

Case Report

Multimodal Imaging in a Case of Idiopathic Neuroretinitis

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Keywords

Neuroretinitis · Multimodal imaging · Choroidal involvement

Abstract

A 37-year-old woman presented with painful visual loss in the left eye for 2 weeks. The best-corrected visual acuity was 20/200. Ophthalmic examination of the left eye revealed vitreous cells, optic disc swelling, serous retinal detachment, and macular star-figure hard exudates. Swept-source optical coherence tomography showed both inner and outer retinas were swollen, the choroid was thickened, and the corresponding retinal pigment epithelium was elevated. Wide-field indocyanine green angiography disclosed multiple hypofluorescent spots in the mid-periphery. Taken together, an involvement of optic disc, entire retina, and choroid was considered in the current case.

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Introduction

Neuroretinitis is characterized by acute visual loss accompanied by optic disc swelling, serous retinal detachment (sRD), and hard exudates arranged in a star figure around the fovea [1, 2]. The etiology is thought to be infection or an immune-mediated process that may be precipitated by different agents [3]. Most patients achieve excellent visual recovery with or without treatment [1, 2]. Gass [4] speculated that the optic disc swelling preceded macular exudates and abnormal permeability from the deep capillaries within the optic disc resulted

in sRD, suggesting the term “neuroretinitis.” Here, we report multimodal imaging in a case of idiopathic neuroretinitis and consequent speculation about the pathophysiology in this disease.

Case Report

A 37-year-old woman presented with painful unilateral visual loss for 2 weeks in the left eye. The patient had fever accompanied by ocular pain several days prior to the visual onset. Any other neurological disorders were not present. The best-corrected visual acuity of the right eye and the left eye was 20/20 and 20/200, respectively. There was a relative afferent pupillary defect in the left eye. Slit-lamp examination revealed a clear anterior segment with occasional cells in the vitreous cavity of the left eye. A fundus picture taken by a fundus camera (TRC-50DX, Topcon, Tokyo, Japan) showed sRD and hard exudates arranged in a star figure around the fovea in the left eye (Fig. 1). Swept-source optical coherence tomography (DRI OCT Triton, Topcon, Tokyo, Japan) showed subretinal fluid between the optic disc and the macula (Fig. 1). Both inner and outer retinas were swollen and hyperreflective foci were detected from the inner nuclear layer through the outer nuclear layer (Fig. 1). Moreover, the choroid was thickened, accompanied by blurry visible choroidal lumina, and the corresponding retinal pigment epithelium (RPE) line was elevated (Fig. 1). Fluorescein angiograms (Heidelberg Retina Angiograph 2, Heidelberg Engineering, Heidelberg, Germany) showed hyperfluorescence at the disc and hypofluorescence corresponding to the area of sRD (Fig. 1). Wide-field indocyanine green angiograms (WFICGA) (Optos California, Optos plc, Dunfermline, UK) showed multiple hypofluorescent spots in both the posterior pole and the mid-periphery of the left eye (Fig. 1). Visual field examination (Humphrey Field Analyzer, Carl Zeiss Meditec, Jena, Germany) of the left eye demonstrated a dense centrocecal scotoma (Fig. 1). Any abnormal findings on head magnetic resonance imaging were not present. All blood tests including various antibody titers against cytomegalovirus, herpes simplex virus, varicella zoster virus, toxoplasma, toxocariasis, and *Bartonella henselae* gave normal results. The patient did not have a history of sustaining a scratch or bite from a cat or dog. The patient was diagnosed with idiopathic neuroretinitis. Azithromycin dehydrate (500 mg/day) was administered orally for 3 days. Since the ocular symptoms and findings gradually resolved, any other treatments including steroid were not received. Five months after the initial visit, ocular findings had almost resolved (Fig. 2). The best-corrected visual acuity recovered to 20/20, although the RPE in the left eye was atrophic (Fig. 2). The hypofluorescent spots on WFICGA were still present at the convalescent stage (Fig. 2).

Discussion

Here, we report multimodal images of not only the posterior pole but also the wide-field area in a case of idiopathic neuroretinitis. There was leakage from the optic disc on fluorescein angiogram, and consequently both inner and outer retinas also became swollen, hyperreflective foci were located from the inner nuclear layer through the outer nuclear layer, sRD was observed from the optic disc to the macula, and the choroidal was thickened, accompanied by an elevation of the RPE line. Furthermore, multiple hypofluorescent dark spots on WFICGA were detected in the mid-periphery as well as in the posterior pole. Taken together, the condition in the current case was thought to be neurochorioretinitis. A previous report showed

choroidal involvement in eyes with neuroretinitis [5], although only posterior pole lesions were described, which supports the current findings.

