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Diffuse Unilateral Subacute Neuroretinitis with Typical Serpiginous Retinal Pigment Epithelial Atrophy

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Abstract

We report a case of diffuse unilateral subacute neuroretinitis (DUSN) with atrophy of the serpiginous retinal pigment epithelium (RPE) which has not yet been reported in the Korean medical literature. A 41-year-old man complained a progressive visual loss and ocular pain in the left eye. The patient has a history of exposure to live doe's bloods. On the slit lamp microscopy, a mild severity of inflammatory cells was observed in the vitreous cavity. Ophthalmoscopy showed the cluster of gray-white RPE atrophy, many tortuous serpiginous tract with sparing of the macular region in the left eye.

Although the location of the nematode could not be identified, the appearance of serpiginous tract atrophy on subRPE and a history of exposure to live animal supported the diagnosis of DUSN.

Key Words : Diffuse unilateral subacute neruoretinitis, subretinal nematode, typical serpiginous retinal pigment epithelial atrophy

Introduction

Diffuse unilateral subacute neuroretinitis (DUSN) is an infectious disease affecting the outer retina and retinal pigment epithelium (RPE) caused by a nematode worm that may wander in the subretinal space. Since this disease was first reported by Gass and Scelfo [1] in 1978, these authors [2] noted that the etiology of this disease is a motile subretinal nematode. According to Gass and Braunstein [3], two different sizes of nematodes (400-1000 ㎛ and 1500-2000 ㎛) were found in the disease. It has been thought that the small worm is Ancylostoma caninum and a larval form of Toxocara canis, the large worm is due to Baylisascaris procyonis [3-5]. The places where this disease has developed include the

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United States, Venezuela, Brazil, India and China, but this disease has not yet been reported in Korea [3,6-9]. However, a subretinal nematode can be identified inapproximately one third of the cases in many reports [2,10,11]. We describe a case of DUSN with the late serpiginous pigment epithelial degenerative changes except for failure to detect the subretinal nematode.

Case

A 41-year-old man visited our ophthalmology department presenting blurred vision, ocular pain and conjunctival injection in his left eye. The patient had an experience of drinking doe's blood three years ago.

The visual acuity was 20/20 in the right eye and 20/100 in the left eye. On slit lamp microscopy, the left eye had 3+ inflammatory cells in the anterior chamber and mild inflammatory cells in the vitreous cavity. On fundoscopy, there was typical, tortuous RPE atrophy that spared the macular region, RPE hypertrophy around the RPE atrophy in the overall area of the retina in the left eye, and mild severity of the narrowing of the retinal vessel (Fig. 1A). Even on performing fluorescein angiography, there was tortuous hyper-fluorescence due to a window defect that corresponded to the RPE atropy, yet there were no findings suggestive of leaking (Fig. 1B-D). DUSN was clinically suspected. This led to performing meticulous fundoscopy in the overall area of the retina, but any nematode was not detected. Except for a high absolute neutrophil count on the blood tests, the chest PA, ESR, CRP, antinuclear antibody, toxoplasma antibody were negative.

To control the inflammation that developed in the anterior chamber and vitreous body, oral prednisolone therapy were initiated from level of 40 mg. Thereafter, the inflammatory reaction in the anterior chamber and vitreous body was decreased. On fundoscopy, however, the tourtuous lesions did not change. A repeated fundoscopy also couldn't identify the location of a subretinal nematode. Four months following a monitoring of clinical course, the atrophic lesions in the retina did not change, but the inflammatory findings that were previously present in the anterior chamber and vitreous body disappeared. The patient's visual acuity improved to 20/20.

Discussion

DUSN was first described by Gass and Scelfo [1], and these authors noted that it is typically characterized by the following findings: (1) an insidious, usually severe loss of peripheral and central vision; (2) vitritis; (3) diffuse and focal pigmented epithelial derangement with relative sparing of the macula; (4) narrowing of the retinal vessels; (5) optic atrophy; (6) an increased retinal circulation time and (7) subnormal electro-retinographic findings.

Later, Gass *et al.* [2] reported that a motile subretinal nematode was detected while monitoring the clinical course during the early stage in two patients among twenty-five DUSN patients. Thus, these authors added (8) subretinal nematode to the diagnostic criteria for DUSN.

