



Case report

Surgical interventions for late ocular complications of relapsing polychondritis

Yuan He^{*}, Huifeng Liu, Zhi Ji, Binliang Zhu

Department of Ophthalmology, The Second Affiliated Hospital of Xi'an Medical University, Ocular Immunology and Inflammation Institute, Shaanxi Provincial Clinical Research Center for Ophthalmology, Xi'an, China

ARTICLE INFO

Article history:

Received 24 February 2016
 Received in revised form
 17 November 2016
 Accepted 7 December 2016
 Available online 10 December 2016

Keywords:

Relapsing polychondritis
 Late ocular complications
 Surgical intervention

ABSTRACT

Purpose: To report a case of surgical interventions for a patient with relapsing polychondritis who presented with late ocular complications.

Observations: A 44-year-old male was diagnosed to have relapsing polychondritis on the basis of recurrent acute auricular chondritis, deformity of the ear, saddle nose deformity and painful nasal chondritis, acute ocular inflammation with conjunctivitis, episcleritis and keratouveitis, laryngotracheal chondritis, erythema nodosum in the skin, a history of polyarthritis, and abnormal blood examination findings. The acute ocular and auricular inflammation was resolved with oral corticosteroid treatment. Intraocular pressure (IOP) of the left eye was 60 mmHg as measured by Goldmann applanation tonometer. Gonioscopic observation revealed the presence of peripheral anterior synechiae and plateau iris configuration. Express drainage screw implantation was applied to the left eye, because topical and systemic medicines failed to decrease the IOP. After 12 months, complicated cataract aggravated in the right eye, and phacoemulsification operation was performed with corticosteroids administered during the perioperative period. His corrected visual acuity was 20/20 for the right eye, and the IOP remained below 21 mmHg for the left eye. The patient has been healthy, without any recurrence, for 36 months.

Conclusions and importance: The present case of relapsing polychondritis is the first to be reported wherein late ocular complications were alleviated by surgical interventions. Routine use of corticosteroids is necessary for successful anti-glaucoma and phacoemulsification operations.

© 2016 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Relapsing polychondritis is an unusual multiorgan autoimmune disease characterized by recurrent inflammation and progressive destruction of cartilaginous structures and collagen-rich tissues, including the eyes, ears, nose, skin, renal, musculoskeletal, heart, and respiratory systems.^{1–7} Ocular manifestations, including proptosis, eyelid edema, conjunctivitis, episcleritis, keratitis, uveitis, secondary open glaucoma, and retinopathy often occur in more than half of patients with relapsing polychondritis.^{1–8} We experienced the case of a patient with relapsing polychondritis who presented with ocular complications, including conjunctivitis, episcleritis, keratouveitis, secondary angle closure glaucoma, and complicated cataract. Among the published reports of relapsing

polychondritis with ocular manifestations, very few have described surgical management and outcome for late ocular complications. Here, we report the case of successful express drainage screw implantation and phacoemulsification operations in a patient with relapsing polychondritis with late ocular complications.

2. Case report

A 44-year-old man visited a local ophthalmic clinic with the complaint of recurrent bilateral and painful red eyes, swollen eyelids, and blurred vision that had started 5 months earlier. He had been treated with antibiotics and anti-allergy medicine topically and systemically for bacterial and allergic conjunctivitis; however, the treatment could not alleviate the inflammation. On the contrary, the symptoms of redness and pain in the eyes were aggravated. He was also being treated with topical corticosteroids for interstitial keratitis for a long period of time, and the inflammation was controlled only for a very short duration. An ophthalmic examination showed acute ocular inflammation with redness and

^{*} Corresponding author. The Second Affiliated Hospital of Xi'an College of Medicine, 167 Fang Dong Street, Xi'an 710038, China.

