

Case Report

## A Case of Retinitis Pigmentosa Diagnosed with Severe Anterior Capsule Contraction after Cataract Surgery

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A 66-year-old woman presented with significant anterior capsule contraction and intraocular lens dislocation in both eyes 4 months after cataract surgery. Postoperative examinations such as fluorescein angiography, Goldmann perimetry, and electroretinography revealed retinitis pigmentosa (RP). Patients with significant anterior capsule contraction after cataract surgery should be closely examined because RP may be a contributing factor.

**Key words:** retinitis pigmentosa, intraocular lens, anterior capsule contraction

**R**etinitis pigmentosa (RP) is an inherited retinal dystrophy characterized by the progressive and extensive degeneration of photoreceptor cells and retinal pigment epithelial cells. The global prevalence of RP is estimated to be approximately 1:4000 [1, 2]. The main symptoms of RP are night blindness, photophobia, progressive peripheral visual field loss, and visual acuity loss. Common findings on ophthalmoscopic examination are peripheral retinal spicule pigmentation, retinal arterial and venous narrowing, and waxy pallor of the optic disc [3].

Lens abnormalities in RP can include posterior subcapsular cataracts [4-6], lens dislocation due to zonular weakness [7-9], anterior capsule contraction after cataract surgery, and intraocular lens (IOL) dislocation [9].

In this report, we describe a case of RP caused by severe anterior lens capsule contraction after cataract surgery.

### Case Presentation

A 66-year-old Japanese woman presented to our hospital complaining of blurred binocular vision. She had no known systemic or ocular illness or family history of retinal disease. She had never had a driver's license. At the time of the initial examination, her right eye and left eye had 20/500 (−5.00 Diopter (D) sphere (sph)) and 20/250 (−6.00 D sph) visual acuity, and 17 mmHg and 14 mmHg intraocular pressure, respectively. No inflammatory cells were observed in the anterior chamber of either eye. Both lenses showed grade II nuclear sclerosis based on the Emery-Little classification with cortical opacity. Ophthalmoscopy revealed no abnormalities in the disc or macula of either eye; however, the peripheral retina could not be adequately examined because of the presence of cataracts. Optical coherence tomography (DRI-OCT Triton; Topcon, Tokyo) showed that the central foveal depression was maintained, but the retinal layers were

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slightly thinned and some irregularities of the ellipsoid zone / interdigitation zone were observed.

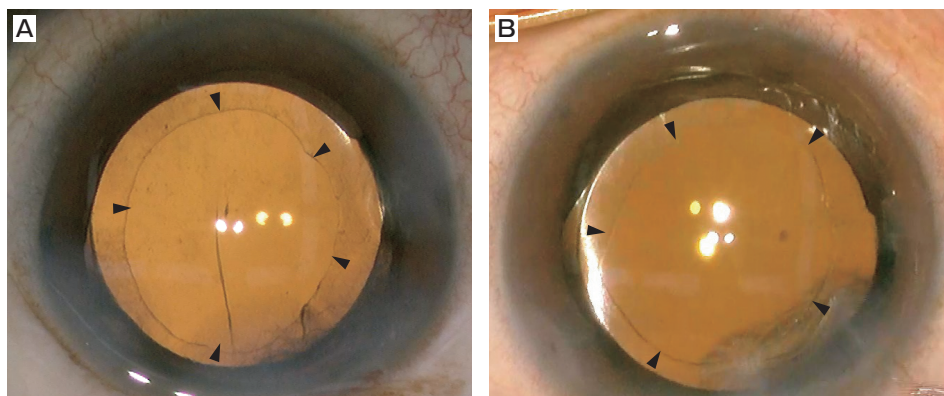
Her symptoms were considered to be due to bilateral cataracts. Phacoemulsification and intraocular lens implantation were performed in both eyes by the same experienced surgeon. At the time of surgery, a continuous curvilinear capsulorhexis measuring 4.5 mm in diameter was created in the anterior capsule of both eyes and an acrylic IOL (CNA0T0; Alcon Laboratories) was inserted in each eye (Fig. 1). The operation times were 15 min for the right eye and 10 min for the left eye, and no posterior capsule rupture, corneal disorders, iris damage or other complications occurred. There were no findings suggestive of zonular weakness, such as lens dislocation or lens dislocation, during surgery.

