

Retinitis Pigmentosa Combined with Macular Schisis and Macular Holes in Both Eyes: A Case Report

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Abstract

A 29-year-old woman with an 8-year history of retinitis pigmentosa presented with decreased visual acuity in both eyes. We collected her examinations and diagnosed her with retinitis pigmentosa, macular schisis and lamellar macular holes in both eyes. We proposed the poor condition of retina, caused by retinal pigmentosa, resulted in macular schisis and lamellar macular holes.

Keywords: Retinitis pigmentosa • Macular schisis • Macular hole

Abbreviations: RP: Retinitis Pigmentosa; OCT: Optical Coherence Tomography; ERG: Electroretinography; RPE: Retinal Pigment Epithelium; PPV: Pars Plana Vitrectomy

Introduction

Retinitis pigmentosa is the most common form of hereditary retinal degeneration that causes blindness. Macular schisis generally occurs in patients with a long-term history of myopia. Macular hole usually reflects the low level of the central light sensitivity. Therefore, a patient with these three eye diseases needs long-term follow-up and analysis.

Case Report

In May 2008, a young woman developed abnormal vision in both eyes, especially in the evening. She was physically well and had no history of ocular diseases or injuries. On initial examination, best-corrected visual acuity was 20/50 in the right eye and 20/2000 in the left. Intraocular pressure was 18 mmHg and 15 mmHg in the right and left eyes, respectively. The direct light reflex was normal in both eyes and the relative afferent pupillary defect was negative. Ocular fundus examination revealed scattered pigment particles (bone spicules) throughout the entire retina of both eyes except macula. OCT showed macular schisis and lamellar macular holes in both eyes and neuroepithelial detachments were observed in the left eye. She was diagnosed with Retinitis Pigmentosa (RP), macular schisis and lamellar macular holes in both eyes. In May 2009, OCT (Figure 1) revealed macular schisis and lamellar macular holes in both eyes. We observed macular membrane and neuroepithelial detachment in the left eye. Perimetry (Figure 1) showed tubular vision in both eyes. ERG displayed severely reduced amplitudes, indicating damaged rod and cone cell responses.

In 2010, OCT (Figure 2) showed a crack-like or bridge-like connection in the centre of the macular area, cystoid macular oedema and lamellar macular holes in both eyes. In 2011, OCT (Figure 3) showed the same features. In 2012, Fundus photography (Figure 4) showed scattered pigment particles deposited throughout the retina except macular. OCT revealed macular schisis, lamellar macular holes in two eyes. Until 2015, the patient returned to our hospital and

wanted to improve her poor vision. Best-corrected visual acuity was 20/40 (-1.25/-1.00 × 20) in the right eye and 20/400 (-3.00/-1.00 × 155) in the left eye. Intraocular pressure was at a normal level. Ophthalmoscopy revealed macular schisis and lamellar macular holes in both eyes with retinal detachment in the periphery of the macula. Moreover, ocular fundus examination showed the same results as those seen previously. OCT (Figure 5) also showed macular schisis and lamellar macular holes. Ultrasonography revealed vitreous opacities.

Discussion

RP is a common form of hereditary retinal degenerative disease that manifests as the gradual loss of function in photoreceptor cells and retinal pigment epithelial cells [1]. The prevalence of the disease is approximately 1/3000, and there are no significant gender differences; moreover, this disease is the leading cause of blindness worldwide [2]. The majority of cases (50-60%) are autosomal recessive, sporadic or simplex. Besides, families with autosomal dominant (30-40%) and X-linked inheritance (5-20%) are frequently observed. Maternal (mitochondrial) inheritance is very rare in RP [3]. Treatment for this disease is still unknown. The patient was diagnosed with typical RP, but we had no condition to test her genes. Recent study showed macular schisis generally attributed to traction in the retina and posterior pole [4]. The lamellar macular holes in both eyes of our patient may be the result of a complication of long-term traction. In 2010, we observed a small and convex triangle under the bridge-like connection in OCT of the right eye. This finding was unchanged in 2015. Theodossiadis [5] also observed a patient with macular schisis revealed a triangle-like full-thickness hole under an inner crack-like connection in OCT. Besides, Matsumura [6] had reported two patients who had macular holes and macular schisis, and proposed the tangential traction could be transferred through a bridge-like connection, inducing the development of macular holes, as well as partial Posterior Vitreous Detachment (PVD). We speculate that macular schisis weakens connections between the Retinal Pigment Epithelium (RPE) and neuroepithelial layer, resulting in the formation of retinal detachment and macular hole.

