

Sympathetic ophthalmia in eye with pathologic myopia

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ABSTRACT

Purpose: To present our findings in a case of SO that developed in an eye with pathologic myopia.

Observations: The patient was an 83-year-old woman who was examined one month after an ocular trauma to the right eye. She was found to have signs of uveitis with multiple serous retinal detachments in the non-injured contralateral left eye. In addition, she had hearing loss and mononuclear pleocytosis of the spinal fluids. Swept-source OCT images showed focal and choroidal thickening in areas with abrupt edges that was restricted to the regions with more normal appearing choroid. Bruch's membrane was damaged at the edge of the focal choroidal thickening. The ocular and systemic findings were rapidly resolved after systemic corticosteroid therapy.

Conclusions and importance: The pathobiological and clinical course of SO is nearly identical to Vogt-Koyanagi Harada disease (VKH) although its pathogenesis of autoimmunity had not been definitively established. In eyes with pathologic myopia, the choroid is extremely thin and sometimes completely absent. The findings in this rare case indicate that in eyes with thin choroid, the OCT findings typical of SO might be different from those seen in non-highly myopic eyes. Thus, the pre-status of the choroid may affect the choroidal thickening in pathological conditions. This case gives us a valuable insight in understanding the pathology of SO and characteristic of pathologic myopia.

1. Introduction

Sympathetic ophthalmia (SO) is a sight-threatening and systemic disorder of one eye caused by ocular trauma or surgery to the contralateral eye. The characteristic ocular findings in the non-injured eye consist of granulomatous uveitis and multiple serous retinal detachments with vision reduction. The typical systemic findings are headaches, hearing loss, vitiligo, and poliosis. After the onset, the clinical course and pathology is nearly identical to Vogt-Koyanagi-Harada (VKH) disease.^{1,2} In SO, melanocyte specific immunity is acquired by injury to uveal tissues following a penetrating ocular trauma or surgery. Therefore, melanocyte-rich tissues are the major sites of the autoimmune inflammation, e.g., the ocular uvea, dermis of the skin, meninges of the brain, and the inner ear. All of the changes at these sites are considered to be melanocyte-specific autoimmune reactions.

A choroidal thickening due to the infiltration of inflammatory cells as seen in optical coherence tomographic (OCT) images has been reported to be a useful marker for diagnosing VKH and SO.^{3,4} The uniform

choroidal thickening is seen spanning across the posterior fundus in eyes with VKH and SO.³⁻⁶

Pathologic myopia is characterized by the presence of eye abnormalities such as posterior staphylomas,⁷⁻⁹ and by the presence of various lesions in the macula and optic nerve. These alterations can lead to visual impairments.⁹⁻¹² Enhanced depth imaging (EDI)-OCT has shown that the choroid is thinned in highly myopic eyes and the degree of thinning was significantly correlated with an increase of the axial length.¹³ Swept-source OCT examinations showed that even in eyes with diffuse chorioretinal atrophy, i.e., mild stage of myopic maculopathy according to META-PM classification,¹⁴ the choroid is almost absent except for sporadic large choroidal vessels.¹⁵ The choroid in eyes with pathologic myopia is too thin to measure in many cases.¹⁶ A search of PubMed did not extract any publications reporting the occurrence of VKH or SO in patients with pathologic myopia.

The purpose of this report is to present our findings in a case of SO that developed in an eye with pathologic myopia. Swept-source OCT showed unusual findings including focal and abrupt thickening of the

; SO, Sympathetic Ophthalmia; VKH, Vogt-Koyanagi Harada Disease; OCT, Optical Coherence Tomography.

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choroid only in the areas of remaining choroid.

2. Case report

An 83-year-old woman was referred to the Department of Ophthalmology and Visual Science of the Tokyo Medical and Dental University Hospital complaining of blurred vision in both eyes and hearing loss bilaterally. A month before the initial examination in our department, she had fallen and hit her right eye. She was examined by a local ophthalmologist who diagnosed her with vitreous hemorrhages and a subconjunctival herniation of the iris of the right eye. She was treated by observation only without surgery or systemic administration of corticosteroids. Her visual symptoms gradually improved in the right eye, however she developed a reduction of vision in the left eye at about 5 days before and a month after the initial visit to our hospital.