One month after surgery, the postoperative visual acuity was 20/50 ( $-3.00$  D sph  $-0.50$  D cylinder (cyl)  $\times 180^\circ$ ) in the right eye and 20/40 ( $-2.50$  D sph  $-0.50$  D cyl  $\times 120^\circ$ ) in the left eye. We recommended that she undergo further testing to determine the cause of her poor visual acuity. However, she was satisfied with her postoperative vision and had no symptoms of night blindness or visual field loss. Therefore, a detailed examination for retinal diseases was not performed. At that time, no anterior capsule contraction, IOL dislocation, or intraocular inflammation was observed (Fig. 2).

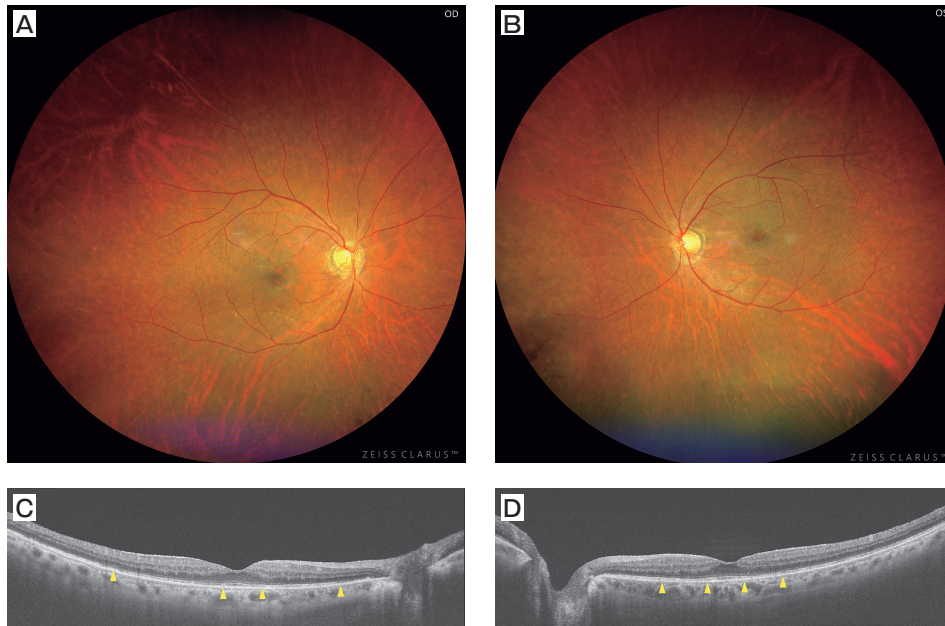
Over the ensuing weeks the patient gradually became aware of vision loss in both eyes, and at 4 months after surgery she returned to our hospital. Her right eye and left eye had 20/250 ( $-3.00$  D sph) and 20/1,000 ( $-2.50$  D sph) visual acuity and 24 mmHg and 22 mmHg intra-