Many researchers believe that the initial damaged site in RP is RPE layer. The damaged RPE layer can result in loss of nutrients in the photoreceptor cell layer and gradual degeneration, which gives rise to retinal dysfunction. Testa F [7] showed that the frequency of patients with full-thickness or lamellar macular holes was nearly 1% among 581 RP patients. Professor Ryan B Rush [8] has performed PPV and intravitreal gas infusion surgery on an RP patient with lamellar macular holes in both eyes. After 6 months, visual acuity was significantly improved. They suggested the potential mechanism underlying RP with bilateral lamellar macular holes is as follows: contraction or rupture of

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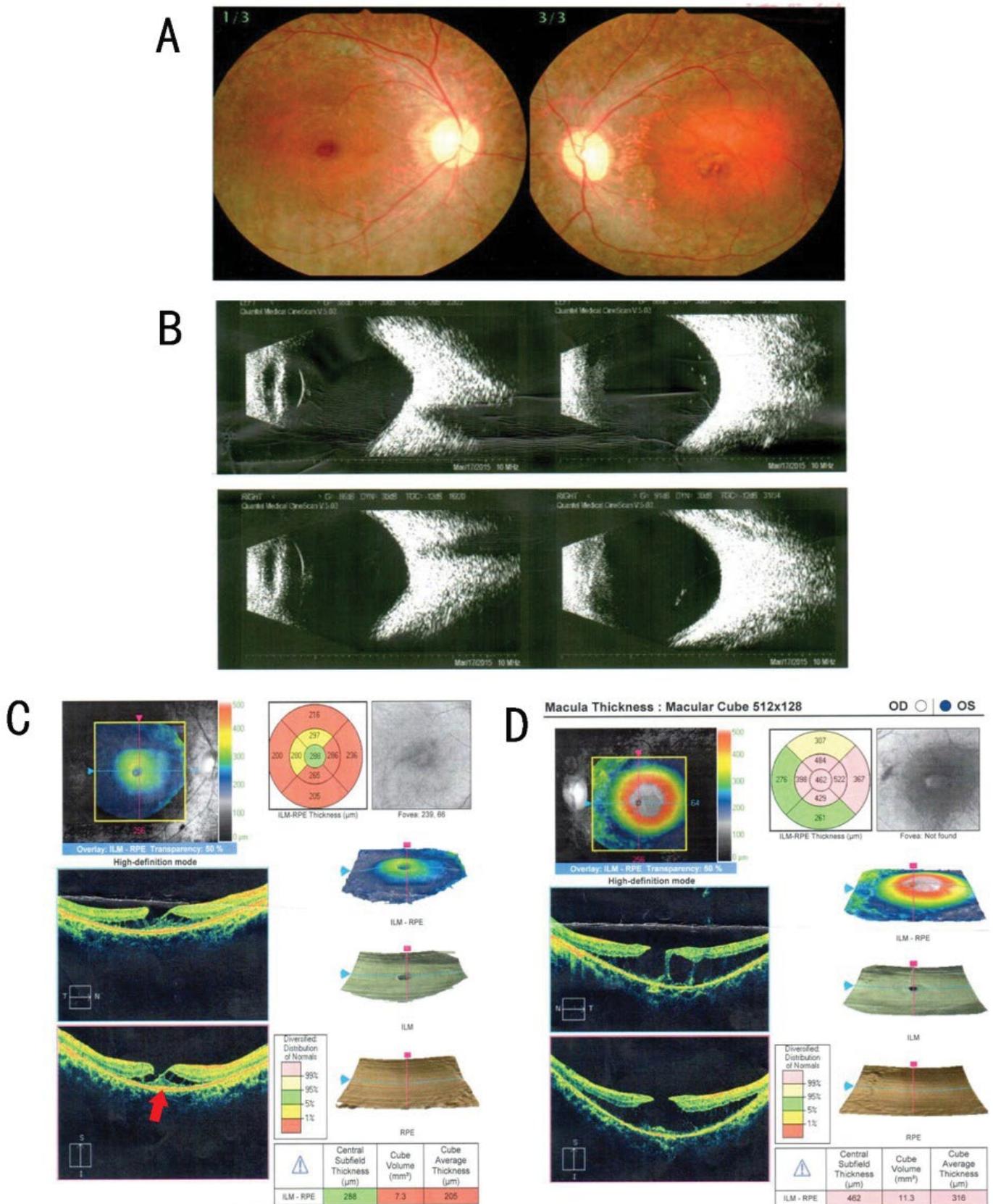


