

# Postscleral Fenestration Zonulysis in A Patient with Idiopathic Uveal Effusion Syndrome Misdiagnosed as A Ring Melanoma of Ciliary Body

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## Abstract

**Purpose:** To describe the case of a 21-year-old patient with uveal effusion with no microphthalmia and any systemic disease that was treated with scleral window surgery and topical administration of mitomycin C (MMC) and zonulysis which was misdiagnosed as a ring melanoma of ciliary body appeared shortly after the operation

**Case report:** A uveal effusion was detected in the eye. Partial-thickness scleral flap with sclerostomy was performed and topical MMC was administered to inferotemporal quadrant of the equatorial sclera. The subretinal fluid resorbed gradually. In a short period after fenestration procedure and temporal zonulysis appeared in the eye.

**Conclusion:** In a patient with idiopathic uveal effusion syndrome, a significant zonulysis without severe intraocular pressure (IOP) changes can appear after scleral fenestration procedure.

**Keywords:** Uveal Effusion Syndrome, Zonulysis, Ring Melanoma of Ciliary Body

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## Introduction

Schepen and Brockhurst were the first who described idiopathic uveal effusion syndrome as a rare condition causing annular cilio-choroidal detachment and unremarkable inflammation in the anterior segment. The fundus change is characterized by the "leopard-spot".<sup>1</sup> It can occur in nanophthalmic and normal eyes with variable scleral thickness.<sup>2</sup>

Lens subluxation is due to rupture or weakening of the zonules that can occur in Marfan syndrome, homocystinuria, Weil Marchesani syndrome, sulfide oxidase deficiency, syphilis, intraocular neoplasia, aniridia, microspherophakia, and iatrogenic causes such as prior vitrectomy or filtration procedures and trauma.<sup>3</sup>

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No proprietary interest.

Herein we describe a patient with idiopathic uveal effusion syndrome who experienced zonulysis after scleral fenestration surgery, which was misdiagnosed as a ring melanoma of ciliary body.

### Case report

A 21-year-old male was referred for evaluation of a peripheral, elevated, melanotic fundus mass, suspected to be a diffuse ciliochoroidal melanoma, in the left eye (OS). He was presented by vision loss in OS since 2 months before. On examination, the patient's best corrected visual acuity (BCVA) was  $20/25$  in the right eye (OD) and  $20/40$  in OS with 2+ relative afferent pupillary defect in OD. His refraction was plano in OD and -3.00 in OS.

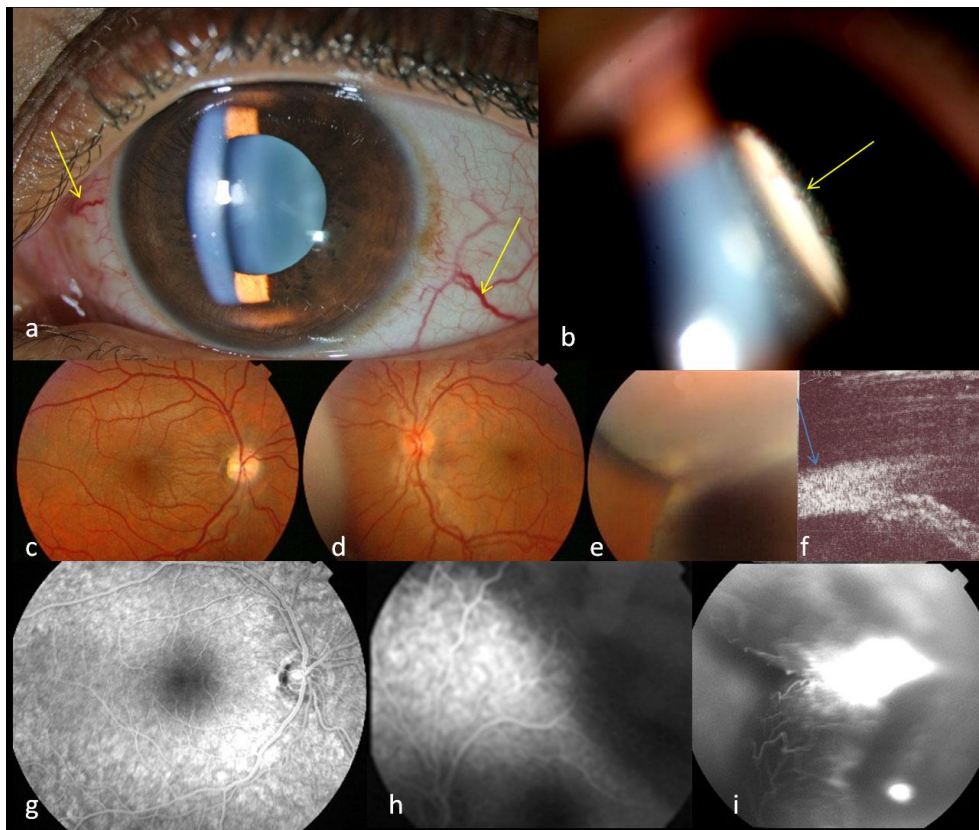
Slit-lamp examination showed bilateral normal sclera, moderately shallow left anterior chambers, and bilateral normal lens and dilated episcleral vessels in both eyes. Intraocular pressure (IOP) was 10 mmHg in OU. There were little pigmentation and trace cells in OS anterior vitreous face and a bulged mass in the back of crystalline lens nasally (Figure 1).

