSN is very rare in Europe, which makes this report an exceptional one.

Case Report

A 15-year-old Caucasian male was evaluated in the Department of Ophthalmology of the “La Fe” University Hospital in Valencia (Spain) with a one-month his-

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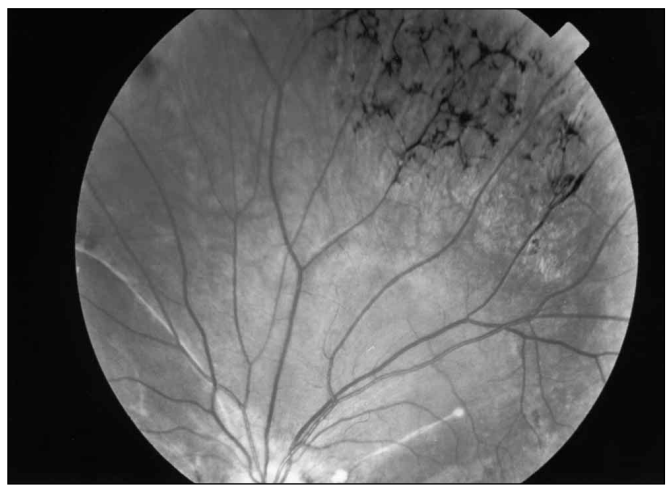
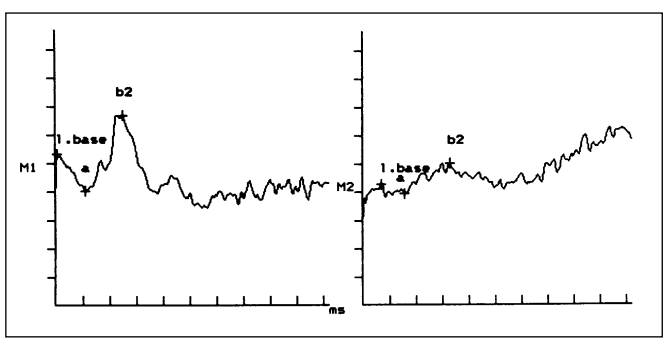


Fig. 1 - ***Left****, Composite photographs of the fundus at initial exploration. Multiple tracks and marked diffuse subretinal change in pigmentation are evident. Two worms are seen below the inferior temporal vascular arcade (arrows). (See Fig. 3)* ***Right****, Detail of retinitis pigmentosa-like changes in RPE.*



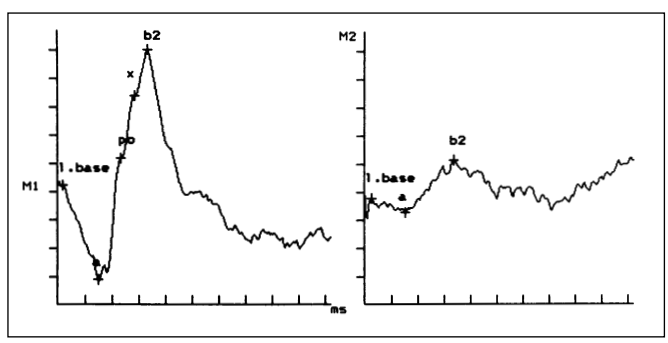


Fig. 2 - *Electroretinogram showing a marked reduction in the photopic and scotopic a- and b- wave amplitudes. (****Left****: white flash;* ***Right****: blue flash).*

tory of painless visual loss in his left eye. Other than exposure to dogs, cats and farm animals, his previous medical history was unremarkable; there was no history of cutanea larva migrans, and the patient had not travelled outside of Europe. Best corrected visual acuity was 20/20 OD and 10/20 OS. Pupillary reflexes were normal in both eyes. The biomicroscopic examination showed quiet anterior segments with clear lenses in both eyes. Intraocular pressure measured by aplanation tonometry was within normal limits (14 mmHg OD, 15 mmHg OS).

Color vision testing, determined by the Farnsworth-Munsell 100-hue test, disclosed defective blue-yellow discrimination in the left eye. Visual field examination showed a relative central scotoma and concentric reduction in the left eye. Visual field testing of the right eye was normal.

The vitreous was clear bilaterally. Fundus exami-

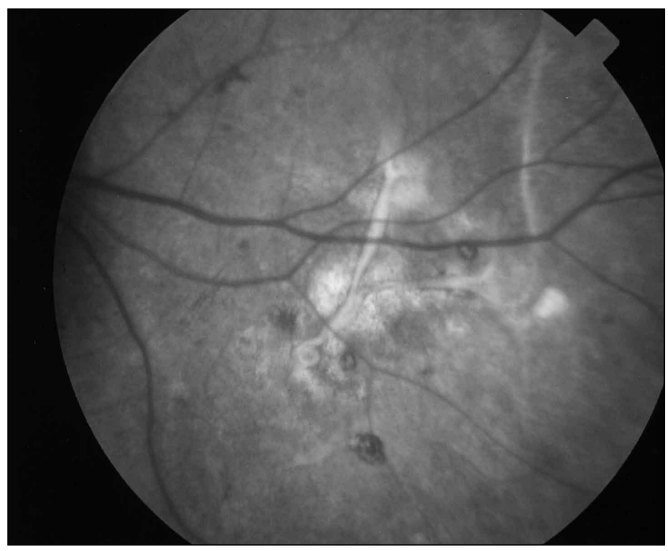


Fig. 3 - *Amplified image showing two worms surrounded by laser photocoagulation scars.*

nation revealed mild optic disk pallor and diffuse degeneration of the retinal pigment epithelium (RPE), that was most marked in the superior peripheral fundus simulating retinitis pigmentosa (Fig. 1, right). Several variably sized hypopigmented tracks were observed in the nasal and inferior retina. Inferotemporal to the macula there were two worms measuring approximately 2000 µm (Fig. 1, left). The inferonasal aspect of these worms was coiled. The fluorescein angiogram disclosed scattered areas of irregular hyperfluorescence related to the pigmentary changes seen in the fundus examination.