At our initial examination, her best-corrected visual acuity (BCVA) was hand motion in the right eye and her decimal BCVA was 0.3 in the left eye. Both eyes were highly myopic with axial lengths of 31.4 mm for the right eye and 33.7 mm for the left eye. In addition, the fundus of both eyes showed multiples areas of patchy chorioretinal atrophy on a background of yellowish diffuse atrophy. The patient had been diagnosed with myopic choroidal neovascularization in the right eye 10 years earlier, and the BCVA had been poor since then. Both eyes were considered to have pathologic myopia because of the presence of the lesions specific to myopic maculopathy. She reported that her left eye had retained relatively good decimal BCVA of about 0.7. Our findings showed that the intraocular pressure was 14 mmHg in the right eye and 15 mmHg in the left eye.

Slit-lamp examination of the right eye showed diffuse subconjunctival hemorrhage and a herniation of iris into the subconjunctival space over a wide area resulting in aniridia (Fig. 1). Her right fundus was blurred due to the vitreous hemorrhage. In the left eye, anterior uveitis together with multiple areas of serous retinal detachments were seen.

Fluorescein angiography (FA) of the left eye showed multiple hyperfluorescent spots in the early phase followed by dye leakage and pooling in the late phase (Fig. 2B and C). Indocyanine green angiography (ICGA) of the left eye showed a decrease in the number of choroidal vessels in the venous phase (Fig. 2E). Dark hypofluorescent spots were also observed diffusely in the late phase of ICGA (Fig. 2F and G). OCT examinations (HD-OCT Cirrus®, Zeiss Co., Germany or DRI-OCT Atlantis®, Topcon Co., Japan) showed serous retinal detachments and marked thickening of the choroid of the left eye (Fig. 3A, B, 3C). The choroidal thickening was focal and the borders of the thickened choroid were sharp (Fig. 3C). There was an abrupt transition from an area whose choroid was completely lost within the area of patchy chorioretinal atrophy to an area of marked thickening of the choroid. In the peripapillary gamma zone, the area around the optic nerve, Bruch's membrane was torn and its disrupted ends were visible (Fig. 3C). Systemic evaluations showed sensorineural hearing reduction and pleocytosis of the cerebrospinal fluid (33/μl). Considering all of these findings, the patient

was diagnosed with SO.

Systemic administration of corticosteroid was started with intravenous 1000 mg of methylprednisolone for 3 days followed by oral prednisolone 1mg/kg with gradual tapering along with cyclosporine. Surgical removal of the subconjunctivally herniated iris and pars plana vitrectomy for the vitreous hemorrhage were also performed. It was strongly suspected that the iris tissue was herniated from previous scleral wound of extra capsular cataract extraction, despite the wound was closed at the time of surgery. The steroid therapy and surgical treatment rapidly improved the ocular signs as well as the hearing reduction. Six weeks later, the serous retinal detachment had almost disappeared with multiple hyperfluorescent spots with late pooling in FA (Fig. 2D). There was a reduction in the number of hypofluorescent dark spots in ICGA images (Fig. 2H). Swept-source OCT examinations showed that the choroidal thickening was markedly decreased, and the areas with thickened choroid were more restricted (Fig. 3C and D). Three months later, OCT examinations showed no retinal detachment or choroidal thickening. The choroid was extremely thin over the entire posterior pole of the fundus. The decimal BCVA of the left eye improved to 0.7.

3. Discussion

Recent studies have reported on the helpfulness of OCT in diagnosing VKH and SO.⁴⁻⁶ In the acute phase of VKH, the subfoveal choroid is thickened^{5,6} which was considered to be due to the massive infiltration of inflammatory cells into the choroid. The choroidal thickening is generally diffuse and seen over the entire extent of the choroid in OCT sections across the macula. A boundary of the choroidal thickening was not obvious.

Our case had choroidal thickening that was detected in the OCT images, however the OCT findings were unusual and different from that seen in non-highly myopic eyes. The left eye had patchy chorioretinal atrophy and a large peripapillary gamma zone due to the pathologic myopia. It was recently reported that these two types of lesions were both characterized by Bruch's membrane defects.¹⁷ In the area with patchy atrophy or with a large peripapillary gamma zone, the entire thickness of the choroid was lost, and the inner retina sits directly on the sclera. Even in the mild stage of myopic maculopathy, e.g., diffuse chorioretinal atrophy, the choroid is markedly thinned with sporadically remaining large vessels.

