



## Case report

## Two cases with retinitis pigmentosa that developed severe retinal atrophy long after vitreo-retinal surgery



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## ABSTRACT

**Purpose:** To report two cases with retinitis pigmentosa (RP) who underwent vitreo-retinal surgery and developed severe retinal atrophy.

**Observations:** Case 1 was a 36-year old man who underwent 20-gauge pars plana vitrectomy (PPV) for a macular hole (MH), and Case 2 was a 71-year-old man who also underwent 20-gauge PPV for an epiretinal membrane (ERM). During 13 years follow-up period, severe retinal atrophy developed near the macula that progressed rapidly and the best-corrected visual acuity (BCVA) was reduced to 1.1 logarithm of the minimum angle of resolution (logMAR) units in the MH case and to no light perception in the ERM case.

**Conclusions and importance:** An unexpected severe retinal atrophy can develop long after vitreo-retinal surgery in RP patients.

## 1. Introduction

It is known that some RP patients can develop macular disorders such as an epiretinal membrane and macular hole. We have had two patients with retinitis pigmentosa who underwent vitreo-retinal surgery for a MH and an ERM who were followed for over 13-years. Both eyes developed severe retinal atrophy long after the surgery. A search of Medline/PubMed did not extract any publications of more than 10 years follow-up after vitreo-retinal surgery for a macular disorder in RP patients. The purpose of this report is to present our findings in these two RP patients who underwent vitrectomy and developed severe retinal atrophy long after the surgery.

## 2. Findings

## 2.1. Case 1

A 36-year-old man was referred to our department for surgery for a MH. He was aware of having night blindness since he was 15-years-old, and he was diagnosed with RP at the previous hospital. His older brother was also diagnosed with RP, however his sister, parents, and son were not affected. The ocular conditions of his grandparents were not known. Consanguineous marriage was not present in his family, and he had no history of ocular diseases other than RP or any ocular trauma.

At the preoperative examination, his best-corrected visual acuity (BCVA) was 0.4 logarithm of the minimum angle of resolution (logMAR) units in his right eye and 0 logMAR units in the left eye. The axial length of the right eye was 24.28 mm, and cataracts were not observed in both eyes. Fundus examinations revealed intraretinal bone-spicule pigmentations in the midperiphery, waxy disc pallor, and narrowing of the retinal vessels in both eyes. Both the scotopic and photopic electroretinograms (ERGs) were non-recordable in both eyes. Optical coherence tomography (OCT) showed a full-thickness macular hole in the right eye, and a posterior vitreous detachment (PVD) was not observed. No signs of ocular inflammation were observed. In January 2006, he underwent 20-gauge pars plana vitrectomy with Indocyanine green (ICG)-assisted internal limiting membrane (ILM) peeling and 20% sulfur hexafluoride (SF6) gas tamponade. The total operation time was 60 minutes. After one week of face down positioning, the OCT images confirmed a closure of the MH. Three years after the surgery, his BCVA in the right eye had improved to 0.1 logMAR units but there were retinal atrophic lesions around the macula that did not exist before the surgery (Fig. 1). The lesions gradually expanded and at 9 years after the surgery, his BCVA had decreased to 0.3 logMAR units. At 12 years after the surgery, the atrophy had expanded to the edge of the fovea, and the BCVA had further decreased to 1.1 logMAR units. The BCVA in the fellow eye was 0.1 logMAR units. The central foveal thickness (CFT) of the right and left eyes at the final

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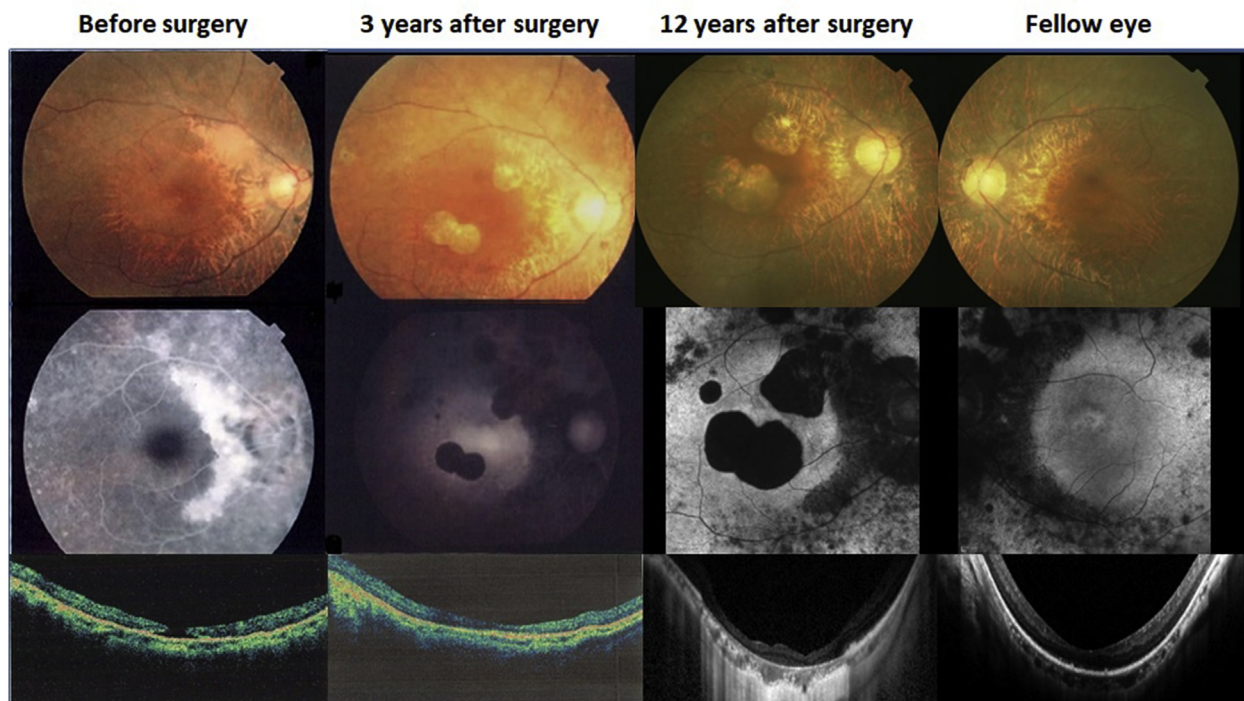