Funduscopy showed paler optic disk in OD, mild engorgement and tortuosity of large vessels in OS, a peripheral, large, smooth, brown mass lesion stretching the ora serrata, extending 360 degrees meridian to the equatorial region, with localized pockets of nonshifting subretinal fluid at its base with some telangiectatic and aneurysmal vessels all over on the stretched retina specially in the temporal side with mild vitreous reaction (Figure 1). Indentation funduscopy of the OD revealed 2 similar small elevations at the extreme temporal and nasal fundus periphery. Ultrasound revealed retinochoroidal complex detachment and normal axial length (24 mm) and posterior sclera with higher echogenicity implying calcification of the sclera. No solid lesion was found. Ultrasound biomicroscopy demonstrated fluid accumulation in peripheral suprachoroidal space, elevating the ciliary body and peripheral choroid from the sclera and a thick sclera peripherally in OS and limited similar changes in OD (Figure 1: F). Fluorescein angiogram in both eyes showed leopard-spot hyperfluorescence indicating some degenerative change in the retinal

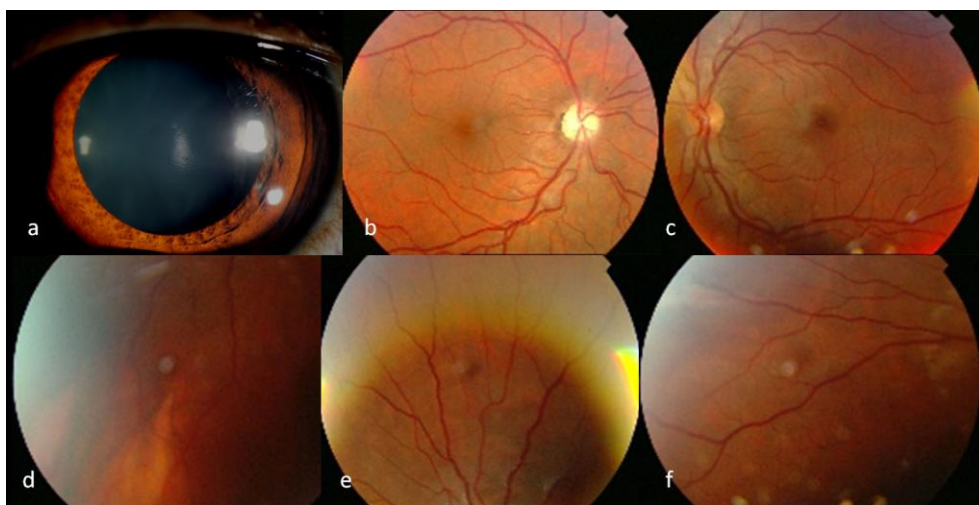
pigment epithelium and leaking vessels on the periphery of the retina, over the elevated retinochoroid layer (Figure 1: G-I).

Cerebrospinal fluid and neuroimaging were not remarkable. There was no evidence of other causes of zonulysis within the eye. The situation was explained to the patient and scleral fenestration was suggested as treatment. The patient refused the operation for nearly one year. After that time, the visual acuity (VA) in OS dropped to finger count and the patient accepted the operation and a rectangular 10x5 mm flap, 0.5 mm thick, first was created in equatorial area infero-temporally. The sclera was thicker than normal (up to 2 mm) and abnormally rigid and we had to change the blade twice. Two flaps were removed in two steps until choroid hue was appeared and suprachoroidal fluid was oozed out at this time. Then the flap was replaced and sutured with two 10-0 nylon sutures in two corners. After removal of the second scleral block -before opening of the sclera- 0.02% mitomycin C (MMC) was applied to the bed for 1.5 minute. Histologically, scleral flaps had degenerated fibro-collagenous tissue without calcification. The patient was treated with topical cycloplegics and steroids for 6 weeks.