Figure 1. In 2009, OCT showed macular schisis and lamellar macular holes in the left eye (A) and in the right eye (B), and neuroepithelial detachment in the left eye(B). Perimetry showed tubular vision in the right eye (C) and on the temporal side of the left eye (D). ERG showed severe reduced amplitude (E and F).

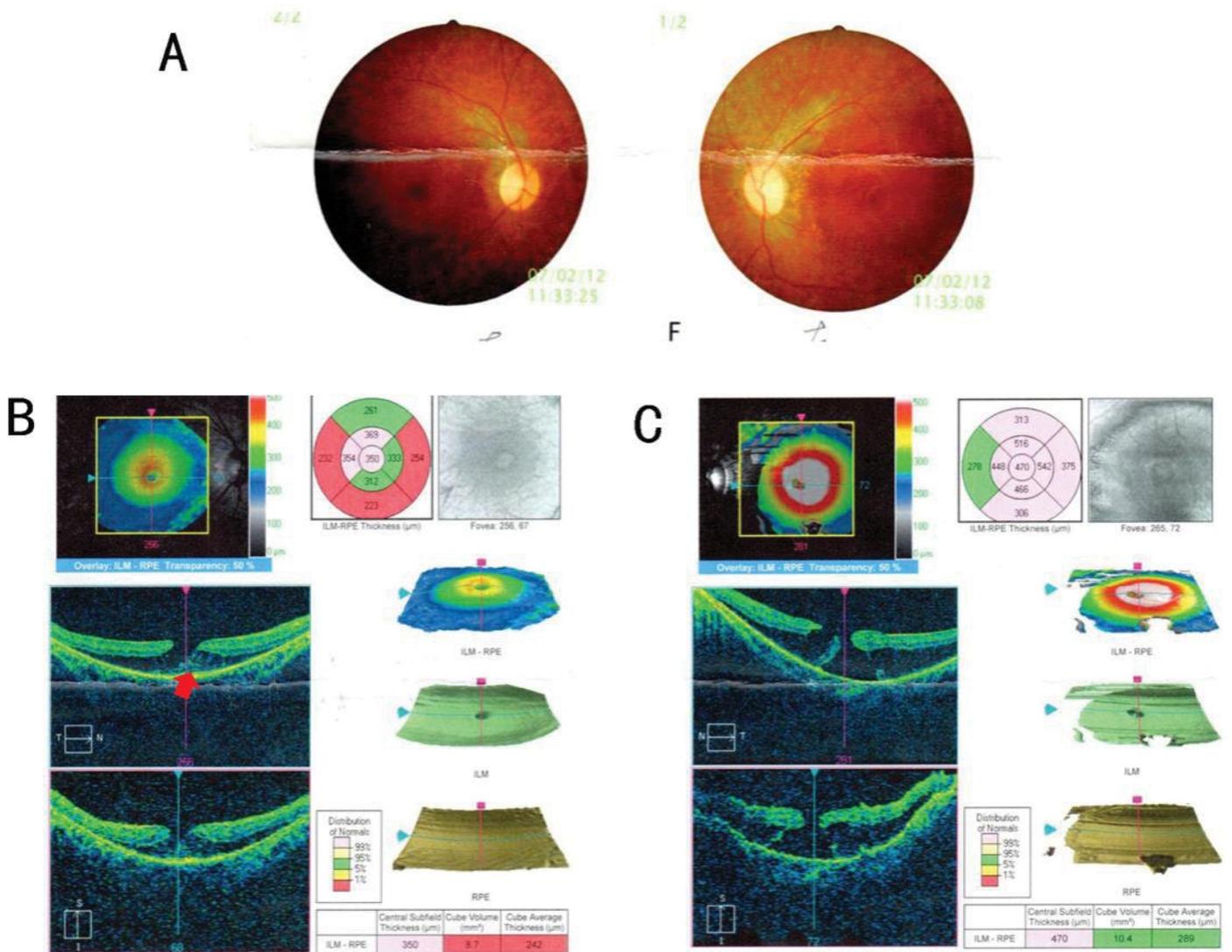


Figure 2. In 2010, OCT showed a crack-like or bridge-like connection on the centre of macular area in both eyes (A and B) and lamellar macular holes in the right eye (A) and in the left eye (B).