It typically affects healthy children or young adult. The mean age of onset is 16.7 years (range: 5 to 22 years) according to a

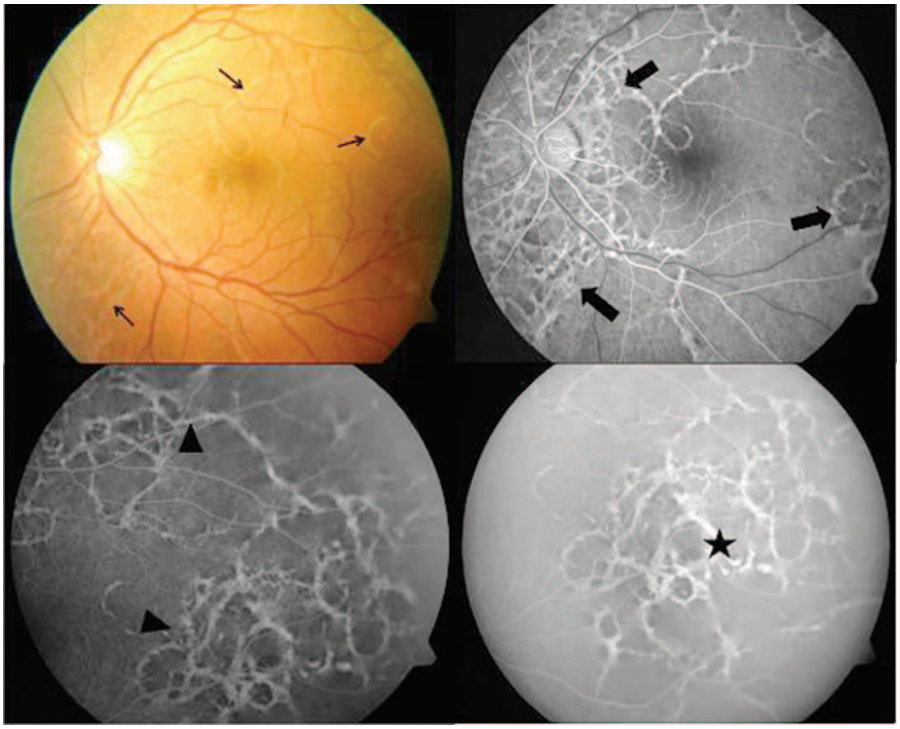

Fig. 1. (A) Fundus photograph of the left eye. Note diffuse linear retinal pigment epithelium changes, and the gray-white outer retinal lesions (small arrows). (B) Early phase fluorescein angiography shows diffuse linear window defect secondary to diffuse retinal pigment epithelium atrophy (large arrows). (C and D) Diffuse serpiginous linear retinal pigment epithelial atrophy without leakage was seen in peripheral on late phase fluorescein angiography (arrow heads, star).

recent report by Cortez *et al.* [12]. In more than 83% of the patients, DUSN occurs before 20 years of age. In this case, the age of patient (41 years) was much higher than the general age of onset that's been commonly reported for patients with DUSN. Serologic studies, stool examinations and peripheral blood test have little value in making the diagnosis of DSUN, however, we need to run a few test to discriminate DSUN from many other infectious and inflammatory conditions.

During its early stages, symptoms include mild to moderate vitritis, mild optic disc swelling, recurrent evanescent multifocal gray-white lesion at the level of the outer retina. Failure to recognize and to destroy the subretinal nematode with medical therapy or photocoagulation for a long period consequences in progressive optic atrophy, narrowing of the retinal arteries, diffuse degeneration in outer retina and RPE, ill-defined areas of pigment epithelial depigmentation.

A summary of the recent reports has confirmed that subretinal nematode is detected in approximately 25-40% of the total cases of DUSN. According to Anshu and Chee [13], even for the cases in which a subretinal nematode could not be detected, a diagnosis of DUSN can be made if the criteria of Gass and Scelfo [1,2] were satisfied. Venkatesh *et al.* [11] reported on two cases

for which laser photocoagulation could not be performed, although the nematode was identified at the initial diagnosis because the nematode migrated as time went by. Based on these two cases, they maintained that the motility of the nematode is the cause of not detecting a nematode on a fundus examination.

From the same perspective, a subretinal nematode could not be identified in our current case on several fundoscopy exams, although the typical tortuous findings suggestive of track-like serpiginous retinal pigmented epithelial atrophy were present. Such findings as a lack of optic atrophy, no papillitis, mild severity of visual disturbance and its subsequent recovery were inconsistent with the previous reports. Presumably, these findings might be due to the difference of the disease course in DUSN patients who come from such areas as a prairie region, a seaside area or Venezuela, whose climate and environment are different from those of Korea.

This case highlights the need for a high suspicion in making the diagnosis of DSUN, a history of exposure to live doe's bloods, the appearance of subRPE serpiginous tract and peripheral RPE hypopigmentation due to mechanical disruption of motile worm and variable inflammation due to toxic reaction even the nematode was not detected subretinal space.

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