In 3 weeks postoperative follow-up the retina and choroid were dried up with some leaf-like subretinal pigmentary and atrophic changes pointing to posterior pole in the midperiphery (Figure 2). Two months later the patient presented with low vision in OS. His BCVA was  $20/25$  OD and  $20/30$  OS. IOP was normal OU. In the slit exam, a zonulysis extending between the 2 and 6 o'clock meridian was shown with trace cells in the anterior vitreous. The fundus had no new changes. The OCT showed normal findings. The telangiectatic far peripheral vessels were disappeared now with some ghost vessels remnants. After 4 months BCVA was  $20/25$  (-0.5x15) in OD and  $20/20$  (-1.75x-2.75x105) in OS without any sign of reaccumulation of suprachoroidal fluid but the zonulysis had extended to nearly half of the zonules with phacodonesis now. After then the patient was followed for 18 months without any further extension of the zonulysis.



**Figure 1.** The ocular appearances of a patient with suprachoroidal effusion syndrome. A: dilated episcleral vessels temporally and nasally without any sign of zonulysis. B: Dilated, aneurysmal changes and sheathed peripheral vessels (arterioles and venules). C-E: Fundus photograph of both eyes showing mild pale disk of right eye and mildly engorged vessels. The peripheral part of fundus showed suprachoroidal effusion with small pockets of subretinal fluid. F: UBM showed thick and dense sclera anteriorly. The choroid is elevated but the ciliary body in this cut is not detached but is rolled anteriorly. G-I: The fluorescein angiogram of patient shows Leopard spots in both eyes. The peripheral part of retina is defocused with subretinal and suprachoroidal fluid 360 in left eye. Leaking telangiectatic and aneurysmal vessels on the elevated part is easily visible throughout the elevated areas but well prominent in temporal area.



**Figure 2.** Slit photo and fundus photograph of patient with idiopathic suprachoroidal effusion syndrome after treatment. A: The slit photo of the patient 2 months after scleral fenestration shows zonulysis from 2.00 to 6.00 o'clock (temporally). B-F: Fundus photo of right eye shows pale disk. In the left eye, there is no suprachoroidal and subretinal fluid anymore and he has leaf like areas of pigmentary changes in the midperiphery of fundus in the area of posterior part of previously elevated choroid (D,F).

## Discussion

The uveal effusion syndrome must be distinguished from a solid tumor to prevent erroneous enucleation.<sup>4</sup> The presence of abnormally dense sclera obstructing venous outflow leads to uveal fluid retention with subsequent uveal swelling and subretinal transudation often associated with low-grade inflammation. It can be managed by sclerectomy to create a bypass outflow.<sup>5</sup> Secondary uveal effusion may result from ocular hypotony or inflammations such as uveitis or scleritis.<sup>6</sup> In our patient there was no ocular problem before.

To the best of our knowledge, presumed spontaneous zonulysis after scleral fenestration procedure is not reported before. Davis recently has studied late in-the-bag spontaneous intraocular lens (IOL) dislocation in 86 consecutive cases. The main conditions associated with IOL dislocation were pseudoexfoliation, prior vitreoretinal surgery, trauma, uveitis, and unknown reason.<sup>7</sup> Gimbel stated that pseudoexfoliation, uveitis, myopia are the predisposing conditions for progressive zonular weakening.<sup>8</sup>

Our patient had no known medically associated condition associated with

spontaneous zonulysis and any trauma history, accounting for a high proportion of acquired cases of subluxation. Inflammation in chronic uveitis is a less recognized cause for destruction of zonules.<sup>9</sup> Other rare reported causes include syphilis, buphthalmos, ciliary body tumor and severe or pathological myopia.<sup>10</sup>

It is hypothesized that long standing low grade inflammation in anterior vitreous and zonules in our patient had caused some restrictive changes in zonules causing their instability and shortening. Suprachoroidal fluid drainage at the time of surgery and then consecutive complete and rapid absorption of suprachoroidal fluid in postoperation period could cause tractions on the already shortened or inflamed zonules causing progressive zonulysis as a complication.

## Conclusion

This report describes a progressive zonulysis after scleral fenestration without severe IOP changes in a patient with uveal effusion syndrome misdiagnosed as ring melanoma of ciliary body.

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