A Case of Refractory Posterior Scleritis with Marked Retinochoroidal Detachment Associated with Panuveitis

Yoshihiro NAKAGAWA, Takahiro SUZUKI, Ami SAHASHI,
Xue TAN and Yasuyuki SUZUKI

Department of Ophthalmology, Tokai University School of Medicine

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An 84-year-old man presented with decreased right-eye visual acuity. Upon initial examination, the right- and left-eye visual acuities were 0.03 and 1.2, respectively; moreover, the right- and left-eye intraocular pressure was 12 mmHg and 13 mmHg, respectively. Examination revealed a shallow anterior chamber of the right eye, anterior chamber inflammation, vitreous opacity, and marked retinochoroidal detachment. Optical coherence tomography (OCT) revealed retinal detachment (RD) and choroidal folds; moreover, B-scan ultrasonography (B-scan) showed RD as well as thickened sclera with fluid in Tenon's space. Fluorescent fundus angiography revealed hyperfluorescence in the optic disc and vascular hyperpermeability in the right eye. The left eye lacked extra-ocular symptoms or abnormalities. The right ocular axis measured 23.4 mm with no apparent subretinal fluid migration due to positional changes. Accordingly, the patient was diagnosed with panuveitis associated with posterior scleritis and immediately started on 40 mg prednisolone, which improved his symptoms. However, at 3 post-treatment months, choroidal folds were observed and was restarted on 20 mg prednisolone. The choroidal folds subsequently disappeared, with a current visual acuity of 0.3 in the right eye and no recurrence. Our findings indicated the utility of accurate diagnosis of posterior scleritis by B-scan and prompt systemic steroid administration.

Key words: posterior scleritis, retinochoroidal detachment, panuveitis, steroid therapy, B-scan ultrasonography

INTRODUCTION

Posterior scleritis is a rare disease primarily involving the posterior sclera that affects surrounding areas in various ways [1, 2]. Inflammation of the adjacent choroid, retina, optic nerve, external ophthalmoplegia, and orbit, as well as the diversity of clinical findings, including subretinal masses, choroidal hilum, and exudative retinal detachment, often impede diagnosis [3]; moreover, prompt treatment is crucial for a favorable prognosis [2]. We encountered a case of refractory posterior scleritis, which was difficult to differentiate from other diseases such as rhegmatogenous retinal detachment, uveal effusion syndrome, and Harada's disease; however, prompt steroid administration improved the panuveitis symptoms. It is noted in the medical record that consent has been obtained from this patient for academic publication in a non-personally identifiable form.

CASE REPORT

An 84-year-old man, who was 165 cm height and weighted 63 kg, had been treated with 0.1% dexamethasone eye drops for iritis of the right eye by his family doctor for ≈6 months. He was referred to our hospital for close examination and treatment due to exacerbation of iritis. His medical and family history was unremarkable. On initial examination, the visual acuity in the right and left eyes was 0.03 and 1.2, respectively; moreover, the intraocular pressure in the right and left eyes was 12 mmHg and 13 mmHg, respectively. The right eye showed mild conjunctival hyperemia, a slightly shallower anterior chamber than the left eye, posterior iris adhesion, and mild cataract (Fig. 1). There are 1+ cells in the anterior chamber. Retinochoroidal

Fig. 1 Anterior segment photography: The anterior chamber in the right eye was slightly shallow with posterior iris adhesions and mild cataracts.

Takahiro SUZUKI, Department of Ophthalmology, Tokai University School of Medicine, 143 Shimokasuya, Isehara, Kanagawa 259-1193, Japan
Tel: +81-463-93-1121 Fax: +81-463-91-9328 E-mail: taka-su@tokai.ac.jp

