CASE REPORT

Bilateral Neuroretinitis in Cat Scratch Disease with Exudative, Obliterative Vasculitis in the Optic Disc

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ABSTRACT

A 29-year-old fisherman exhibited optic disc oedema and peripapillary retinal detachment in the right eye, whereas in the left eye, optic atrophy and intraretinal exudates were already observed on first examination. About 6 months earlier, he noticed blurred vision of the left eye but took no medication. Visual acuity was 0.4 OD and 0.01 OS. Perimetry showed a large lower-half field defect with sparing 10° central field in the right eye and a large central scotoma in the left eye. Fluorescein angiography showed existence of arteriole or capillary nonperfusion and hyperpermeability of surrounding capillaries. Since serological examinations showed positive *Bartonella* immunoglobulin G (IgG) and other causes of neuroretinitis (NR) were excluded, NR in the present case was caused by cat scratch disease (CSD). Optic atrophy appeared 2 weeks after onset. Optical coherence tomography 13 weeks after onset revealed severe loss of retinal nerve fibre layer (RNFL) superior and nasal to the optic disc in both eyes and temporal in the left eye. Visual acuity of the right eye improved to 1.2 by the treatment, whereas visual field defects were persistent. CSD-NR in the present case developed abrupt appearance of optic atrophy with severe RNFL loss in the right eye, which was elicited by exudative, obliterative vasculitis in the superficial layer of the optic disc.

Keywords: cat scratch disease, neuroretinitis, optical coherence tomography, retinal nerve fibre layer

INTRODUCTION

Neuroretinitis (NR) is characterised by optic disc oedema, serous retinal detachment, and macular star, which is attributed to not only idiopathic but also infectious condition.¹ In contrast to severity of fundus features and visual symptoms, this disease has a good visual prognosis. On the other hand, some NR patients show little recovery from visual dysfunctions.^{1,2} However, the pathomechanism in NR that develops irreversible impairment within the optic nerve or the retina remains unsolved.

We report a patient with bilateral NR in cat scratch disease (CSD-NR) who exhibited persistent visual dysfunctions in both eyes. Present fluorescein angiography showed inflammatory optic neuropathy with exudative, obliterative vasculitis on the superficial layer of the optic disc, and optical coherence

tomography (OCT) revealed direct evidence of severe and abrupt loss of the retinal ganglion cell axons in both eyes, which accounted for poor visual recovery.

CASE REPORT

A 29-year-old fisherman was referred to our hospital for treatment for a 3-day history of blurred vision in the right eye. About 6 months earlier, he noticed severe visual loss in the left eye but did not treat this eye, as he had no ocular pain and systemic symptoms. On first examination, corrected visual acuity was 0.4 OD and 0.01 OS. Direct light pupillary reflex was weak in the right eye but more sluggish in the left eye. Slit-lamp and ophthalmoscopic examinations showed swollen optic disc with haemorrhages on its surface

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FIGURE 1 Ophthalmoscopic and fluorescein angiographic findings on first examination (A–F) and 2 weeks later (G). (A) The right eye shows optic disc oedema including white to waxy patch and spots (arrows). (B) FA of the right eye revealed hypofluorescent areas (arrows) corresponding to white patch and spots shown in A during the early arteriovenous phase. (C) During the late arteriovenous phase, the hypofluorescent areas were stained by dye leaking from surrounding capillaries. Black parts were due to haemorrhages. (D) Eleven minutes after injection, maximum leakage from the optic disc and peripapillary serous retinal detachment were noted. On the other hand, the present FA findings showed that the intrachoroidal circulation had no filling delay or defects. (E) The left eye showed optic atrophy and retinal lipid-rich exudates. (F) FA of the left eye showed vertical hypofluorescent area in the optic disc during the arteriovenous phase. (G) Two weeks later, ophthalmoscopic findings of the right eye showed a typical macular star configuration with optic atrophy.

and posterior vitreous cells in the right eye (Figure 1A). The retinal veins were engorged, whereas the retinal arteries were attenuated. Retinal vessels were almost tortuous. It, furthermore, was discernible that white to waxy spots and patch were noted at 1, 3, 7, and 10 o'clock positions of the optic disc surface (shown by arrows in Figure 1A) and indefinitely demarcated exudates were located at the optic disc centre. Fluorescein angiography (FA) revealed massive leakage from the optic disc at fully stainable stage in the right eye (Figure 1C, D). However, during the arteriovenous phase, the white to waxy patch and spots observed ophthalmoscopically (arrows in Figure 1A) showed hypofluorescence (Figure 1B, arrows) but changed to hyperfluorescence at the late arteriovenous phase (Figure 1C). The central portion of the optic disc was also hypofluorescent where the underlying vessels were masked (Figure 1B). FA 11 minutes after the injection revealed massive leakage of dye from the optic disc to peripapillary regions with a diffuse and a dome-like expansion pattern in an inferotemporal and a superonasal direction, respectively (Figure 1D). The arm-to-retina circulation time was 37 seconds. On the other hand, optic atrophy and white-yellowish intraretinal exudates appeared in the left eye (Figure 1E). FA of the left eye showed

hypofluorescent areas in the optic disc during the arteriovenous phase (Figure 1F). OCT detected fluid retention in the outer nuclear-plexiform space and retinal detachment in the right eye, which were almost identical to those in Leber idiopathic stellate neuroretinitis.³ Goldmann's perimetry showed a large lower visual field defect with sparing 10° central field in addition to concentric contraction of upper field in the right eye and a large lower visual field defect with central scotoma in the left eye; however, this large visual field defect in both eyes was different from altitudinal hemianopsia. He was immediately administered with 1000 mg methylprednisolone intravenously daily for 3 days, which was thereafter changed to oral prednisolone of 30 mg, persisting for three successive days. This treatment was carried out three times. After completion of the third treatment, oral prednisolone was gradually tapered. Two weeks after onset, lipid-rich exudates were accumulated to make macular star and the optic disc was pale in colour with resolution of oedema (Figure 1G). Serological examinations on admission revealed that *Bartonella* immunoglobulin G (IgG) was 1:256, but those for collagen diseases, autoimmune diseases, and other infections were negative. The patient did not sustain animal injury previously. As CSD caused this bilateral