Swept-source OCT showed that the choroidal thickening was focal and not diffuse. It was different from the diffuse thickening of the choroid that is usually seen in VKH or SO disorders.⁴⁻⁶ The transition from the area with no choroid, in the area of patchy atrophy or gamma zone, to the area of thickened choroid was abrupt, which caused a tear of Bruch's membrane along the boundary of the thickened choroid. The OCT findings showed that the remaining choroid outside the patchy atrophy or gamma zone was thickened. However, the adjacent areas without choroid prior to the onset of SO remained the same.



Fig. 1. Slit-lamp photographs of the injured right eye. The herniated iris tissue is seen in the nasal superior subconjunctival space of the anterior globe. Intraocular blood coagulant can also be seen.

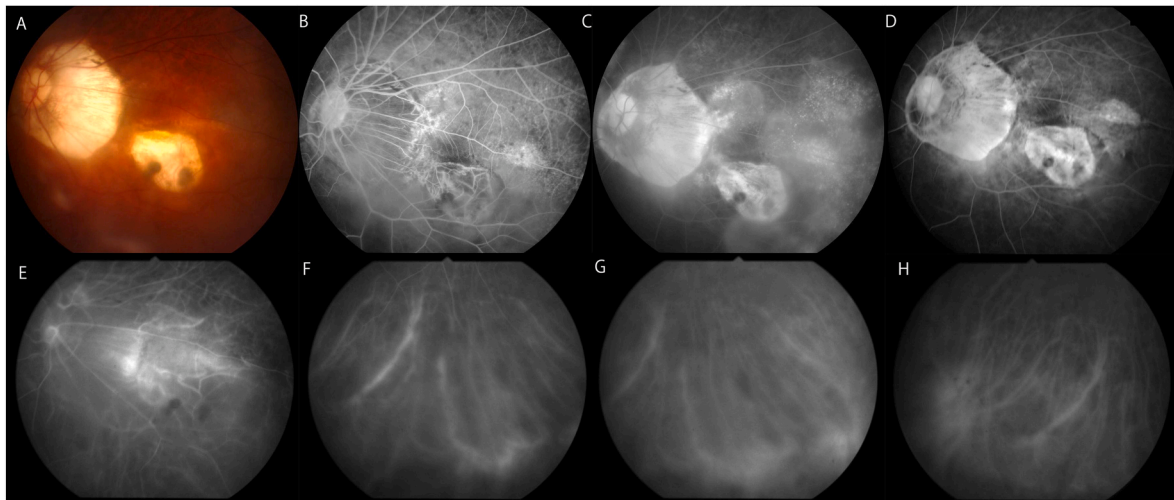


Fig. 2. Fundus photographs, fluorescein angiograms (FA), and indocyanine green angiography (ICGA) of the left uninjured eye at the initial visit (A-C, E-G) and 6 weeks later (D, H). A. Color fundus photograph of the left uninjured eye at initial visit shows patchy atrophy in the macula and peripapillary atrophy with suspected serous retinal detachment. B. FA image of the left uninjured eye at the initial examination showing multiple hyperfluorescent spots in the early phase. C. Late phase of FA of the left eye at the initial visit with dye leakage and pooling from the hyperfluorescent spots. D. FA of late phase of the left eye at 6 weeks after the initial visit following treatment. No apparent dye leakage and pooling. E. ICGA image of the venous phase showing a fewer number of visible choroidal vessels. F. and G. Hypofluorescent dark spots can be seen diffusely at the peripheral fundus in the late phase of ICGA. H. Remaining but less apparent hypofluorescent dark spots in the ICGA at 6 weeks after the initial examination and following treatment. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

