Case report

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

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A R T I C L E  I N F O

Article history:
Received 31 March 2014
Received in revised form
14 May 2014
Accepted 25 June 2014
Available online 18 September 2014

Keywords:
malformation of the lens–zonule–ciliary body complex
pars plicata break
retinal detachment
scleral buckle

A B S T R A C T

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 performing segmental scleral buckling, cryopexy, and laser photocoagulation.

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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 in the right eye and −8.0 to 1.75 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 pigmented 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...
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...
remained at 6/60 owing to long-term macular detachment prior to definite diagnosis and treatment.

3. Discussion

Retinal detachment with a pars plicata break associated with lens–zonule–ciliary body congenital malformation is rarely reported.\(^3\)–\(^5\) It may be overlooked on examination because the detachment is typically shallow, and the break is concealed at the distal periphery behind the iris with only subtle lens anomaly, visible only during maximal pupil dilation. The most important procedure to detect the pars plicata break is binocular indirect ophthalmoscopy with scleral indentation, along with an ultrasound biomicroscopy, if available.\(^6\) Previous reported cases of pars plicata breaks are in young Japanese male patients, most of whom have a history of atopic dermatitis.\(^3\)–\(^4\) To the best of our knowledge, this is the first reported case in a Chinese patient and one without any history of atopic dermatitis.

The pathogenesis of spontaneous break in the pars plicata nonpigmented epithelium in patients with congenital malformation of lens–zonule–ciliary body complex remains unknown. All of the reported cases have a pars plicata break located in a region of

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**Fig. 3.** Ten days after performing the scleral buckling and cryopexy, the retina is well attached. A break (arrow) anterior to ora serrata is found at the temporal lower quadrant.

**Fig. 4.** Gonioscopy reveals a break at the temporal lower nonpigmented epithelium of pars plicata (arrow) with its edge pulled to the lens.

**Fig. 5.** Diffuse light with retroillumination during maximal pupil dilatation shows a subtle lens defect with segmental flattening adjacent to the pars plicata break.
segmental ciliary process hypoplasia. Embryology shows that nonpigmented and pigmented epithelium of ciliary body is the anterior extension of the neural retina and retinal pigment epithelium, respectively. The two ciliary body epithelial layers are normally tightly attached by intercellular junctions. Congenital ciliary process hypoplasia may be associated with degenerative epithelium, which has a loose intercellular junction in an animal study. Thus, the hypoplastic ciliary process may be more vulnerable to traction of the zonules, which is generally believed to cause the pars plicata break and lead to retinal detachment.

Retinal detachment associated with a pars plicata break shares several similarities to retinal detachment caused by retinal dialysis; both occur predominately in young males, most have an inferotemporal break, and are easy to overlook owing to the challenge of examination. In retinal dialysis, the break occurs at the ora serrata and is usually associated with trauma and lacks any lens–zonule–ciliary body anomaly.

Retinal detachment related to pars plicata breaks is usually treated with anterior scleral buckling at the ora serrata and cryopexy or diathermy to seal the breaks. Some authors recommend performing the segmental intrascleral buckling directly above the break, but this may result in marked astigmatism. For larger tears, encircling buckling is recommended to reconstruct the artificial ora serrata and release the zonular traction. Pars plana vitrectomy with lens extraction is considered in recurrent cases or cases with particularly large tears to alleviate the tractional force. In this patient, high segmental scleral buckling targeting the ora serrata combined with cryopexy at the peripheral retina and pars plana adjacent to the break were adequate to reattach the retina. Additional laser photocoagulation at the buckle confined the shallow subretinal fluid following the later episode of head trauma.

In conclusion, the present report marks the first Chinese case of spontaneous retinal detachment with a pars plicata break associated with malformation of the lens–zonule–ciliary body complex. Although the retina was successfully surgically reattached, the visual prognosis is guarded because of the chronicity of the macular detachment. Detailed retinal examination including binocular indirect ophthalmoscopy with scleral indentation and gonioscopy of pars plicata should be considered in cases of retinal detachment without a visible break, even in patients without previous atopic dermatitis.

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