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Case report

Retinal detachment with a break at pars plicata associated with congenital malformation of the lens–zonule–ciliary body complex

Fang-Yi Tsai [a], Ling-Ing Lau [a, b, *], Shih-Jen Chen [a, b], Fenq-Lih Lee [a, b]

[a] Department of Ophthalmology, Taipei Veterans General Hospital, Taipei, Taiwan
[b] Department of Ophthalmology, School of Medicine, National Yang-Ming University, Taipei, Taiwan

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ABSTRACT

Retinal detachment with a break at the pars plicata associated with congenital malformation of lens–zonule–ciliary body complex is rare; most reports are of young Japanese male patients with atopic dermatitis. The present case report is the first to describe the condition in a Chinese patient with no atopic dermatitis or trauma history. A 22-year-old male presented with blurred vision in the left eye for 4 months. Fundus examination revealed shallow lower temporal retinal detachment. Further examination with scleral indentation under maximal pupil dilatation identified a break at the far periphery beyond the ora serrata and pars plana. Gonioscopy revealed a pars plicata break at the nonpigmented ciliary epithelium associated with congenital ciliary process hypoplasia and subtle lens defect at the same meridian. The retina was successfully reattached after segmental scleral buckling, cryopexy, and laser photocoagulation.

1. Introduction

Retinal detachment with a break at the pars plicata was first described in 1953 in a case report of perforating trauma.[1] A multicenter study showed that 4.8% of retinal detachment in patients with atopic dermatitis was caused by breaks at the pars plicata.[2] Retinal detachment with a pars plicata break associated with lens coloboma and adjacent hypoplastic ciliary processes is rarely reported, and most reports are of Japanese patients with a history of atopic dermatitis.[3,4] The present case report describes unilateral shallow retinal detachment with a break at the pars plicata and associated congenital malformation of lens coloboma and rudimentary ciliary process without atopic dermatitis in a Chinese patient. The retina was reattached after performing segmental scleral buckling, cryopexy, and laser photocoagulation.

2. Case Report

A 22-year-old Chinese male developed progressive blurred vision in the left eye during 4 months. He was previously examined by several ophthalmologists without a definite diagnosis and was referred to our clinic for further evaluation and management. Upon examination, his best-corrected visual acuity was 6/6 in the right eye and 3/60 in the left eye. The refractive error was −7.5 to 0.5 × 180 in the right eye and −8.0 to 1.75 × 170 in the left eye. He had no history of previous ocular trauma or systemic disease including atopic dermatitis. Slit-lamp microscopy showed clear lens and silent anterior chamber bilaterally. Binocular ophthalmoscopy of the left eye showed a shallow retinal detachment at the temporal lower quadrant in the 2 to 5 o'clock meridian with macular involvement but without a definite retinal break (Fig. 1). Fluorescein angiography showed a silent optic disc and macula without any sign of exudative retinal detachment. Optical coherence tomography also revealed a detached neurosensory retina from the retinal pigment epithelium at the macula (Fig. 2A). A detailed retinal binocular examination with the contact lens under microscopy also failed to demonstrate a retinal break to the ora serrata.

After an extensive discussion with the patient, a segmental scleral buckling was recommended. Intraoperatively, the scleral indentation revealed a break beyond the ora serrata and pars plana

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* Corresponding author. Department of Ophthalmology, Taipei Veterans General Hospital, Number 201, Section 2, Shih-Pai Road, Taipei, Taiwan.
E-mail address: leliew@vghtpe.gov.tw (L.-I. Lau).