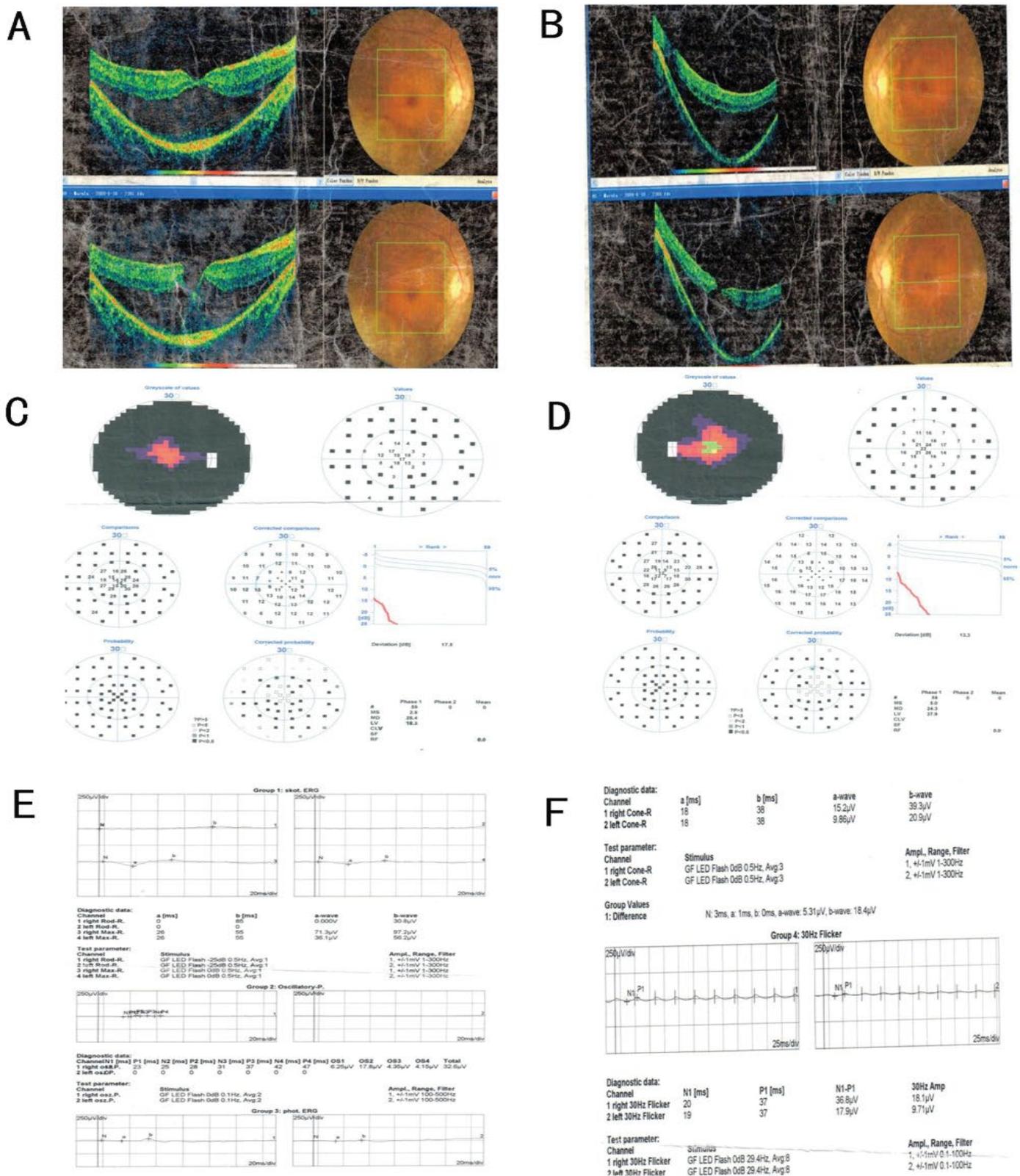


Figure 3. In 2011, OCT showed macular schisis in both eyes (A and B); lamellar macular holes in the right eye (A), and in the left eye (B).

