

Case Report

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# Retinal Pigment Epithelial Detachment Associated with Immunoglobulin A Nephropathy: A Case Report

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## Keywords

Pigment epithelial detachment · IgA nephropathy · Pulse steroid therapy · Retinal pigment epithelium tear

## Abstract

Uveitis and scleritis are eye diseases associated with immunoglobulin A (IgA) nephropathy, but reports on retinal pigment epithelial detachment (PED) in relation to IgA nephropathy are scarce. We have experienced a case of PED associated with IgA nephropathy that was improved by pulse steroid treatment. A 68-year-old woman underwent examination for visual loss in the right eye. Her corrected visual acuity was 20/20 on both sides, and serous PED was observed in both eyes. One month later, the PED improved in both eyes but recurred 3 months later. Results of blood examination raised suspicion of IgA nephropathy, and she was referred to a nephrologist. Two weeks later, the PED in both eyes worsened, and a retinal pigment epithelium (RPE) tear appeared in the right eye. A sub-Tenon's injection of triamcinolone acetonide was performed to address the PED, but it was not effective; thus, pulse steroid therapy was performed twice. The PED disappeared from both eyes, and the visual acuity in her left eye was maintained at 20/20, but it decreased to 20/200 in her right eye due to macular atrophy after the RPE tear. The PED had not recurred despite having no improvement in renal function. In conclusion, in IgA nephropathy, deposition of immune complexes on the RPE causes its inflammation, which may lead to PED. In cases of unexplained PED, the possibility of a systemic disease as the cause should be considered.

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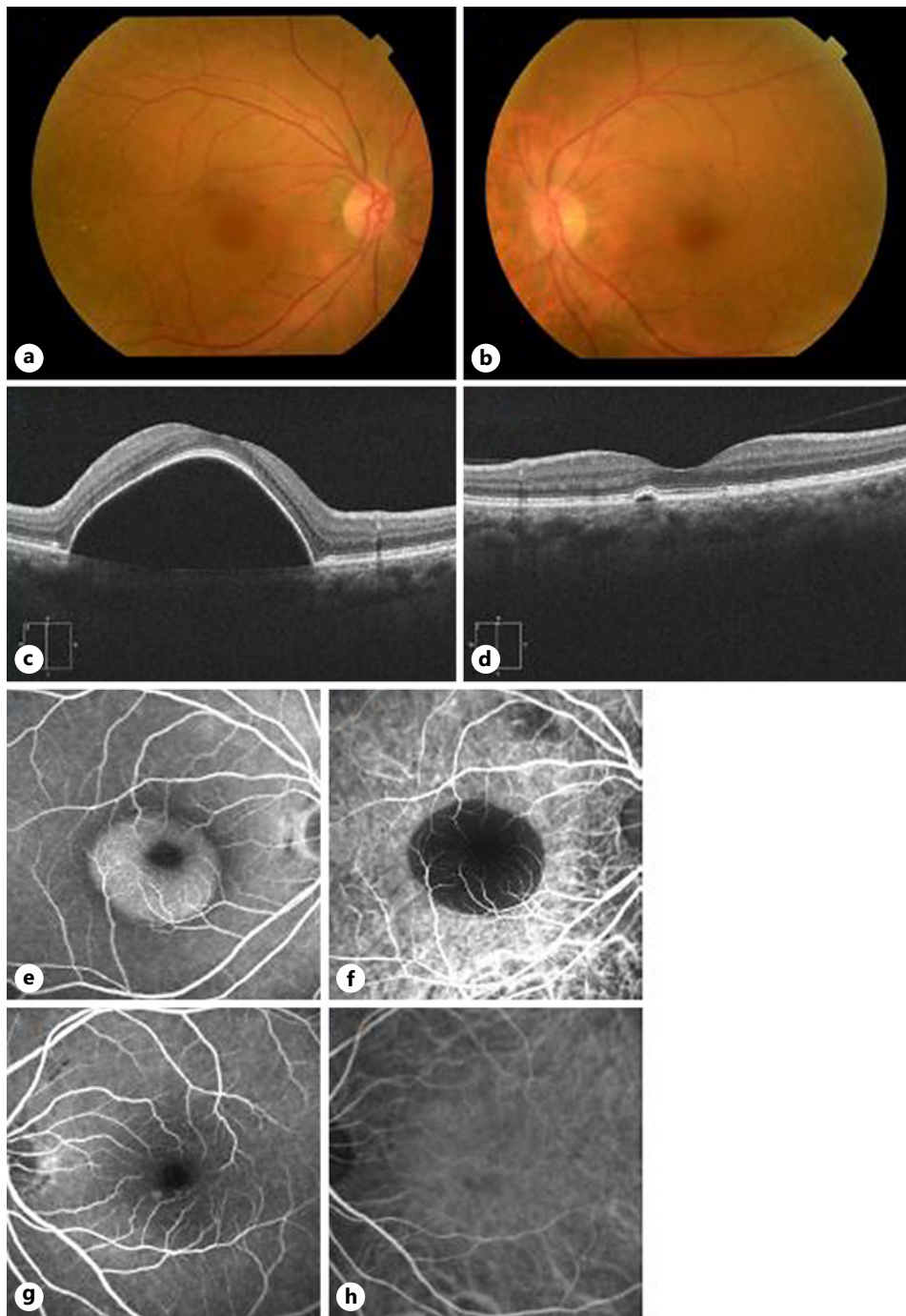
## Introduction