E-mail address: openji7127@hotmail.com (Y. He).

mixed conjunctival congestion in both eyes with dilated vessels, conjunctival swelling (Figs. 1 and 2), and auricular chondritis that led to a swollen, inflamed, painful, and soft left ear (Fig. 3). There was also a saddle nose deformity and erythema nodosum in the skin (Fig. 4). His corrected visual acuity was hand motions (HM)/30 cm for the right eye and 20/20 for the left eye. The intraocular pressure (IOP) for the two eyes was 16 mmHg (right) and 60 mmHg (left), as measured by a Goldmann applanation tonometer. An ocular examination revealed diffuse conjunctivitis, episcleritis, and keratouveitis with dense whole intrastromal corneal infiltrate in the right eye (Fig. 1). The left eye presented with diffuse conjunctivitis and episcleritis (Fig. 2). The posterior section of the right eye could not be observed clearly because of the cloudy cornea. The anterior chamber angles were shallow for both the eyes. Ultra-biomicroscopy (UBM) (Fig. 5) revealed narrow anterior chamber angles for both the eyes and accumulation of inflammatory cells at the corner of the anterior chamber angles. The flow of inflammatory cells in the aqueous humor also indicated anterior uveitis. Both narrow anterior chamber angles and anterior uveitis were caused by this secondary IOP elevation. Gonioscopic observation revealed the presence of bridge-like peripheral anterior synechiae that discontinuously occupied more than 60% of the angle and plateau iris configuration in the left eye.

A corneal scrape was taken at the presentation time but failed to show any growth on standard culture media. A blood examination revealed that serum total protein levels, total hemolytic complement activity (CH50), complement C3, C-reactive protein levels (39.5 mg/L, the normal value is less than 5 mg/L), and Westergren sedimentation rate (46 mm/h, the normal value is less than 20 mm/h) were elevated with mild leukocytosis. The test results for rheumatoid factor, antinuclear antibodies, antimitochondrial antibodies, HLA-DR4-DNA, HLA-DW53-DNA, and HLA-B27-DNA were normal. A computed tomography (CT) scan showed lower airway stenosis (Fig. 6) and calcified laryngeal cartilages (Fig. 7).

A diagnosis of relapsing polychondritis was made on the basis of recurrent acute auricular chondritis with marked redness; swelling, warmth, and tenderness of the pinna; deformity of the ear that followed recurrent acute attacks; saddle nose deformity and painful nasal chondritis; acute ocular inflammation; laryngo-tracheal chondritis; erythema nodosum in the skin; a history of polyarthritis; and abnormal blood examination findings.^{9,10} The acute ocular and auricular inflammation was resolved by treatment with 1 mg/kg/day oral prednisone (Fig. 8).⁸ Methotrexate was started at a dose of 7.5 mg/week, and the corticosteroids were tapered.⁸ Because 0.5% timolol ophthalmic solution, travoprost, brinzolamide, and 2% brimonidinetartrate failed to decrease the

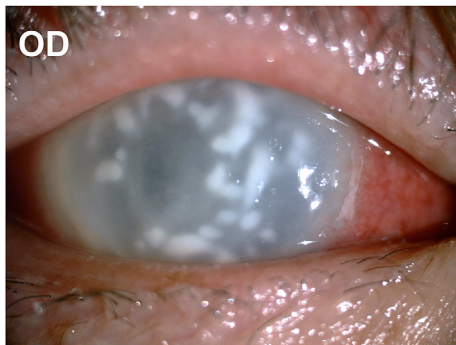


Fig. 1. Red right eye with conjunctival and episcleral congestion. Ocular examination revealed diffuse conjunctivitis, episcleritis, and dense whole intrastromal corneal infiltrate in the right eye. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

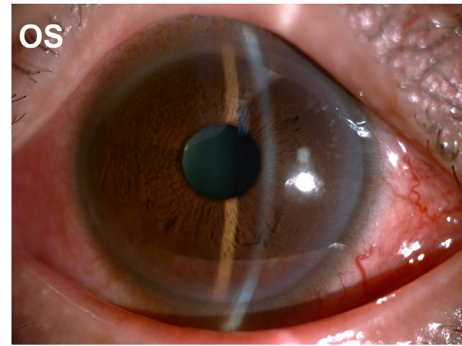


Fig. 2. Diffused conjunctivitis and episcleritis in the left eye.