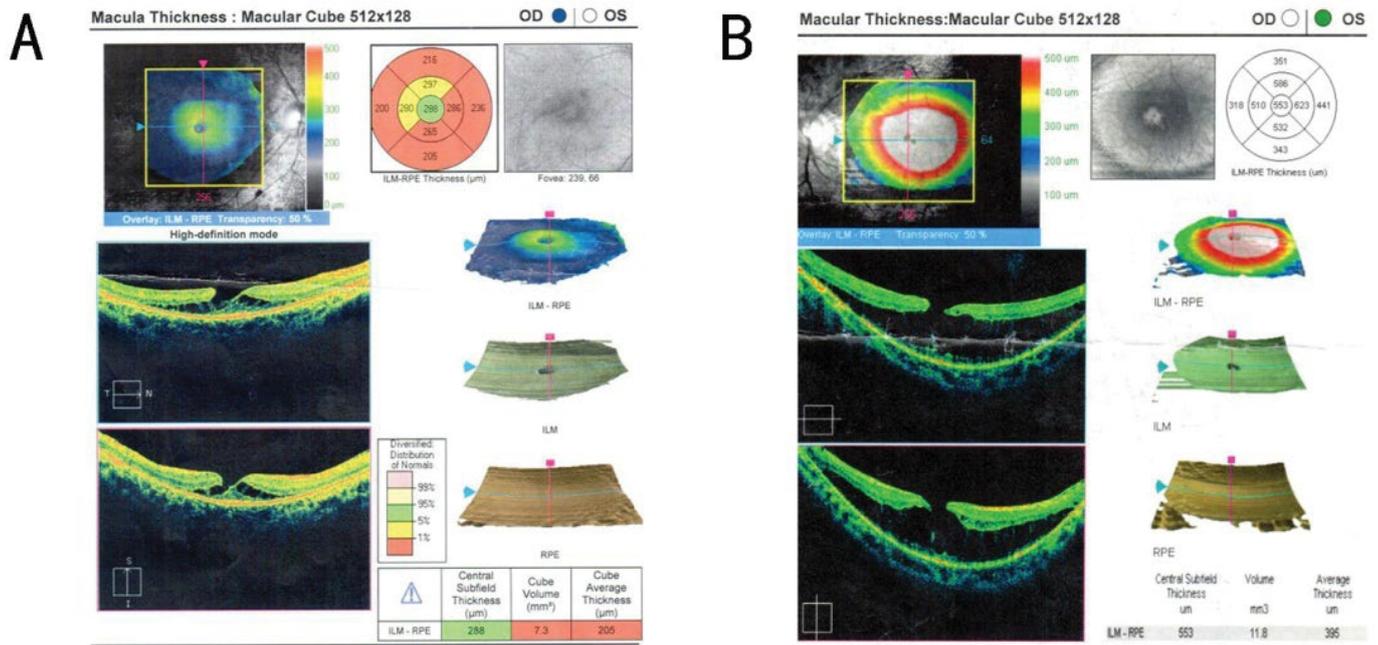


Figure 4. In 2012, Fundus photography showed scattered pigment particles were deposited in the retina (A). OCT showed macular schisis in both eyes (A and B), lamellar macular holes in two eyes (B and C). A convex triangle was present in the macular centre (B).