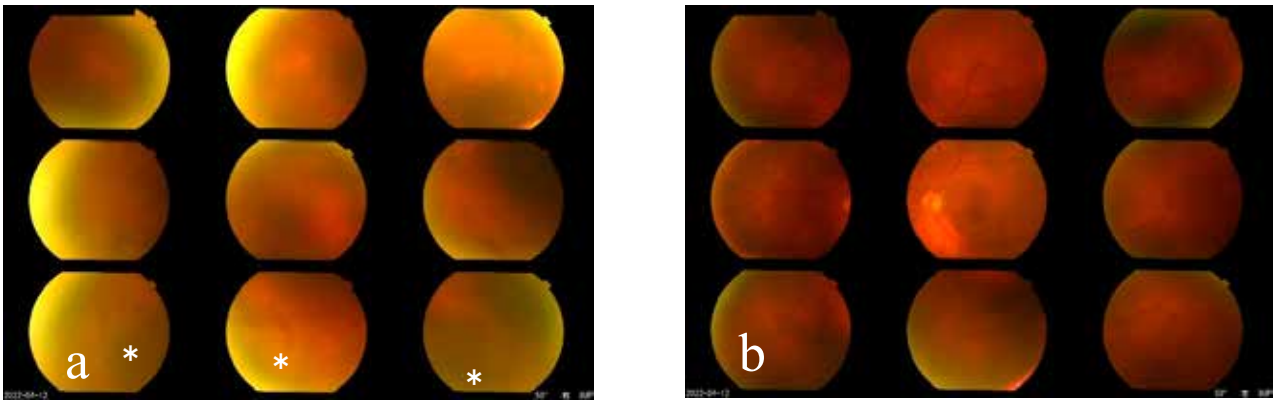

Fig. 2 Fundus photograph at the first visit: Vitreous opacities and inferior retinochoroidal detachment (asterisk) in the right eye (a). The left eye shows no abnormality (b).

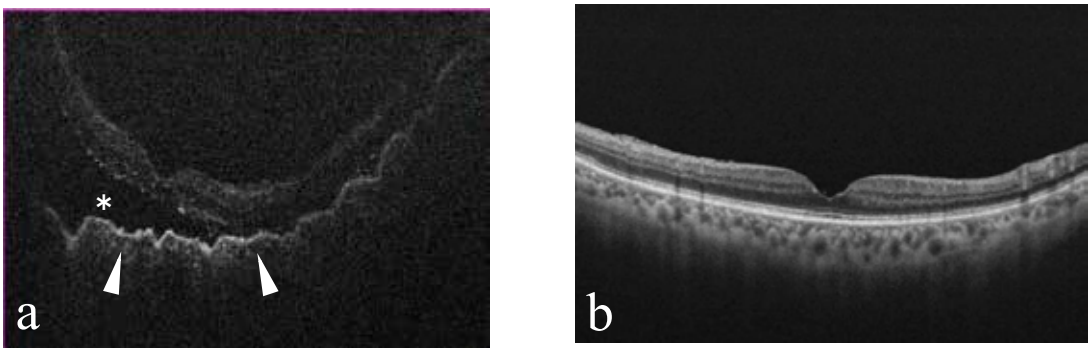

Fig. 3 OCT at the first visit: OCT shows RD (asterisk) and choroidal folds (arrowhead) in the right eye (a). The left eye shows no abnormalities (b).

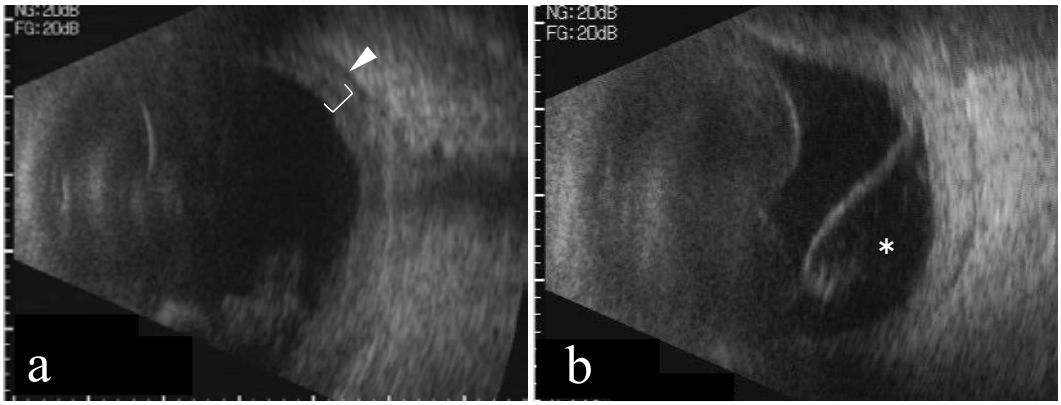

Fig. 4 B-scan in the right eye: B-scan in the horizontal section shows a thickened sclera (white line) with fluid in Tenon's space (arrowhead) (T-sign) (a). B-scan in vertical section shows RD (asterisk) (b).

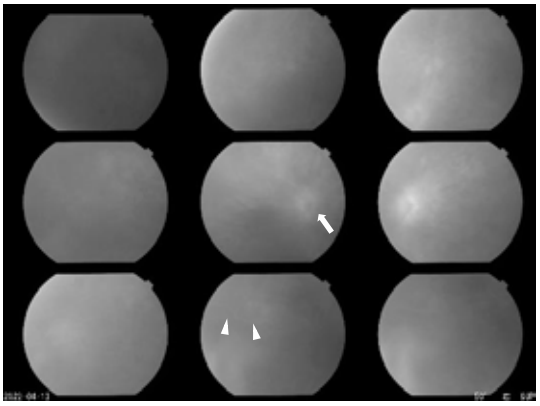

Fig. 5 Fluorescein angiography: Fluorescence leakage from the optic nerve papillae in the right eye (arrow).