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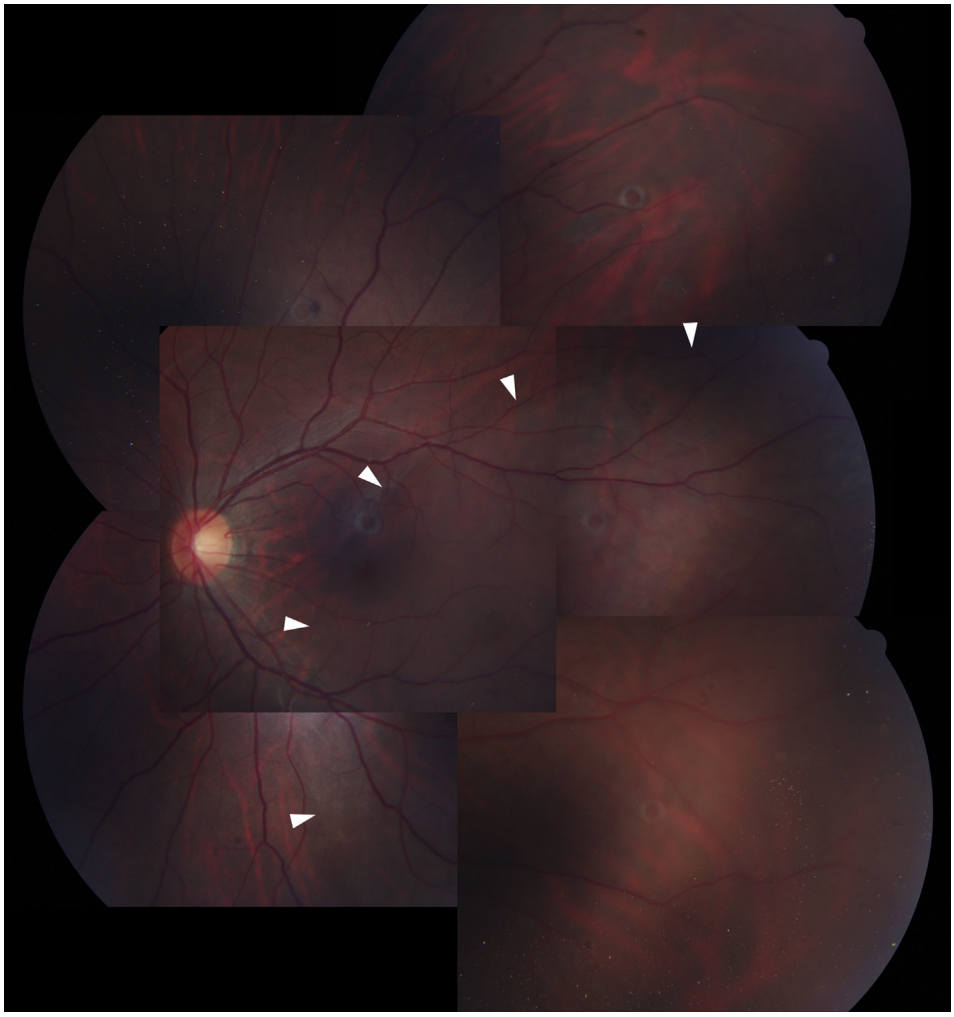

Fig. 1. Shallow retinal detachment at the temporal lower quadrant from the 2 to 5 o'clock meridian (arrowheads) involving macula with no visible retinal break in the corresponding area.

at the temporal lower quadrant from the 3:30 to 4:30 o'clock meridian. Cryopexy was performed at the peripheral retina and pars plana adjacent to the break, and a high segmental buckle was applied at the ora serrata posterior to the break. The subretinal fluid resolved completely 10 days postoperatively (Fig. 3), and the optical coherence tomography showed an attached macula (Fig. 2B). However, postoperative slit-lamp microscopy with goniolens revealed a break at the pars plicata nonpigmented epithelium with its edge pulled to the lens (Fig. 4). The surrounding ciliary process was rudimentary, indicating a focal hypoplastic ciliary body. The detached membrane of pars plicata extended posteriorly and was continuous with the detached pars plana and retina. Diffuse light with retroillumination during maximal pupil dilatation showed a subtle lens defect with segmental flattening adjacent to the pars plicata break (Fig. 5). The patient was diagnosed with a retinal detachment with a pars plicata break associated with congenital malformation of the lens–zonule–ciliary body complex.

Three months later, the patient experienced head trauma by bumping into a door. Fundus examination revealed localized shallow subretinal fluid surrounding the pars plicata break, and no additional break was noted. Laser photocoagulation was applied directly onto the scleral buckle and its posterior edge to confine the subretinal fluid (Fig. 6A). His condition remained stable during the 3-year follow-up (Fig. 6B), and the best-corrected visual acuity

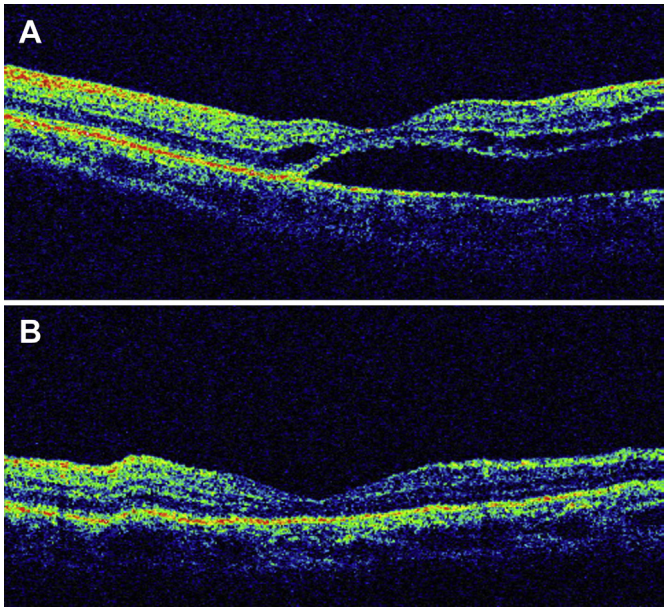

Fig. 2. Optical coherence tomography of (A) the detached neurosensory retina at presentation and (B) the attached macula postoperatively.