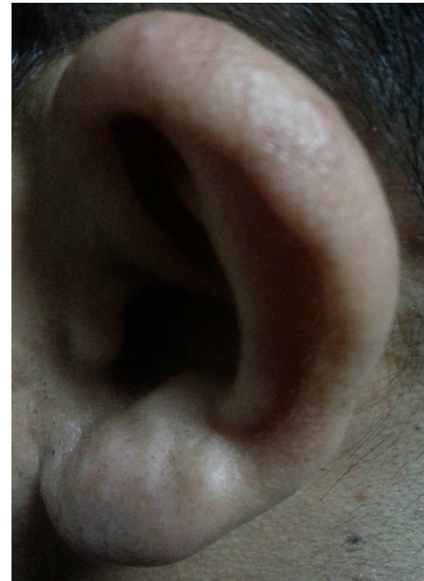


Fig. 3. An inflamed, swollen, and tender left ear.



Fig. 4. Saddle nose deformity and inflammation. A type of inflammation with nodular, erythema, and pustule present in the skin.

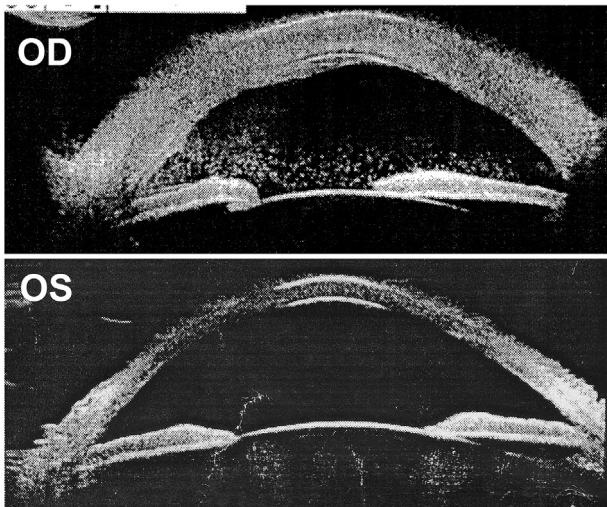


Fig. 5. Ultra-biomicroscopy revealed narrow anterior chamber angles and flow of inflammatory cells in the aqueous humor.

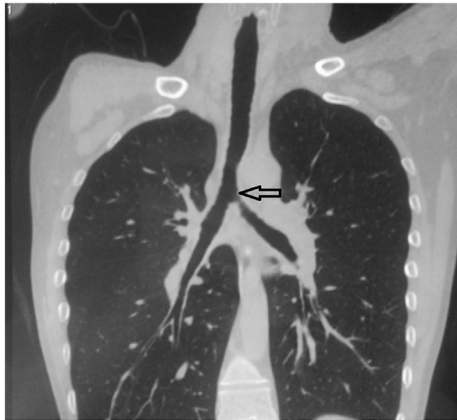


Fig. 6. Computed tomography scan showed lower airway stenosis (arrow).

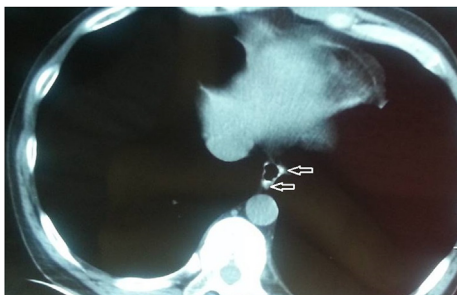


Fig. 7. Computed tomography scan showed calcified laryngeal cartilages (arrow).