Immunoglobulin A (IgA) nephropathy is the most common chronic glomerulonephritis worldwide, and it has clinical findings of hematuria and proteinuria [1, 2]. In general, they are often asymptomatic and are often found by accident during school and workplace examinations. This disease affects relatively young individuals (teens as well as adults ~20 years old at the time of discovery), but it can occur in all ages. Treatment for IgA nephropathy mainly focuses on lifestyle regulation and diet, and for patients with hypertension, their blood pressure should be controlled with antihypertensive drugs. Corticosteroids are also effective for maintaining and improving renal function, and the combination therapy of tonsillectomy with pulse steroid therapy has been performed in patients with IgA nephropathy in Japan [3].

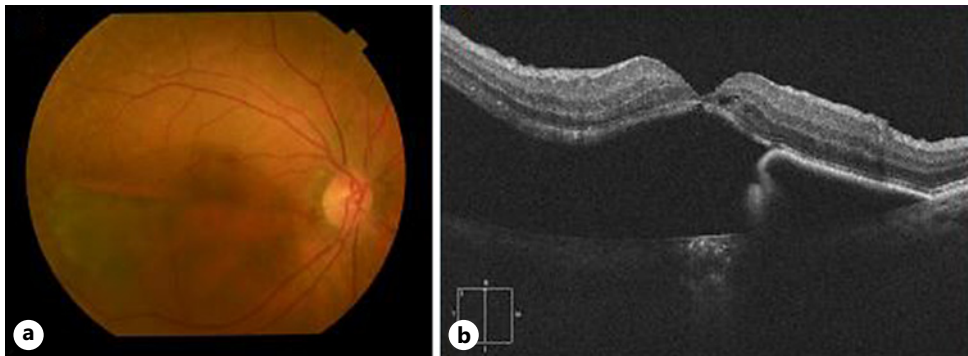
Uveitis [4] and scleritis [5] are eye diseases associated with IgA nephropathy, but reports on serous retinal detachment are scarce [6–8]. We herein report a case of serous retinal detachment associated with retinal pigment epithelial detachment (PED) in both eyes that was related to IgA nephropathy and that disappeared with pulse steroid therapy.