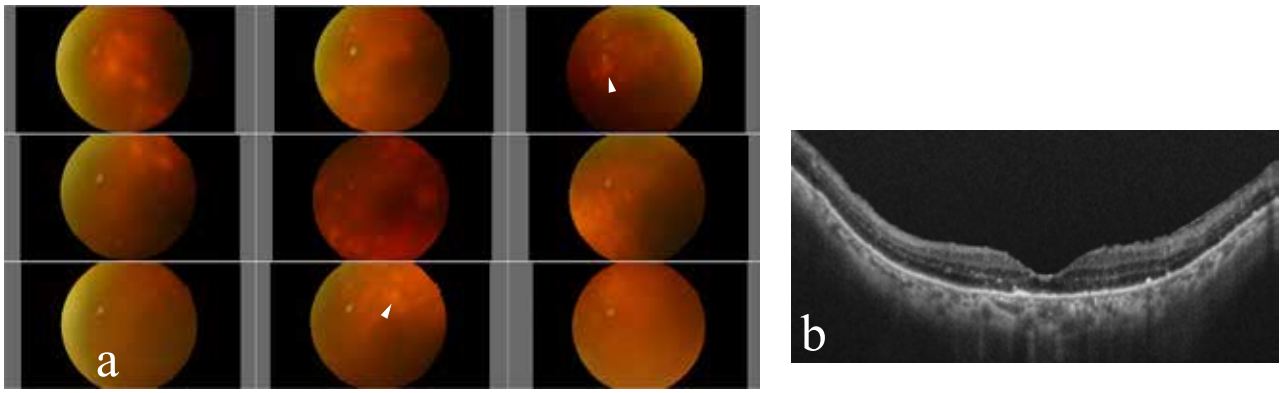

Fig. 6 Fundus photograph and OCT after 5 months of treatment: The fundus photograph shows improved vitreous opacities and evident depigmented plaques (arrowhead) (a). OCT images revealed disappearance of RD and improved choroidal folds.

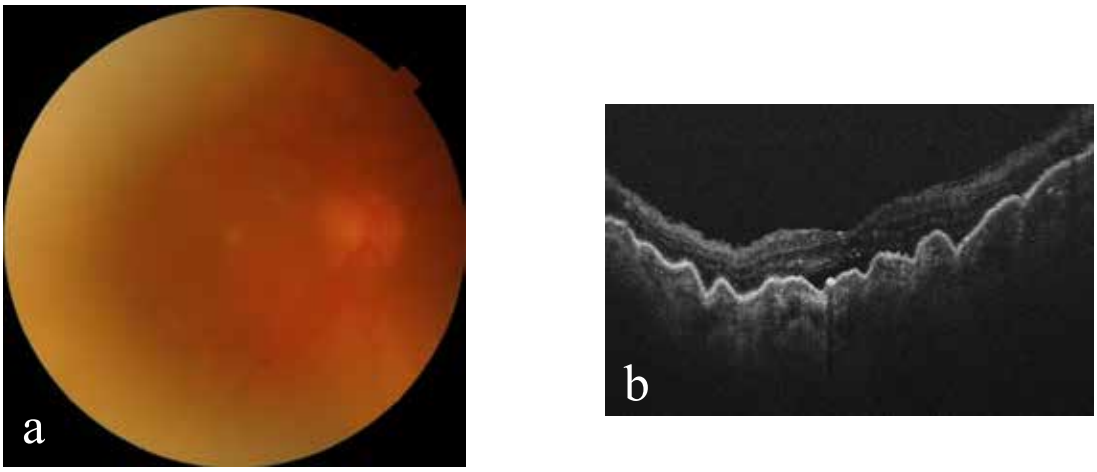

Fig. 7 Fundus photograph and OCT after 8 months of treatment: Fundus photograph shows recurrence of vitreous opacities (a). OCT shows RD, choroidal folds (b).