ocular pressure, respectively. The anterior capsule had contracted significantly in both eyes, and the anterior lens capsule margin was 1.3 mm in diameter in the right eye and 0.8 mm in diameter in the left eye. In both eyes, the IOL loops were twisted and deformed, the IOL optics deviated slightly to the inferior nasal side, and ciliary zonules were observed on the superior and temporal sides (Fig. 3). Since the anterior capsule contraction in both eyes was considered the cause of vision loss, we decided to perform a reoperation to improve her vision. Reoperation was performed on the left eye first. We attempted to use a vitreous cutter to incise the contracted anterior capsule edge; however, the anterior capsule was severely fibrotic and could not be treated. Therefore, the IOL enclosed in the lens capsule was extracted, and vitrectomy and intrascleral fixation of the IOL were conducted. As no intraocular inflammation or dislocation was observed 1 month postoperatively, IOL extraction and intrascleral IOL fixation were performed on the right eye. One month after the second reoperation, her visual acuity improved to 20/25 ( $-3.00$  D sph  $-0.50$  D cyl  $\times 115^\circ$ ) in the right eye and 20/25 ( $-2.50$  D sph  $-0.50$  D cyl  $\times 135^\circ$ ) in the left eye, and intraocular pressure was 16 mmHg in the right eye and 17 mmHg in the left eye.

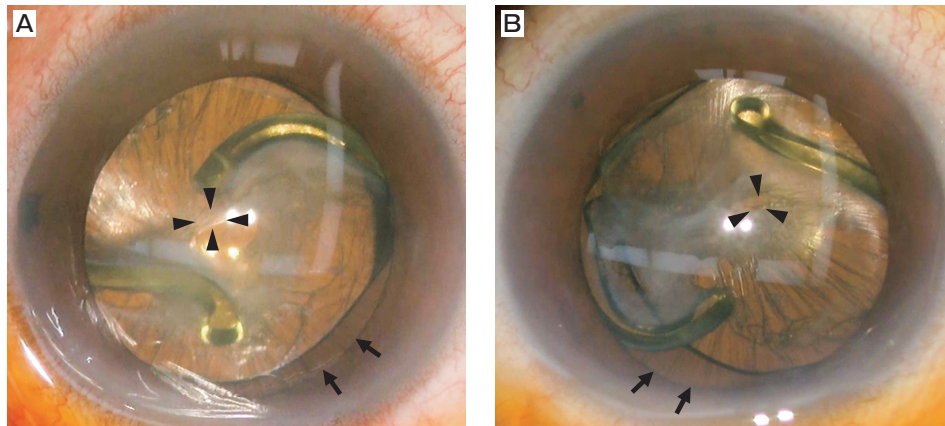
To investigate the cause of the severe anterior capsule contraction, fluorescein angiography was performed. The results showed hyperfluorescence due to window defects scattered throughout all quadrants of the peripheral retina in both eyes, as well as multiple mild leakages from the peripheral retinal blood vessels



**Fig. 1** Anterior segment photographs of both eyes at the end of cataract surgery (intraoperative photos: surgeon's view). A continuous curvilinear capsulorhexis (arrowheads) with a diameter of 4.5 mm was created in both the right (A) and left (B) eyes. An intraocular lens was inserted into the lens capsule of each eye.



**Fig. 2** Fundus photographs and optical coherence tomography images of both eyes 1 month after cataract surgery. There were no abnormalities in the optic disc or macula, retinal pigmentation, or vascular abnormalities in the peripheral retina in either the right (A) or left (B) eye. Optical coherence tomography of the macula showed that the central foveal depression was preserved in both the right (C) and left (D) eyes, but the retinal layers were slightly thinned and some irregularities of the ellipsoid zone / interdigitation zone (arrowheads) were observed.



**Fig. 3** Anterior segment photographs of both eyes 4 months after cataract surgery (intraoperative photos: surgeon's view). The anterior capsule of both the right (A) and left (B) eyes was severely contracted, reducing the diameter of the anterior lens capsule margin to 1.3 mm in the right eye and 0.8 mm in the left eye (arrowheads). In both eyes, the intraocular lens (IOL) loops were deformed in a twisting fashion, the IOL optics was deviated slightly to the inferior nasal side, and ciliary zonules (arrows) were observed on the superior and temporal sides.