## Case Report

A 68-year-old woman consulted an ophthalmologist with chief complaints of central scotoma and poor vision in the right eye. She had high blood pressure. At the first consultation, her corrected visual acuity of both eyes was 20/20, and the intraocular pressures of the right and left eyes were 20 and 17 mm Hg, respectively, by noncontact tonometer. No inflammation was observed in the anterior eye or in the optic media. A well-defined retinal pigment epithelial (RPE) elevation lesion in the macula and some hard drusens near the temporal arcade blood vessels were observed in the fundus examination of the right eye, and a small lesion of pigment abnormality was observed on the inferior of fovea of the left eye (Fig. 1a, b). Optical coherence tomography revealed a raised dome-shaped PED in the right eye and a small elevation of the RPE on the inferior of fovea in the left eye (Fig. 1c, d). Fluorescein angiography (FA) showed a pooling of fluorescein consistent with the domed lesion of optical coherence tomography in the right eye (Fig. 1e). No abnormal findings suggestive of pigment epitheliopathy, such as clear leakage from abnormal blood vessels or window defects, were found in the peripheral part of the retina in FA. Indocyanine green angiography revealed a consistently hypofluorescent lesion, indicating PED, in the right eye (Fig. 1f). No hyperfluorescence indicating choroidal neovascularization was observed. In the left eye, spot fluorescence retentions were observed slightly below the macula in the FA (Fig. 1g). Indocyanine green angiography of the left eye revealed a blockage of fluorescence, probably by the small PED, but it was not as pronounced as that in the right eye (Fig. 1h). Blood pressure at this point was 172/73 mm Hg, which was a high blood pressure. Based on these clinical findings, age-related macular degeneration, central serous chorioretinopathy, multifocal posterior pigment epitheliopathy were excluded, but the definitive diagnosis was unclear. One month later, the PED in both eyes became smaller, albeit remained. Three months later, PED recurred in both eyes, and visual acuity was reduced. The corrected visual acuity of both eyes was 20/50. A blood examination revealed a high level of IgA, mild renal dysfunction (creatinine, 1.65 mg/dL; blood urea nitrogen [BUN], 31 mg/dL), and hypoalbuminemia (albumin 2.7 g/dL). Two weeks later, PED worsened in both eyes, and an RPE tear appeared in the right eye (Fig. 2). Because central serous chorioretinopathy and multifocal posterior pigment epitheliopathy were excluded and PED was accompanied by inflammation, sub-Tenon's injection of triamcinolone acetonide (STTA) was performed on both eyes to treat the inflammation. She was referred to the renal department for renal dysfunction, and



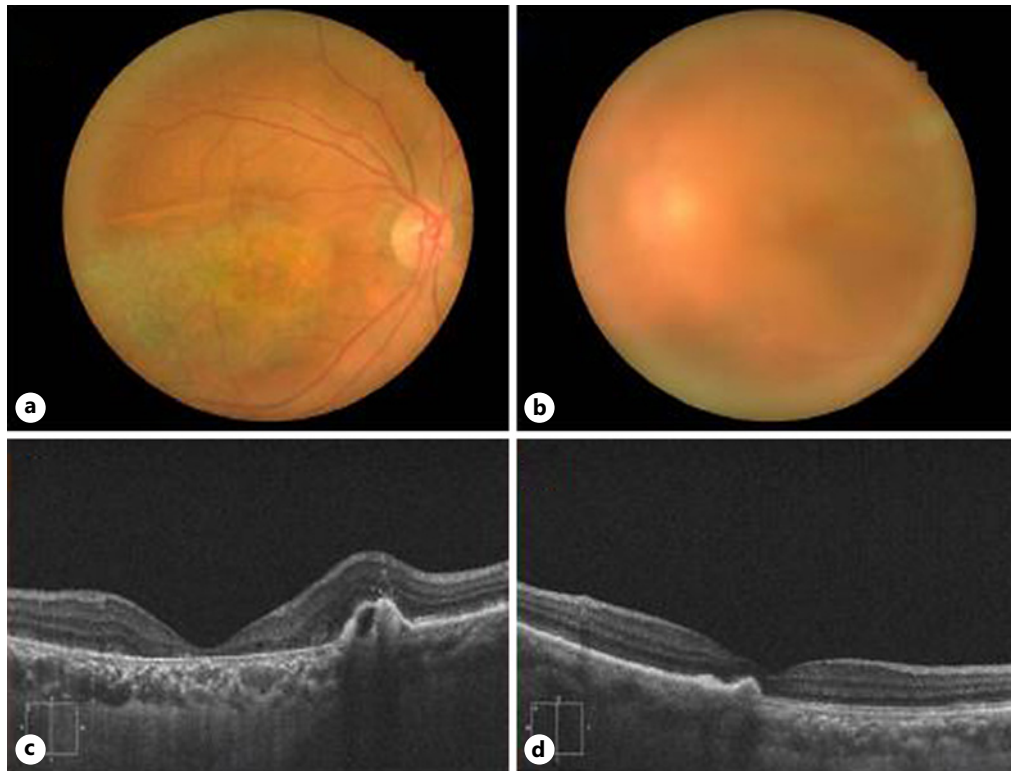
**Fig. 1.** Findings at first consultation. **a** Fundus photograph of the right eye. Drusen was found around the ear arcade, and a prominent lesion was seen in the macula. **b** Optical coherence tomography (OCT) image of the right eye. A dome-shaped raised pigment epithelial detachment (PED) was observed. **c** Fundus photograph of the left eye. Drusen was observed around the arcade. **d** OCT image of the left eye. A small PED was observed on the inferior of fovea. **e** Fluorescein angiography (FA) image of the right eye (late). **f** FA image of the left eye (early). **g** Indocyanine green angiography (IGA) image of the right eye (early). The fluorescence block, indicating PED, is recognized. **h** IGA image of the left eye (late).