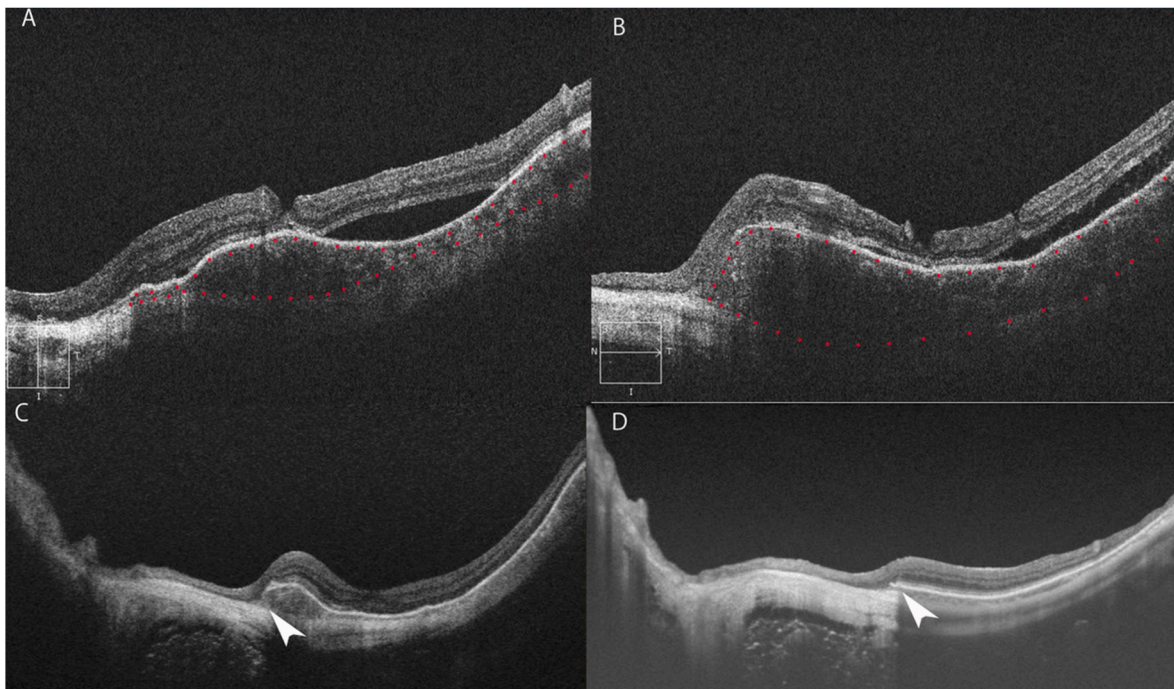


Fig. 3. Optical coherent tomographic (OCT) images of the non-injured left eye at the initial visit and after treatment. A. OCT longitudinal image shows subretinal fluid (SRF) inferior to macula with possible thickened choroid. Thickened choroid in image A is outlined by red dots. B. OCT lateral section showing SRF temporally adjacent to macula. An elevation of the nasal retina is also seen in the macula with suspected choroidal thickening. Choroid in image B is outlined by red dots. C. Focal thickened choroid abruptly ends at gamma zone. RPE-Bruch's membrane tear (arrowhead) is seen in the swept source OCT lateral section image nasal to the macula. D. Focal thickening of choroid is not seen in the swept source OCT image of post treatment (6 weeks after initial visit). RPE-Bruch's membrane tear is present. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

These findings also suggested that the OCT features which were considered integral for diagnosing VKH or SO may be markedly different according to the original status of the choroid. Thus, caution is needed in the interpretation of the OCT findings in eyes with high myopia because the choroid may not be uniformly thickened.

This patient developed SO, however, it is interesting to know that a PubMed search by using 'Vogt-Koyanagi Harada' and 'high myopia' or 'pathologic myopia' as key words did not extract any publications related to our findings. Although about 4,000 patients with pathologic myopia have been followed in the High Myopia Clinic at Tokyo Medical

and Dental University, none of them has developed VKH or SO. Thus, VKH never or almost never develops in the patients with pathologic myopia. In addition to the extreme thinning of the choroid, choroidal melanocytes might be lost in eyes with pathologic myopia. This should be investigated histologically in the future.

4. Conclusions

We report a case of SO in an eye with pathologic myopia. The patient had pathologic choroidal alterations characteristic of SO. The choroidal thickening in the OCT images, an important marker for diagnosing SO or VKH, was markedly different than that seen in non-highly myopic eyes.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

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Declaration of competing interest

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This case has been reported by Kawashima Y et al. in American Journal of Otolaryngology Head and Neck Surgery 36(4): 606-10,2015. The manuscript was written focusing on the otolaryngologic findings.

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