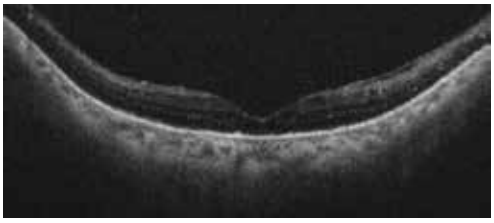

Fig. 8 OCT after 2 months of recurrence: In OCT, RD disappeared and choroidal folds improved.

detachment, and vitreous opacities were observed inferior to the right fundus (Fig. 2); however, there were no evident retinal tears. The left eye showed no abnormalities in the anterior or intermediate translucent body or fundus. Optical coherence tomography (OCT) revealed choroidal folds, and retinal detachment (RD) (Fig. 3); moreover, B-scan ultrasonography (B-scan) in the horizontal section showed a thickened sclera with fluid in Tenon's space (T-sign) (Fig. 4). Fluorescein fluorescence fundus angiography revealed fluorescence leakage from the optic nerve papillae (Fig. 5). No abnormalities were observed in the left eye. Blood tests and chest radiography revealed no abnormalities.

Based on the B-scan suggesting T-sign and various other findings, the patient was considered to have panuveitis associated with posterior scleritis. Accordingly, the patient was immediately started on systemic steroid administration after the patient's visit. On the day of the first visit, the patient received a sub-Tenon's capsule triamcinolone acetonide (STTA) injection; moreover, betamethasone sodium phosphate eye drops and prednisolone 40 mg/day were started as oral titrations. Another STTA was administered for residual RD after 2 months of treatment. The dose of prednisolone was reduced by 5 mg every week up to a daily dose of 15 mg, and then reduced by 5 mg every 3–4 weeks, with the patient's symptoms resolving (Fig. 6). However, recurrence was observed at 3 months after the end of prednisolone treatment (after 8 months of treatment; Fig. 7). Accordingly, 20 mg of prednisolone was started again. The patient underwent 2-month prednisolone treatment and currently has a visual acuity of 0.3 in the right eye, with no recurrence (Fig. 8).

DISCUSSION

Posterior scleritis have varying symptoms; however, rhegmatogenous retinal detachment due to a retinal tear should be promptly ruled out when there is marked retinochoroidal detachment associated with panuveitis. Rhegmatogenous retinal detachment occurs in 3.1% of patients with uveitis [4], which is ≈300 times higher than the incidence of rhegmatogenous retinal detachment in the general population [5]. The only curative treatment alternative for rhegmatogenous retinal detachment is surgery since steroid treatment is ineffective and requires prompt diagnosis. In our case, the diagnosis was difficult since the retinal detachment did not present with ocular pain or scleral hyperemia [6], which are often associated with posterior scleritis. Instead, the inflammatory findings in the anterior eye as well as the optic nerve papillae and retinal vessels on fluorescein funduscopy suggested panuveitis, with no obvious retinal tear was observed. Moreover, B-scan findings suggested thickened sclera with fluid in Tenon's space; therefore, the patient was considered to have retinochoroidal detachment due to panuveitis associated with posterior scleritis, rather than a rhegmatogenous retinal tear. Accordingly, the patient was started on steroids.

Contrastingly, uveal effusion syndrome [7] and Vogt-Koyanagi-Harada disease (VKH) are characterized by retinal detachment and choroidal folds without retinal rupture [8]. In our case, uveal effusion was ruled out because the ocular axis was 23.4 mm, which does not represent microphthalmia; the patient did not present with mobile retinal detachment with positional change, which is characteristic of uveal effusion; and RD improved with systemic corticosteroid administration. Additionally, the patient presented with RD and a thickened choroid in only one eye and did not present with auditory nerve symptoms such as tinnitus and sensorineural hearing loss, skin symptoms such as skin leukoplakia and alopecia, or meningeal symptoms such as headache. Therefore, VKH was considered to be negative. Since prompt initiation of steroid treatment significantly affects visual prognosis, it was important to promptly administer systemic steroid treatment after ruling out rhegmatogenous retinal detachment and accurate examinations, including ultrasonography, and establishing a diagnosis of retinal choroidal detachment.

In addition, the patient experienced recurrence at 3 months after symptom resolution. The recurrence rate of posterior scleritis recurrence has been reported to be 46% [9]; therefore, careful follow-up is necessary given the risk of recurrence. Although the timing of recurrence varies, it is necessary to consider the possibility of recurrence not only after steroid pulse therapy [10] but also after moderate-dose tapering therapy, such as in the present case, at least 3 months after resolution of the inflammation and discontinuation of steroid treatment.

This article reports a case of refractory posterior scleritis presenting as panuveitis with marked retinochoroidal detachment. Prompt diagnosis of posterior scleritis may allow a favorable prognosis. Accurate B-scan for establishing a definitive diagnosis and prompt systemic steroid administration may be useful.

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