(Fig. 4). Visual field testing using Goldmann perimetry showed mild concentric constrictions in both eyes (Fig. 5A, B). In the electroretinogram (ERG) (HE2000; Tomey, Nagoya, Japan), the flash ERG (Dark-adapted 10 ERG) demonstrated decreased a- and b-wave ampli-

tudes in both eyes under the skin electrodes. The 30 Hz flicker ERG and oscillatory potentials showed flat waveforms in both eyes (Fig. 5C-E). Blood tests revealed no abnormalities. She had no known malignant tumors or other systemic illness, and no tumor was detected in the

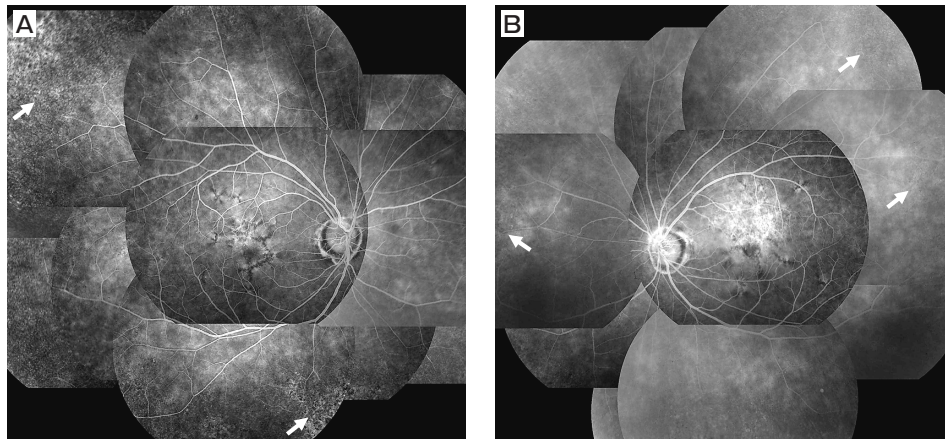


Fig. 4 Fluorescein angiography of both eyes after reoperation. Mild hyperfluorescence due to window defects and leakages (arrows) was observed in all quadrants of the peripheral retina in both the right (A) and left (B) eyes.