**Fig. 2.** Progress after first consultation (4.5 months after the first visit). **a** Fundus photograph of the right eye. Tear formation of the retinal pigment epithelium (RPE) is observed. **b** Optical coherence tomography image of the right eye. In the RPE tear, the RPE has rolled up.

medications, including angiotensin II receptor blocker, were started. Blood pressure was as high as 170/72 mm Hg, and plasma albumin level remained low at 2.7 g/dL. Two weeks later, the PED in the right eye shrank, but visual acuity further decreased to 20/100. Meanwhile, the PED in the left eye disappeared, and visual acuity improved to 20/50. Kidney biopsy revealed a significant deposition of IgA and complement C3 in the mesangial region, and she was diagnosed with IgA nephropathy. Serum complement titers were normal (CH50, 39.5 U/mL; C3, 80.2 mg/dL; C4, 22.5 mg/dL). Thereafter, she underwent lifestyle modification/improvement and diet therapy and took antihypertensive and anticoagulant medications to address the IgA nephropathy. However, 3 months later, PED recurred in the left eye despite improvement of visual acuity to 20/32, and a second STTA was performed on the left eye. One month later, the PED and serous retinal fluid (SRF) in the left eye exacerbated, and cystoid edema appeared in the parafovea in the right eye. Blood and urine tests showed worsening renal function (creatinine, 2.77 mg/dL; BUN, 48 mg/dL; estimated glomerular filtration rate, 14.2 mL/min/1.73 m<sup>2</sup>; urine protein, 878.0 mg/dL; red blood cell count >100/high-power field), which was believed to be associated with worsening retinal exudative lesions in both eyes. Therefore, pulse steroid therapy (500 mg/day Solu-Medrol for 3 days; Pfizer, NY in the USA) was given to enhance the treatment of IgA nephropathy. The systolic blood pressure before the steroid pulse was 140–150 mm Hg, and plasma albumin level was 2.8 g/dL.

After two pulse steroid treatments, PED and SRF in both eyes improved (Fig. 3), but renal function did not considerably improve (creatinine, 2.79 mg/dL; BUN, 65 mg/dL; estimated glomerular filtration rate, 14.1 mL/min/1.73 m<sup>2</sup>; urine protein, 310.7 mg/dL [2+]; red blood cell count, 10–19/high-power field [2+]). Plasma albumin level improved slightly to 3.3 g/dL. Systolic blood pressure remained unchanged at 140–150 mm Hg. Corrected vision was 20/100 for the right eye and 20/66 for the left eye. Over the following year, steroid use was gradually reduced and discontinued. At the end of steroids, renal function was not improved, but plasma albumin levels increased to 3.9 g/dL and blood pressure decreased to 120/54 mm Hg. Approximately 6 years after onset, PED in the right eye has no recurrence, and the corrected vision is 20/200 (Fig. 4). There was no recurrence of PED in the left eye. Visual acuity was reduced by steroid-induced cataract, but it recovered to 20/16 after cataract surgery in the left eye.

