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Case report

Severe panuveitis with relapsing polychondritis

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ABSTRACT

Purpose: Relapsing polychondritis is a rare multiorgan disease characterized by repeated episodes of inflammation and deterioration of cartilages. We report a case of relapsing polychondritis that presented with severe panuveitis.

Observations: A 53-year-old man visited our hospital because of ocular pain in both eyes of 2 weeks' duration. His best-corrected visual acuity was 20/20 in both eyes but he had severe hyperemia of the conjunctiva bilaterally. Inflammation in the anterior segment and vitreous opacity had been getting worse in his right eye. Systemic and topical treatments were not effective, and the visual acuity of the right eye was reduced to hand motion. Thus, pars plana vitrectomy with silicone oil tamponade was performed. After the operation, the vitreous opacities and white lesions in the retina were completely resolved. His visual acuity was improved to 20/20.

Three years later, he developed dizziness and swelling of both auricles of his ears, and he was found to have sensorineural deafness. He was diagnosed with relapsing polychondritis after a laryngoscopic examination.

Twelve months after the diagnosis, scleritis and panuveitis developed in his left eye, and his visual acuity fell to 20/2000. We performed pars plana vitrectomy with silicone oil tamponade on his left eye. After the vitrectomy, the inflammation of the left eye was resolved.

Conclusion: and importance: Ophthalmologist should be aware that severe panuveitis with vitreous opacities may be the initial signs of relapsing polychondritis. In addition, vitrectomy was effective for the treatment of the ocular complications.

1. Introduction

Relapsing polychondritis is a rare multiorgan disease characterized by repeated episodes of inflammation and deterioration of cartilage.[1] The diagnosis of relapsing polychondritis is made on the basis of clinical symptoms. McAdam et al. established 6 clinical criteria: (1) recurrent chondritis of both auricles; (2) nonerosive inflammatory polyarthritis; (3) chondritis of the nasal cartilage; (4) inflammation of the ocular structures; (5) chondritis of the respiratory tract involving laryngeal or tracheal cartilage; and (6) cochlear or vestibular damage manifested by neurosensory hearing loss, tinnitus, or vertigo.[2] Because of its rarity, the pathogenesis of this disease is still unclear, and epidemiological studies are not conclusive. Auricular chondritis and polyarthritis are the most common signs.

The eye is also a target organ.[1] We reported a patient who presented with ocular pain, scleritis, and severe panuveitis as the manifestations of relapsing polychondritis.

2. Case report

A 53-year-old man consulted our hospital with bilateral ocular pain of 2 weeks' duration. At the initial examination, his best-corrected visual acuity was 20/20 in both eyes but he had severe hyperemia of the conjunctiva and sclera bilaterally (Fig. 1). Ophthalmoscopy was essentially normal in both eyes. He was diagnosed with scleritis in both eyes and was treated with topical 0.1% betametazon four times a day. His laboratory results were within the normal limits, and the results for antineutrophil antibody, rheumatoid factor, toxoplasma serologic, and syphilis were negative. A general physical examination by an internist showed no systemic diseases.

Despite the treatments, the inflammation of the anterior segment worsened and an inflammation of the posterior segment in the right eye developed. Anterior chamber examination revealed 2 + cells, and fundus examination showed severe vitreous opacities and ill-defined white lesions along the retinal veins (Fig. 2A). He was then treated with systemic antibiotics, antiviral drug, and predonisone intravenously. However, his visual acuity decreased to 20/2000 within a few days.

Thus, we performed pars plana vitrectomy with silicone oil

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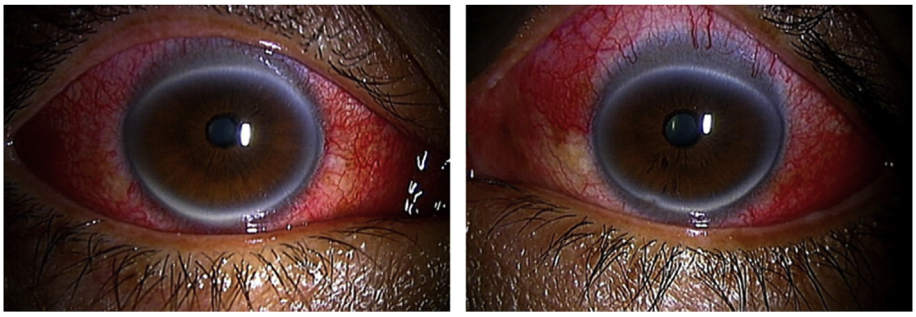

Fig. 1. Slit-lamp photograph of our patient at the first visit showing severe hyperemia bilaterally. His best-corrected visual acuity was both 20/20 in both eyes.