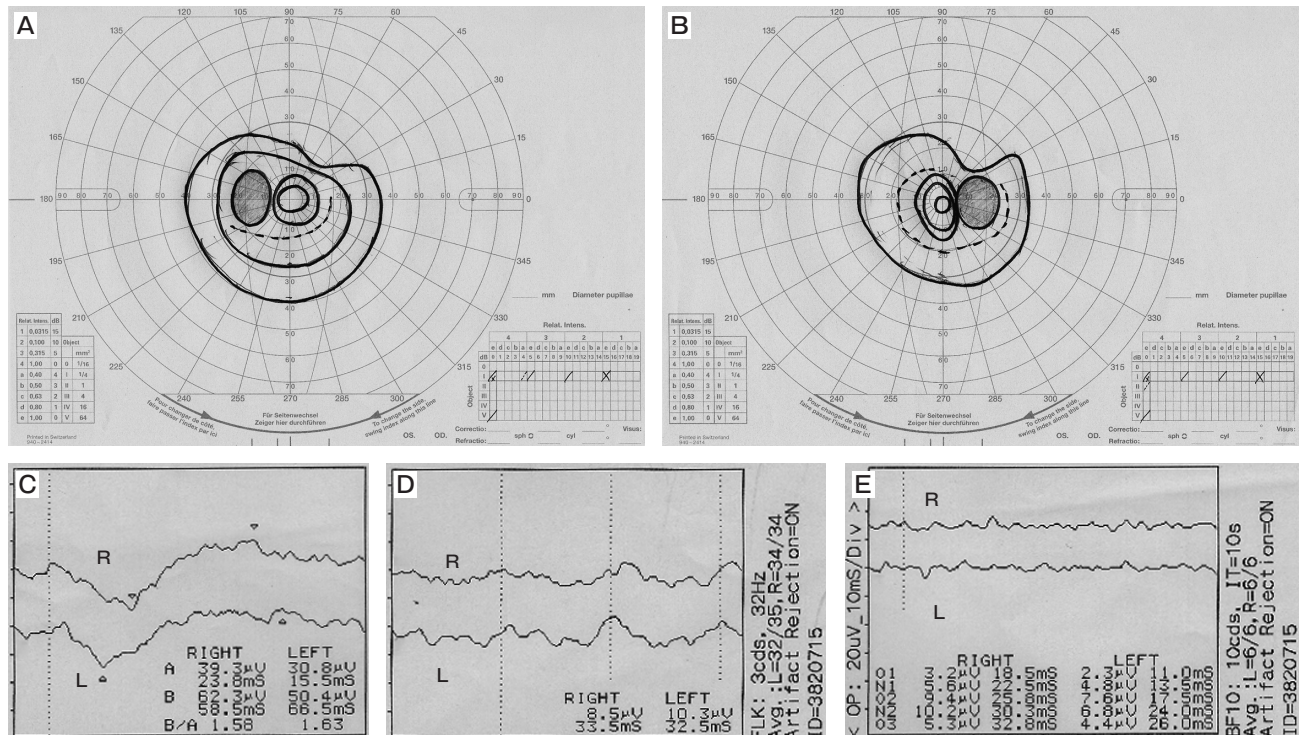


Fig. 5 Goldmann perimetry and electroretinogram of both eyes after reoperation. Goldmann perimetry showed concentric visual field constriction in both the right (A) and left (B) eyes. The dark-adapted flash electroretinogram (ERG) showed decreased a- and b-wave amplitudes in both eyes (C). The 30 Hz flicker ERG (D) and oscillatory potentials (E) showed flat waveforms in both eyes. R, right eye; L, left eye.

examination. Carcinoma associated retinopathy was thus ruled out. Based on these findings, we made a diagnosis of RP.

The patient provided written informed consent for the publication of her case and images.

### Discussion

Anterior capsule contraction is a major postoperative complication of cataract surgery, and is defined as an exaggerated reduction in the margin of the anterior lens

capsule [10-13]. Severe anterior capsule contraction results in IOL dislocation and reduced visual contrast sensitivity, leading to decreased visual function [14,15]. RP, pseudoexfoliation syndrome, uveitis, prior retinal surgery, and diabetes are known risk factors for anterior capsule contraction [9,16-18]. To the best of our knowledge, this is the first published case of RP diagnosed as a result of severe anterior capsule contraction.

There are two possible explanations for the greater likelihood of anterior capsule contraction after cataract surgery in patients with versus without RP. First, compared to healthy individuals, patients with RP have higher levels of specific cytokines that induce the migration, proliferation, and fibrous metaplasia of lens epithelial cells in the anterior chamber [19-21]. This is related to impairment of the blood-retinal and blood-aqueous barriers in the eyes of patients with RP [22-24]. Second, the ciliary zonules of patients with RP are often weakened, reducing their ability to generate centrifugal force [6].

The treatment of anterior capsule contractions in patients with RP depends on the contraction severity. Cases have been reported in which visual improvement was achieved after incision of the constricted anterior capsule margin using capsulorhexis forceps or a Nd:YAG laser [14,15]. However, these treatments cannot be administered in patients with severe anterior capsule fibrosis or IOL dislocation. In the present case, because of the severe anterior capsule contraction and IOL dislocation, we decided that radical treatment was necessary and chose to perform IOL extraction.

In this patient, RP was diagnosed at an early stage, when she was asymptomatic. RP causes progressive loss of vision and the visual field, and historically there has been no effective treatment. In recent years, new gene therapy [25-27] and stem cell [28,29] treatments have been developed. If RP is detected at an early stage, it is now possible to initiate an appropriate early-stage treatment, including a treatment plan tailored to the causative gene and genetic counseling.

In conclusion, we presented a case of retinitis pigmentosa diagnosed as a result of severe anterior capsule contraction after cataract surgery. The presence of severe anterior capsule contraction in both eyes after cataract surgery should be closely examined because RP may be an underlying factor that has not been noted.

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