Neuroretinitis is characterized by acute visual loss with disc swelling and macular star formation [1, 2]. Gass [4] speculated that the optic disc swelling preceded macular exudates. In the current case, ocular pain preceded visual loss, and both retinal edema and sRD were observed continuously from the optic disc, which are thought to be consistent with Gass's speculation. Gass [4] also described that abnormal permeability from the retinal deep capillaries within the optic disc resulted in sRD and penetration into the outer plexiform layer. In the current case, both inner and outer retinas were swollen and the choroid was thickened, accompanied by blurry visible choroidal lumina. We speculate that lymphocytes or other inflammatory cells recruited at the optic disc might diffusely infiltrate into the retina and the choroid. WFICGA disclosed multiple hypofluorescent dark spots in the mid-periphery of the left eye, which are usually observed in eyes with diffuse choroiditis such as Vogt-Koyanagi-Harada disease [6]. Taken all together, the current case appeared to have an involvement of optic disc, entire retina, and choroid. Therefore, this condition in the current case should be called neurochorioretinitis.

The visual prognosis of patients with neuroretinitis is generally excellent as the disease is self-limiting, but some cases without visual improvement because of optic disc atrophy were reported [1]. Treatment for neuroretinitis depends on whether there is an underlying infectious or inflammatory condition that requires therapy. Patients with idiopathic neuroretinitis usually do not need any treatments, although antibiotics, steroids, or both have been used in some cases [1, 3, 7, 8]. Dreyer et al. [1] reported that steroid therapy did not hasten the recovery or attain superiority in the visual prognosis compared to no treatments. In the current case, we treated with oral azithromycin dehydrate but only for 3 days because the visual acuity gradually improved immediately after the administration. Therefore, it was unclear whether the antibiotics were effective or not in the current case.

In conclusion, we report multimodal imaging in a case of idiopathic neuroretinitis, which indicates an involvement of optic disc, entire retina, and choroid. Future studies with a large sample size are needed for further understanding of the pathogenesis of idiopathic neuroretinitis.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

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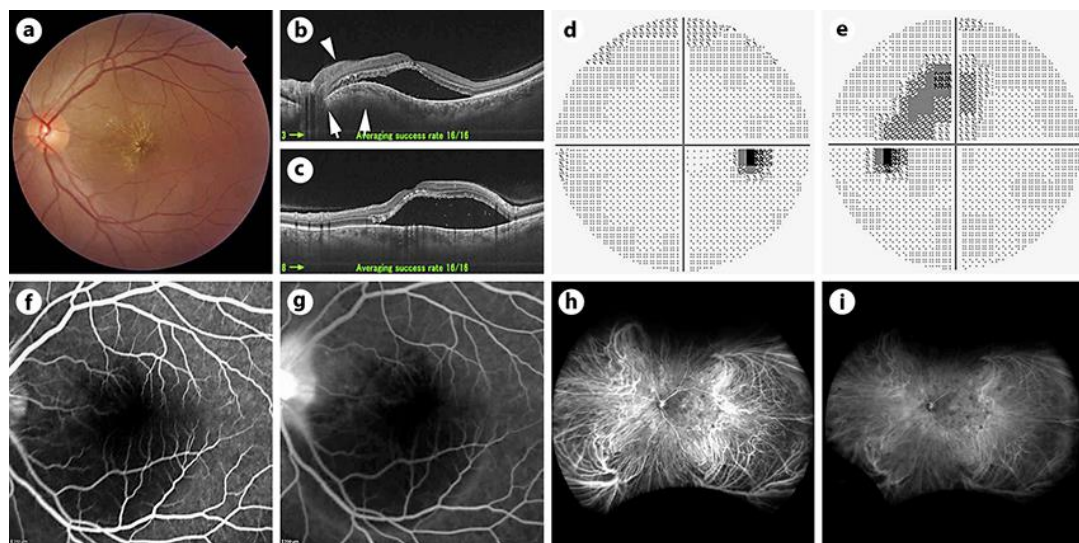


Fig. 1. Findings at the acute stage. **a** Fundus photography findings. Serous retinal detachment (sRD) at peripapilla and macular star-shaped hard exudates were observed in the left eye. **b, c** Optical coherence tomography findings. **b** Horizontal scan (5-line cross, 12 mm). **c** Vertical scan (5-line cross, 12 mm). Both inner and outer retinas were edematous (arrowhead) and hyperreflective foci were observed from the inner nuclear layer through the outer nuclear layer. The choroid was thickened (the central subchoroidal thickness was 425 μm), accompanied by blurry visible choroidal lumina (arrows), and the corresponding retinal pigment epithelium line was elevated. **d, e** Visual field findings measured by the C30-2 program of the Humphrey Field Analyzer. **d** Right eye. **e** Left eye. The visual field of the right eye was normal. A dense centrocecal scotoma was detected in the left eye. **f, g** Fluorescein angiography findings. **f** Early phase. **g** Late phase. Hypofluorescence (block) corresponding to the area of sRD and hyperfluorescence (leakage) at the optic disc were observed. **h, i** Wide-field indocyanine green angiography findings. **h** Early phase. **i** Late phase. Multiple hypofluorescent spots were observed in the mid-periphery.