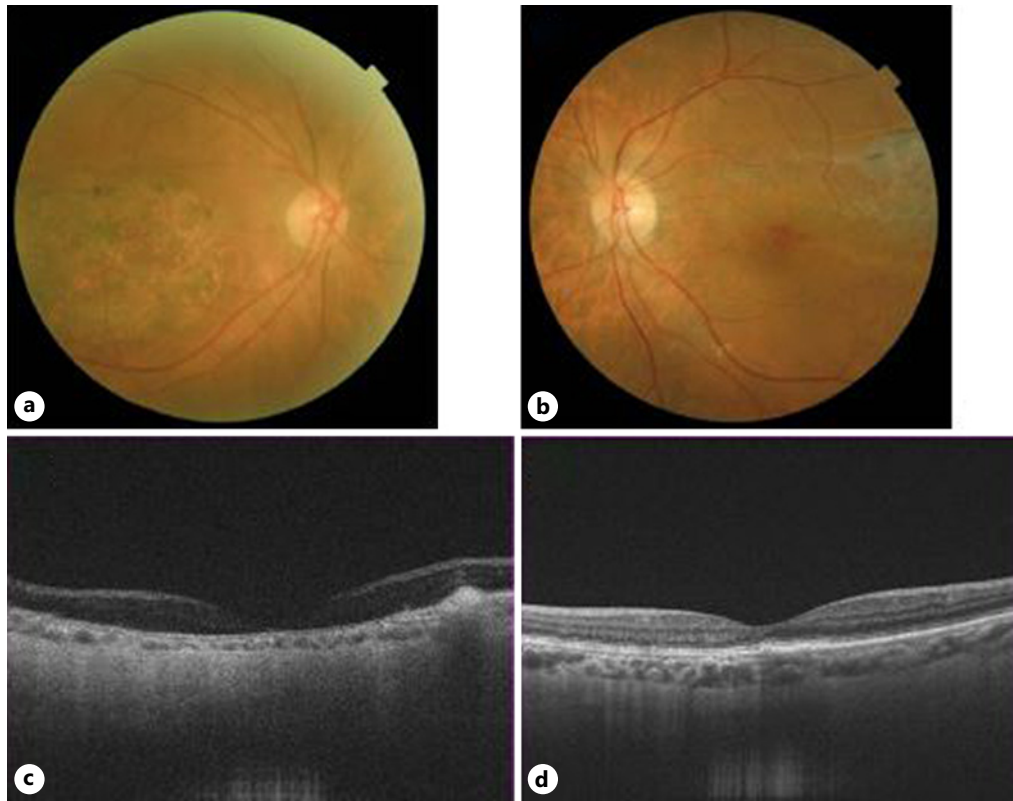


**Fig. 3.** Progress after pulse steroid therapy (1 year and 2 months after the first visit). **a** Fundus photograph of the right eye. The macula had extensive atrophic lesions. **b** Fundus photograph of the left eye. This is unclear because of steroid-induced cataract, but atrophic lesions are seen on the ear side. **c** Optical coherence tomography (OCT) image of the right eye. The ellipsoid zone in the macula has disappeared, and the elevated RPE has increased. **d** OCT image of the left eye.