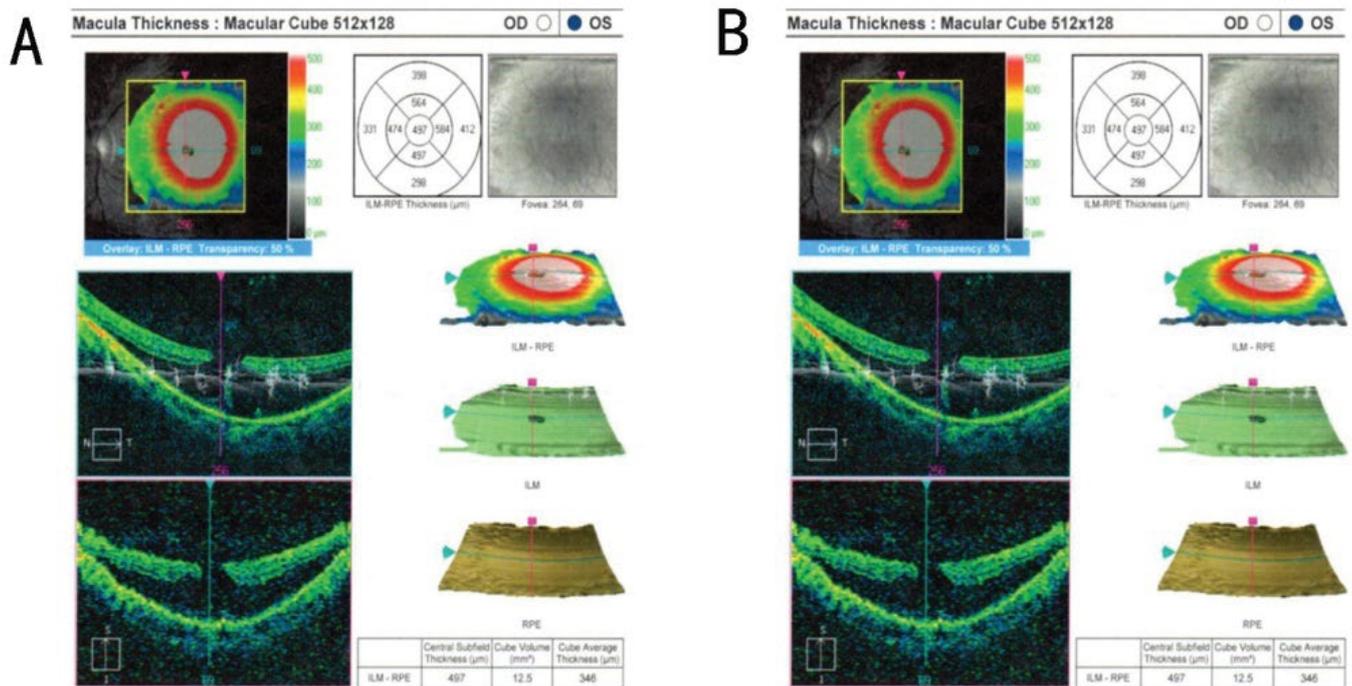


Figure 5. In 2015, Fundus photography showed scattered pigment particles were deposited in the retina (A). Ultrasonography showed vitreous opacities (B). OCT showed a crack-like or bridge-like connection on the centre of macular area and macular holes in both eyes (C and D). A convex triangle was present in the macular centre (C).

an epiretinal membrane or fusion of cysts from RP-associated cystoid macular oedema, inflammatory reactions, RPE and vitreous traction. Besides, Macular schisis involves interlayer separation which happens in the inner plexiform layer and outer plexiform layer, forming one or more non-reflective optical spaces, which leads to macular holes. So, RP and macular schisis have the potential to induce macular holes. Furthermore, Professor Kaori Amemiya [9] had an RP patient with a macular hole in the left eye at 18-year-old. After PPV surgery and removal of a thick, macular retinal membrane around the hole, the patient's vision improved from 20/100 to 20/28. Histopathology revealed that macrophages, Müller cells, glial cells and fibroblasts were present in the removed membrane, which due to vitreous degeneration. Retinitis pigmentosa was the main cause to retina and vitreous degeneration.

Conclusion

We propose that retinitis pigmentosa impacts the state of the whole retina and induces macular schisis and lamellar holes. In the poor state of retinal function, the three diseases interacted, eventually leading to a serious decline in vision. Our patient was treated for 6 years, and her vision remained stable. We suppose:

- RP did not damage the macular area or damaged it slowly;
- RP and macular holes slowly developed and remained stable;
- Due to the lack of myopia, macular schisis remained stable.

Ethical Statement

In this research, we complied with guidelines for human studies in World Medical Association Declaration of Helsinki. Firstly, due to “the health of my patient will be my first consideration”, we followed this patient for 8 years and hoped to promote her vision. Secondly, in WMA, “medical research is subjected to ethical standards that promote and ensure respect for all human subjects and protect their health and rights”, we collected all her examinations but still could not find the main reason about her disease. Our purpose is to discuss with other specialists about this kind of disease. Before we got her information to do research, we have asked for her consent. Unfortunately, we did not sign an informed consent. Because this patient was sad about the final result (her vision will not be improved) and she did not come again. When we want to submit this article, we could not contact her. But we are sure that she agreed to give us information and her private information will not present in this article.

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