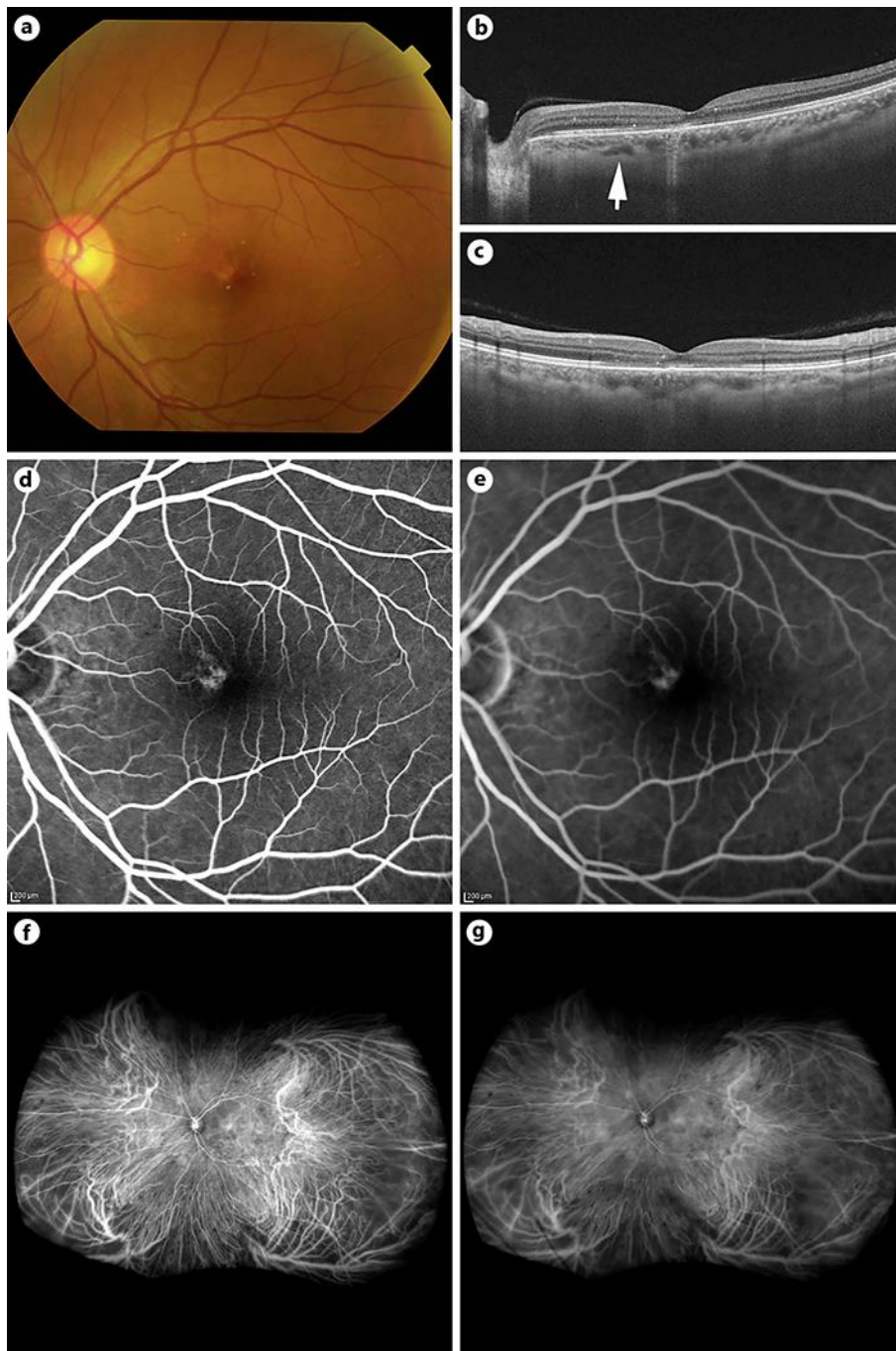


Fig. 2. Findings at the convalescent stage (5 months after disease onset). **a** Fundus photography findings. Serous retinal detachment (sRD) at peripapilla resolved, small hard exudates remained, and the retinal pigment epithelium (RPE) was atrophic. **b, c** Optical coherence tomography findings. **b** Horizontal scan (5-line cross, 12 mm). **c** Vertical scan (5-line cross, 12 mm). sRD completely resolved. The central subchoroidal thickness decreased to 247 μm . An artifact due to the RPE atrophy was observed. The choroidal lumina (**b**) were better observed compared to those at baseline (Fig. 1b). **d, e** Fluorescein angiography findings. **d** Early phase. **e** Late phase. Leakage from the optic disc disappeared. Hyperfluorescence indicating “window defect” due to RPE atrophy was observed at the macula. **f, g** Wide-field indocyanine green angiography findings. **f** Early phase. **g** Late phase. Multiple hypofluorescent spots were still observed in the mid-periphery.