### Discussion

IgA nephropathy may accompany hematuria after upper respiratory tract or gastrointestinal infections and is believed to be associated with mucosal immunity. Although serum IgA may be elevated, a definitive diagnosis of IgA nephropathy requires renal biopsy to show cell proliferation in the mesangial region of the glomeruli and deposition of abnormally glycosylated IgA. In addition, since abnormal deposition of IgG and complement component C3 is recognized together with abnormal glycan, the pathogenesis of IgA nephropathy may be due to the deposition of IgA-type immune complexes on the glomeruli and the resulting inflammatory changes in glomerular tissue. Uveitis is a common ocular disease associated with IgA nephropathy [9], and other inflammatory eye diseases such as scleritis, episcleritis, and retinal vasculitis have been reported as well [10]. Cetin et al. [11] also reported cases of drusen-like deposits in the retina of children with IgA nephropathy. They suggested that IgA and C3 immune complexes could be deposited not only in the mesangial region but also between the RPE and the Bruch membrane, which has a structure that is very similar to that of the glomerular basement membrane [11].

There were few reports of raised PED associated with IgA nephropathy, and the cause was unknown. In this case, dome-shaped serous PED was observed in both eyes, and slight drusen was observed. PED in this case was considered to be caused by inflammation of the IgA and C3 immune complexes deposited between the RPE and the Bruch membrane and, as a result,



**Fig. 4.** Latest progress (5 years and 10 months after the first visit). **a** Fundus photograph of the right eye. **b** Fundus photograph of the left eye after cataract surgery. **c** Optical coherence tomography (OCT) image of the right eye. The height of the ridge is lower. **d** OCT image of the left eye.

disappeared with pulse steroid therapy for IgA nephropathy. The pulse steroid was effective because it may have suppressed the inflammation of the RPE by the immune complex more strongly than STTA. It is also possible that the disruption of water balance between hydrostatic pressure and oncotic pressure in the choroid was because of renal dysfunction, resulting in increased interstitial fluid in the choroid and large accumulation under the RPE [12, 13]. In this case, PED and SRF worsened when the blood pressure was high and improved as the blood pressure decreased. There are some reports of PED and an RPE tear with hypertensive choroidopathy secondary to systemic hypertensive diseases such as pregnancy, renal failure, pheochromocytoma, and malignant hypertension [14–16]. In this case as well, fluctuations in blood pressure may have been involved in the exacerbation or improvement of PED and SRF. Although the albumin level eventually increased, it remained low despite the improvement of PED and SRF, so the association with hypoalbuminemia was considered to be low. Furthermore, lipids accumulate under the RPE due to aging, and the adhesion between the RPE and the Bruch membrane decreases with age [17, 18]. These histological changes in the RPE may have also contributed to PED.

In addition, as a result of the formation of the RPE tear in the right eye, the RPE in the macula was extensively atrophied, resulting in poor vision. Although the mechanism of the RPE tear formation is not well understood, the RPE tear is often seen in age-related macular degeneration in patients with choroidal neovascularization following treatments such as retinal photocoagulation, photodynamic therapy, and administration of anti-vascular endothelial growth factor drugs [19–23]. In this case, there was no previous treatment, as

described above, but it was considered that the RPE was prolonged because of a large amount of liquid stored under the RPE, resulting in a tear. Alternatively, we speculate that there was local fragility in the RPE and Bruch membrane due to deposition and inflammation of IgA and C3 immune complexes. When the tear is formed, the RPE is deficient and atrophied, resulting in retinochoroid atrophy. The risk of the RPE tear formation should be considered in the event of rapid PED increase.

In this case, pulse steroid therapy was effective for PED that may have been associated with IgA nephropathy. In cases with unexplained PED, the possibility of a systemic disease as the cause should be considered.

## Acknowledgments

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## Statement of Ethics

The written informed consent was obtained from the patient for publication of this case report and any accompanying images. This study protocol was exempted from the need for approval by the Institutional Review Board of Yamaguchi University Hospital.

## Conflict of Interest Statement

The authors declare no conflict of interests.

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## Author Contributions

Ayano Sakuma designed the study and wrote the initial draft of the manuscript. Tadahiko Ogata, Makiko Wakuta, and Tomoko Orita have contributed to data collection and interpretation and critically reviewed the manuscript. Kazuhiro Kimura was the corresponding author, contributed to the analysis and interpretation of data, and assisted in the preparation of the manuscript. All the authors read and approved the final manuscript.

## Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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