The ERG (blue and white flash under scotopic and photopic conditions and 30-Hz. flicker) in the involved eye showed subnormal a- and b-waves, with a larger defect in the latter, compared with the normal (i.e., right) eye (Fig. 2, left and right).

The patient presented a normal haematologic profile, with no evidence of eosinophilia. Stool examination for ova and parasites was unrevealing. ELISA testing for Toxocara was negative.

Upon questioning, the patient revealed that no other family members were known to have retinal disease. We considered the entity to be DUSN with fundus changes suggesting long-standing pathology, and decided to treat the patient with intense focal retinal photocoagulation using an argon green laser in the zone where the worms were located (Fig. 3). Drug treatment was not employed. Three months later, the patient was doing well, no signs of inflammation could be seen, and the lesions corresponding to photocoagulation treatment appeared to be hyperpigmented patched areas in the fundus examination, with inactivity of the worms. Follow-up examination 4 years later revealed an unchanged visual acuity of 10/20 in the affected eye, with no evidence of progression of the syndrome.

DISCUSSION

Several cases of DUSN have been reported in the literature since Gass and Scelfo (1) in 1978 first described this peculiar syndrome, consisting of loss of central vision, vitreous inflammation, papillitis and infiltrates affecting the outer retina and RPE in its early stages, followed by progressive loss of visual field, optic atrophy, constriction of arterial vessels, diffuse atrophic RPE changes and ERG abnormalities - the b-wave being more affected than the a-wave (1,2). They suggested that toxic damage to the retina and optic nerve caused by the migration of a single worm in the subretinal space over a long period of time may be the primary cause of this syndrome (1,2). Although the concept of DUSN has been linked to infestation by a solitary intraretinal parasite, in this report we describe a case caused by at least two worms.

The specific organisms responsible for DUSN have been a subject of controversy, and a great variety of different helminths have been implicated (1-7).

The fact that patients usually do not have clinical or laboratory evidence of systemic parasitic infestation at the time of the ocular symptoms causes serologic testing to be of limited use in the precise identification of the etiologic agent.

In most cases of DUSN the nematodes have been described mainly in terms of their size.

Based on this criterion, Gass et al suggested that two different categories of nematodes could be implicated in this disease, and that they are related to endemic areas (2). Thus, the smaller nematodes group (400-1000 µm), which includes *Toxocara canis* and *Ancylostoma caninum*, was more frequently found in patients from the southeastern United States; in comparison, the larger nematodes group (1500-2000 µm), which includes *Baylisascaris procyonis*, was typically seen in patients from the northern midwest region of the United States (1-6).

In our case, the patient had not traveled outside of Europe prior to the examination. This is all the more surprising if we take into account that only a few cases of DUSN have been reported in Europe (8). We used fundus photographs to compare the length of the worms with the diameter of the optic nerve head, assuming this to be 1500 µm. The worms measured about 2000 µm. Thus, although we are not able to definitely identify the variety of parasite affecting our patient, we consider it to belong to the larger nematodes group.

Regarding the etiopathogenic mechanism of this syndrome, it is believed that larval excretory / secretory products, including various enzymes and metabolic waste products could have a local direct toxic effect on the retina and stimulate an inflammatory response principally mediated by eosinophils (2, 9, 10). In our patient, manifestations due to inflammatory response

were limited (no Tyndall effect, no vitreitis...), and visual acuity was surprisingly well-preserved, despite the presence of more than one worm. Funduscopic examination revealed multiple zones of atrophy of the pigment epithelium with migration of pigment into the overlying retina that is obviously of many months' and perhaps years' duration. In spite of this the patient had not previously complained of visual loss. For this reason we agree with those authors that suggest that the inflammatory response to helminth invasion depends on the patient's immune response (1, 6, 9, 10). If this response is not severe, the number of worms will play a minor role in visual impairment in DUSN. Despite this discrete inflammatory response in our case, ERG disclosed subnormal a- and b-wave amplitudes, indicating an alteration in the outer retina. This explains the blue-yellow discrimination defect seen in our patient with the Farnsworth Munsell test, according to Kollner's rule (11) (lesions in the outer retinal layers give rise to blue-yellow defects, whereas damage to the inner retinal layers and optic nerve produces a red-green defect).

Concerning treatment of intraocular helminth larvae, photocoagulation has been demonstrated to be highly effective in destroying the worms, with minimal exacerbation of the inflammatory reaction after successful laser treatment (1, 2, 5, 6, 12, 13). This is probably due to heat-destruction of worm antigens, as has been reported by other authors with other intraocular parasites (14).

We have been successful in treating the areas where the worm was located.

Although it was not necessary in our patient, in those cases in which biomicroscopic identification and localization of the worms is not possible, oral therapy with thiabendazole may be beneficial - particularly among patients with moderately severe vitreous inflammation (15). Observation of a new white retinal lesion 4 to 7 days after medical treatment may indicate death of the helminth.

We conclude that this is an atypical case of DUSN in which more than one worm was found in the fundus examination. Ophthalmologists should be aware of this possibility, and patients should be thoroughly examined to avoid the failure of treatment and progression of the syndrome if one of these worms is not detected.

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