IOP in the left eye, express drainage screw implantation was applied when the acute ocular inflammation was resolved. Mitomycin was not applied during the operation considering the side effects of the inflammation on the pathological change in the conjunctiva, sclera, and cornea. There was aqueous humor leakage from the filtering bleb during the first 3 months (Fig. 9). The IOP was controlled and remained at around 10 mmHg. After 3 months, there was no leakage of the aqueous humor from the filtering bleb, and the IOP remained below 21 mmHg. After 12 months, cataract

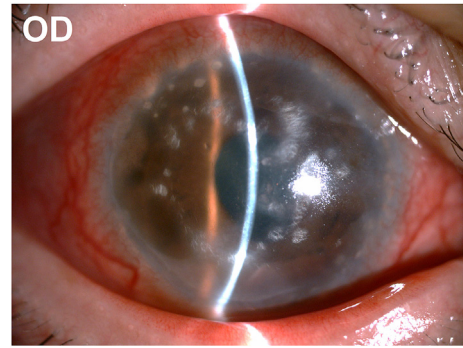


Fig. 8. Ocular examination revealed decreased conjunctivitis, episcleritis, and dense whole intrastromal corneal infiltrate in the right eye with a little scar in the intrastromal cornea. The patient was given oral corticosteroid treatment.

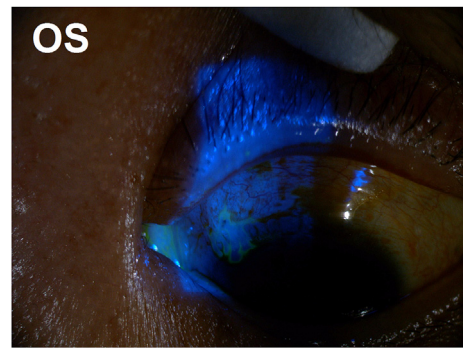


Fig. 9. Aqueous humor leakage from the filtering bleb with fluorescein staining (arrow) in the left eye after express drainage screw implantation.

complication aggravated in the right eye; phacoemulsification operation was performed and corticosteroids were routinely used during the perioperative period. The patient's corrected visual acuity was 20/20 for the right eye, and there was no aqueous humor leakage from the cataract incision at the cornea. The patient has been healthy, without recurrence, for 36 months.

3. Discussion

Relapsing polychondritis mainly affects cartilaginous tissues and collagen-rich tissues.^{1–13} Eyeballs are collagen-rich organs that are unavoidable sites of inflammation in patients with relapsing polychondritis, with the affected areas including the conjunctiva, sclera, cornea, uvea, retina, and optic nerve. Therefore, conjunctivitis, episcleritis or scleritis, keratitis, uveitis, and retinopathy are very common ocular manifestations in these patients.^{2–8,14,15}

The present patient was referred to us by a local ophthalmology clinic 5 months after his first visit, and he presented with a common comprehensive manifestation of relapsing polychondritis. Considering the acute inflammation and progressive destruction of cartilaginous tissues, especially the high risk of tracheobronchial collapse leading to obstructive ventilatory impairment, a systemic demand of corticosteroid or immunomodulatory agents is invaluable to avoid a delayed diagnosis and the subsequent related mortality.^{2,4} In this case, the acute ocular and auricular inflammation was resolved with oral corticosteroid treatment; methotrexate was then started, and the corticosteroids were tapered.

Although there are few reports on relapsing polychondritis occasionally inducing complicated cataract, secondary open angle glaucoma, and angle closure glaucoma, case reports on the surgical

management and outcome for these late ocular complications have rarely been published. In the present case, UBM and gonioscopic examinations revealed the presence of narrow angle, bridge-like peripheral anterior synechiae that was discontinuous with plateau iris configuration, deeply involved in angle closure, and secondary IOP elevation. The UBM also revealed active anterior uveitis with the flow of inflammatory cells in the aqueous humor and subsequent accumulation at the corner of the anterior chamber angles. This provided the evidence for bridge-like peripheral anterior synechiae and a possible trabecular inflammation, which could reduce the outflow by occlusion of the angle, revealing the mechanism of IOP elevation due to secondary angle closure glaucoma.¹⁶ However, any IOP lowering medicine failed to decrease the IOP effectively in the present case. Considering connective tissue inflammatory conditions of conjunctivitis, episcleritis, and stromal keratitis, the aqueous humor leakage from the filtering bleb could not possibly be avoided after any anti-glaucoma surgery. With systemic treatment of the patient, there was no more leakage of the aqueous humor from the filtering bleb after 3 months of express drainage screw implantation. Phacoemulsification operation was also performed successfully with the application of corticosteroids during the perioperative period.