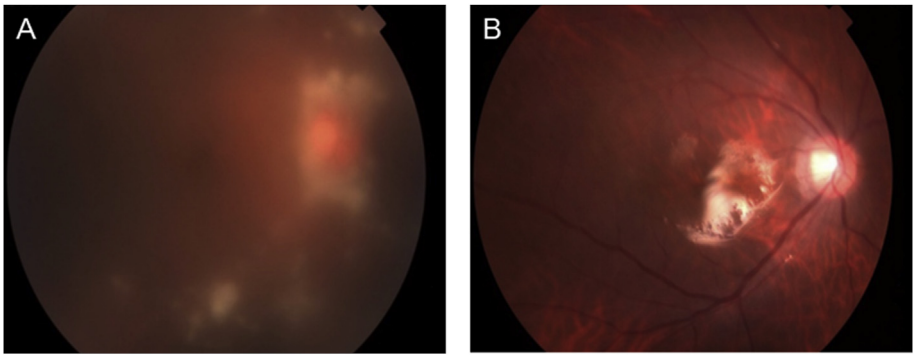

Fig. 2. Fundus photographs showing severe vitreous opacities and ill-defined white lesions located along the retinal veins (A). After pars plana vitrectomy with silicone oil tamponade, the vitreous opacities and white lesions in the retina are resolved (B).

tamponade on his right eye. After the operation, the vitreous opacities and white lesions in the retina were completely resolved (Fig. 2B). The visual acuity of the right eye improved to 20/20, and there were no recurrences for at least 3 years.

Three years after the initial presentation, he developed dizziness, and we referred him to an otolaryngologist in our hospital. The auricles of both ears were swollen (Fig. 3) and he had sensorineural deafness. The patient was diagnosed with relapsing polychondritis by the laryngoscopic examination. His dizziness and swelling of his auricles of ears were resolved with oral predonisone 10 mg/day.

Twelve months after the diagnosis, scleritis and panuveitis developed in his left eye, and his visual acuity fell to 20/2000. The fundus in his left eye was not visible due to vitreous opacities. He was treated with 0.1% betametazon four times a day but the eye did not respond to the therapy. We performed pars plana vitrectomy with silicone oil tamponade on his left eye. After the vitrectomy, the inflammation of the left eye was resolved, and there have been no recurrences with continued treatment of topical 0.1% betametazone four times a day.

3. Discussion

As best we know, there have been no reports of severe panuveitis which required pars plana vitrectomy in cases of relapsing polychondritis. Nicholas et al. reported uveitis with hypopyon in two cases of relapsing polychondritis.[3] They reported that the inflammation was localized to the anterior segment, and dilated ophthalmoscopic examinations showed no vitreous cell, no signs of retinitis, and normal optic discs. In our case, severe panuveitis was detected at the initial examination, and pars plana vitretomy was necessary. Paroli et al. reported uveitis with retinal occlusive vasculitis as a first symptom of relapsing polychondritis.[4] In their case, slit-lamp examination showed 1 + vitreous cells, and fluorescein angiography revealed dye leakages

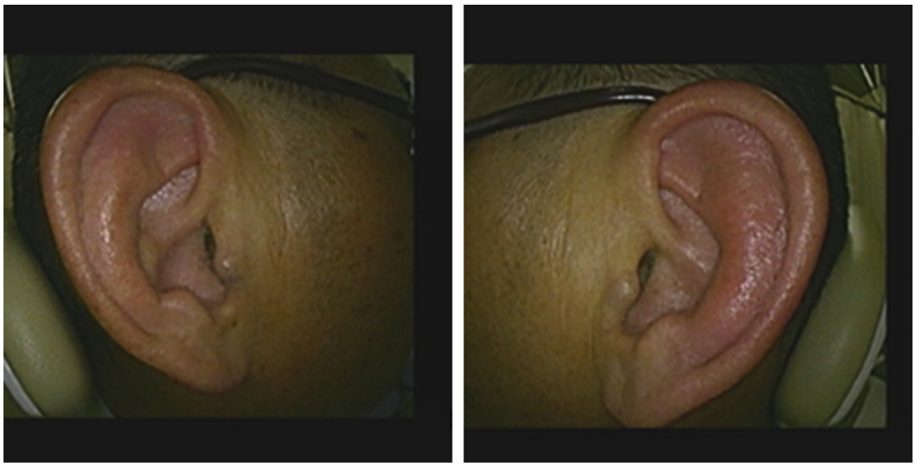

Fig. 3. Photographs of swelling and reddish auricles in both ears.

and vascular staining of both retinal arteries and veins. There was inflammation of the posterior segment but it was not so severe that pars plana vitrectmy was required.

The vitreous consist of 98% of water and extracellular matrix. The major extracellular matrix macromolecules are collagen type II and hyaluronic acid.[5] Several studies have reported the presence of antibodies for cartilage and collagen type II in the serum of patients with relapsing polychondritis.[6,7] It is quite possible that these antibodies for collagen type II caused the inflammation in the vitreous, and thus vitrectomy with the removal of the type II collagen was effective.

In conclusion, severe panuveitis accompanying relapsing polychondritis is a rare condition and vitrectomy is an effective treatment.

Patient consent

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflicts of interest

The authors have no financial disclosures relating this topic.

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