FIGURE 2 OCT findings. (A) OCT of the right eye detected loss of RNFL nasal to the optic disc (arrow) 8 weeks after the onset. (B) Cross-sectional images in temporal-nasal direction through the centre of the optic disc by OCT, 13 weeks after the onset. (C) Cross-sectional images in inferior-superior direction. Left column shows the right eye and right column is the left eye. T=the retinal image temporal to the optic disc; N=nasal; I=inferior; S=superior in B and C. An arrow notes RNFL loss.

NR, he was started with oral administration of 200 mg tetracycline twice daily. Ten days after administration of antibiotic was begun, corrected visual acuity improved to 1.2 OD and 0.1 OS, whereas visual field defects were unchanged in both eyes. Eight weeks after onset, OCT (Cirrus HD-OCT; Carl Zeiss) after pupillary dilation revealed a discernible loss of retinal nerve fibre layer (RNFL) nasal to the optic disc (arrow), but temporal RNFL was preserved in the right eye (Figure 2A).

Goldmann's perimetry and OCT were reexamined 13 weeks after onset. The perimetry showed little alteration of visual field defects in both eyes, compared with those detected on admission. OCT scan was carried out 6 mm in temporal-nasal direction (Figure 2B) and in inferior-superior direction (Figure 2C) through the centre of the optic disc. Present OCT revealed that RNFL was obviously detected at the cross-sectional image temporal (T) to the optic disc. However, the thickness of RNFL 2 mm apart from the optic disc centre was 29 μm ; this value was small, compared with that of three age- and refraction error-matched normal subjects ($n=6$, mean = 57.5 μm). On the other hand, thin RNFL was noted inferior (I) but completely extinguished nasal (N, arrow) and superior (S, arrow) to the disc in the right eye (Figure 2B, C, left). In the left eye, RNFL completely disappeared at the images temporal (T, arrow), nasal (N, arrow), and superior (S, arrow), whereas faint RNFL was observed at the image inferior (I) (Figure 2B, C, right).

DISCUSSION

CSD causes NR characterised by optic disc oedema, serous retinal detachment, and macular star, which has generally a good visual prognosis. However, the present patient exhibited CSD-NR features different from those in the previous reports^{4,5} in fundus appearance and clinical course. Firstly, 2 weeks after onset, optic atrophy started to appear, with resolution of optic disc oedema and occurrence of macular star. Secondly, large lower visual field defect with a contraction of upper field observed on first examination was persistent during a 2-year follow-up period. FA of the right eye revealed vascular abnormalities causative of persistent visual dysfunctions. White to waxy patch and spots seen ophthalmoscopically were hypofluorescent during the early arteriovenous phase but stainable by dye leaking from surrounding capillaries during the late arteriovenous phase, indicating sporadic existence of arteriole or capillary nonperfusion areas and hyperpermeability of the surrounding capillaries with lipid-rich exudates (Figure 1). In fact, FA findings in the left eye preceding the onset of the right eye already showed wide capillary nonperfusion areas on the optic disc during the arteriovenous phase. It, therefore, is likely that the occlusion of small vessels were elicited by exudative, obliterative vasculitis on the superficial layer of the optic disc. *Bartonella* organisms tend to have contact with vascular endothelial cells to aggregate and colonise, consequently producing a necrotising or a vasoproliferative response.^{6,7} Clinically, *Bartonella* infection is associated with ocular vascular abnormalities, i.e. peripapillary angiomas,⁸ retinal and optic nerve head neovascularization,⁹ and retinal artery occlusion.¹⁰

The arm-to-retina circulation time in the present patient was significantly elongated to 37 seconds, suggesting that the prelaminar artery system had heavy resistance to inflow, probably because of *Bartonella* organism itself or decreased internal diameter of the artery following inflammation of vascular endothelium cells. As a result, the stagnation of blood flow and the obliteration in the optic disc circulation system may develop into an ischaemic state to affect the optic nerve fibres.

Interestingly, OCT performed 13 weeks after onset disclosed complete loss of RNFL at the cross-sectional images superior and nasal to the optic disc in both eyes and temporal in the left eye (Figure 2B, C). Discernible RNFL loss nasal to the optic disc in the right eye was already observed 8 weeks after onset (Figure 2A). Thus, this time of the loss is remarkably short, comparing with that in multiple sclerosis neuritis.¹¹ Severe, abrupt RNFL loss in the present patient may result from ischaemic process associated with optic neuritis.

Since CSD-NR has a good visual prognosis,¹ it is difficult to determine the efficacy of its treatment. In the present patient, however, the untreated left eye already showed optic atrophy with prominent loss of the papillomacular axons on first examination. In contrast, the right visual acuity recovered to 1.2 by treatment of corticosteroid with antibiotic, suggesting this treatment was useful to prevent the papillomacular bundle from severe death. Thus, the present study indicates that management of CSD-NR is required, as the condition develops irreversible RNFL loss in some cases.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figure 1 of this article is available in colour online at www.informahealthcare.com/oph

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