Fig. 1. Images of Case 1 before and after surgery.

Upper: Color fundus photographs. Middle: Fundus autofluorescence images. Only the image of before surgery is fluorescein angiographic image. Lower: Optical coherence tomographic images. The images of fellow eye are 12 years after surgery.

visit was 170  $\mu\text{m}$  and 326  $\mu\text{m}$ , respectively.

## 2.2. Case 2

A 71-year-old man was aware of his night blindness and decreased visual acuity, and he was diagnosed with RP when he was 40-years-old. He had no family history of any chorioretinal diseases, and consanguineous marriages were not present in his family. Ophthalmoscopic and ERG findings confirmed the diagnosis of typical RP. His right eye had undergone phacoemulsification and a yellow-colored acrylic foldable intraocular lens had been implanted through a superior sclerocorneal incision without any complications in our department in January 2005. Thereafter, there was a gradual increase in the vitreous traction on the macular in the right eye as seen in the OCT images, and he complained of metamorphopsia. A PVD had already occurred. He was diagnosed with an ERM in the right eye and was scheduled for vitreo-retinal surgery. His preoperative BCVA was 0.7 logMAR units in both eyes, and the axial length was 24.58 mm in the right eye. No signs of ocular inflammation were observed. In May 2005, he underwent 20-gauge pars plana vitrectomy to remove the ERM and release of the retinal tension. Pure SF<sub>6</sub> gas was used to tamponade the retina. The total operation time was 40 minutes. After the surgery, the traction on the macula was removed and an improvement of the macular morphology was observed in the OCT images. However, retinal atrophy including the fovea appeared after the surgery and gradually spread (Fig. 2). The patient lost all light sense in his right eye which was confirmed 8 years after the surgery. Currently 13 years after surgery, the BCVA in the right eye is no light perception (NLP) and the BCVA is 0.0 logMAR units in the left eye. The CFT of right eye was 587  $\mu\text{m}$  before the surgery, and it was reduced to 58  $\mu\text{m}$  at the final visit. On the other hand, the CFT of left eye was 427  $\mu\text{m}$  at the final visit.

## 3. Discussion

Our results showed that the two RP patients who had undergone

vitreo-retinal surgery developed macular atrophy long after the surgery. The atrophy was progressive, and the vision decreased to 1.1 logMAR units in the patient with a MH and to NLP in the patient with an ERM. Hagiwara et al. reported that macular abnormalities such as MH, ERM, and vitreomacular traction syndrome (VMT) are present in 1.9% of RP patients.<sup>1</sup> There have been at least two recent reports of vitreo-retinal surgery performed for macular disorders in RP patients.<sup>2,3</sup> The authors reported a good recovery of the visual acuity and an improvement of the retinal morphology, but there are not many reports that showed a worsening of the BCVA after the surgery as in our two patients.

In Case 1, it was possible to confirm multiple circular degeneration lesions near the macula at 3 years after the surgery (Fig. 1). These lesions gradually enlarged, and the BCVA worsened. What was interesting was that the atrophy spread around the macular but not into the fovea in this patient. In Patient 2, there was a gradual degeneration of the fovea and its surroundings, which led to NLP in 8 years after the surgery. Unlike Patient 1, the degeneration extended into the fovea in Patient 2. However, the morphology of the macula had been improved in the early postoperative period in both cases.

Because cataract surgery was not performed at the time of the vitrectomy in both cases, the retinal atrophy was most likely not related to the cataract surgical procedures.

Ikeda et al. reported that the length of the ellipsoid zone of the photoreceptors was an important factor for predicting the visual acuities after vitrectomy.<sup>3</sup> However, the EZ in the fovea remained intact outside of macular hole in Case 1 and remained intact in Case 2.

The axial length was about 24 mm in both cases, and the two patients were not highly myopic or hyperopic. The vitreo-retinal surgery was completed without complications by the same surgeon in both cases. The operation time was not extremely long.