4. Conclusion

The present case of relapsing polychondritis is unique as a late presentation with ocular complications that required surgical interventions. Routine usage of corticosteroids or an immunomodulator is recommended as part of surgical management for such patients.

4.1. Patient consent

Written consent to publish the details of this case was obtained from the patient. The Ethics Committee approved the report.

Funding

This work was supported by grants from the National Natural Science Foundation of China (No. 81100665), and Science and Technology of Shaanxi Province of China (No. 2013KJXX-31).

Conflict of interest

None.

Authorship

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

Acknowledgments

The authors thank Dr. Kar Wah Leung, The Hong Kong University of Science and Technology, and Elsevier Language Editing Services for editing the manuscript for grammar and style.

References

- McAdam LP, O'Hanlan MA, Bluestone R, et al. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. The arthropathy of relapsing polychondritis. *Med Baltim.* 1976;55:193–215.
- Peebo BB, Peebo M, Frennesson C. Relapsing polychondritis: a rare disease with varying symptoms. *Acta Ophthalmol Scand.* 2004;82:472–475.
- Matoba A, Plager S, Barber J, et al. Keratitis in relapsing polychondritis. *Ann Ophthalmol.* 1984;16:367–370.
- Sallam A, Islam T, Parmar DN, Parmar. Keratouveitis as a first presentation of relapsing polychondritis. *Case Rep Med.* 2010;2010:176514.
- Vroman D, Solomon KD. Relapsing polychondritis. *N Engl J Med.* 2003;349:e3.
- Rodríguez-Colón G, Palestine AG. Images in clinical medicine. Polychondritis with auricular and ocular involvement. *N Engl J Med.* 2015;373:2562.
- Rovenský J, Sedláčková M. Relapsing polychondritis. *Cas Lek Cesk.* 2012;151(2):64–68.
- Yoo JH, Chodosh J, Dana R. Relapsing polychondritis: systemic and ocular manifestations, differential diagnosis, management, and prognosis. *Semin Ophthalmol.* 2011;26(4-5):261–269.
- Staats BA, Utz JP, Michet Jr CJ. Relapsing polychondritis. *Semin Respir Crit Care Med.* 2002;23(2):145–154.
- Arnaud L, Devilliers H, Peng SL, et al. The Relapsing Polychondritis Disease Activity Index: development of a disease activity score for relapsing polychondritis. *Autoimmun Rev.* 2012;12(2):204–209.
- Jaksh-Wartenhorst R. Polychondropathia. *Wiener Arch. Intern Med.* 1923;6:93–100.
- Piette JC, Frances C. Relapsing polychondritis. *N Engl J Med.* 1995;332:580.
- Nakajima T, Sekine Y, Yasuda M, et al. Long-term management of polychondritis with serial tracheobronchial stents. *Ann Thorac Surg.* 2006;81:e24–e26.
- Brink H, Rademakers J, Verbeek A, et al. Ocular manifestations of relapsing polychondritis. Three case histories. *Doc Ophthalmol.* 1994;87:159–166.
- Isaak BL, Liesegang TJ, Michet Jr CJ. Ocular and systemic findings in relapsing polychondritis. Conjunctival and episcleral injection in drug abuse. *Ophthalmology.* 1986;93:681–689.
- Kashiwagi K, Furuya T, Kashiwagi F. Changes in peripheral anterior chamber depth of a case of relapsing polychondritis with recurrent secondary angle closure glaucoma. *Open Ophthalmol J.* 2008;2:1–4.