ICG has been reported to be toxic for the retina,<sup>4,5</sup> but it was used only in Case 1. Nakazawa et al. reported that the visual field defects detected soon after ICG-assisted ILM peeling for MH surgery continued to worsen for 3 years and not thereafter.<sup>6</sup> The retinal degeneration

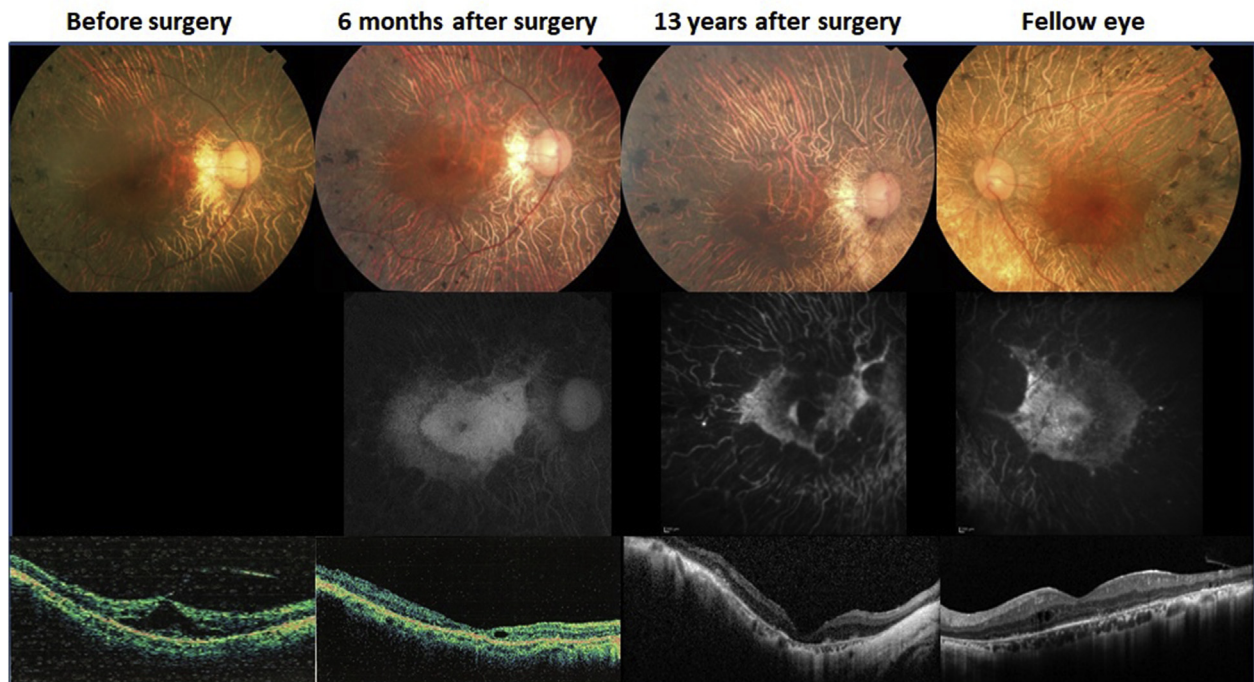


Fig. 2. Images of Case 2 before and after surgery.

Upper: Color fundus photographs. Middle: Fundus autofluorescence images. Lower: Optical coherence tomographic images. The images of fellow eye are 13 years after surgery.

continued even after 3 years in Case 1, however, ICG dye injection cannot be eliminated as one of the causes of the retinal degeneration in this patient.

A reduction of the inner retinal thickness due to ILM peeling has been reported.<sup>7,8</sup> ILM peeling was performed on Case 1, but only the ERM was peeled in Case 2. However, the ILM may have been peeled unintentionally when the ERM was peeled. Therefore, ILM peeling cannot be completely eliminated as the cause of the retinal degeneration in both cases.

Photosensitizing toxicity due to intraocular illumination<sup>9</sup> may have occurred but the illumination source was not placed close to the retina, and no specific area was illuminated for a long period of time during the surgery. Yip et al. reported that intravitreal injection of 1.0 mg/ml ICG may result in photosensitizing toxicity of the RGCs in rats by the illumination.<sup>10</sup>

Thus, there were many factors that may have caused the retinal atrophy in our two cases. But similar vitreous surgical procedures are performed on many patients without RP, and these patients do not develop retinal atrophy. We suggest that there may be some inherent properties of the retina of the RP patients that made them more susceptible to the surgical procedures.

#### 4. Conclusions

An unexpected retinal atrophy can develop near the macular area and occur long after the vitreo-retinal surgery in RP patients. The atrophy can lead to a severe reduction of the BCVA in the postoperative period. The exact reason why the severe retinal degeneration occurred was not conclusively determined but the conditions common to these two cases were surgery using 20-gauge system, SF6 gas tamponade, the same surgeon, and the possibility that ILM was peeled. Long-term careful follow-up is necessary after performing vitreo-retinal surgery on RP patients.

#### Patient consent

Written informed consent was obtained from all of the patients for publication of these case reports and any accompanying images.

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#### Authorship

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

#### Declaration of competing interest

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