CASE REPORT

Asymmetric glaucoma in pseudoplateau iris syndrome

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SUMMARY

A 39-year-old Caucasian man with bilateral narrow angles, a plateau-like iris configuration on gonioscopy and elevated intraocular pressure (IOP) presented with significant asymmetric glaucoma, left eye affected more than right. Initial management with topical medical therapy, laser iridoplasty and peripheral iridotomy in the left eye was ineffective in lowering the IOP or opening the anterior chamber angle. Ultrasound biomicroscopy demonstrated bilateral ciliary body cysts. The patient ultimately required surgical management, consisting of cataract extraction and endoscopic cyclophotocoagulation of ciliary body cysts in the left eye and trabeculectomy in the right eye, for persistent IOP control to prevent further optic nerve damage and subsequent visual field loss.

BACKGROUND

Pseudoplateau iris (PPI) syndrome represents an anterior displacement of the iris insertion secondary to ciliary body cysts leading to a narrow or closed anterior chamber angle that is critical to identify, as it can be refractory to common ophthalmic interventions such as laser peripheral iridotomy (LPI) or iridoplasty. Although the exact prevalence of PPI is unknown, ciliary body cysts are reported to occur in 5%–54% of healthy individuals.^{1,2} An autosomal dominant inheritance pattern has been observed in some patients with PPI.³ PPI clinically masquerades as plateau iris (PI) syndrome,⁴ although definitive treatment can differ between these two entities. Adjunctive testing, such as ultrasound biomicroscopy (UBM), can aid in diagnosis and ultimately guide an appropriate therapeutic strategy.

CASE PRESENTATION

A 39-year-old man presented as a same-day urgent referral from a local optometrist for evaluation of elevated intraocular pressure (IOP) of 27 mm Hg in the right eye (OD) and 48 mm Hg in the left eye (OS) with a non-contact tonometer. At his initial ophthalmic exam, he had noticed a gradual reduction in his vision in the left eye over the last year. He was not on IOP-lowering medications. However, he was aware of an elevated IOP at prior exams. He denied prior or recent trauma, inflammation or surgery. Ocular history was otherwise negative, except his father being followed as a glaucoma suspect. Medical history was significant for an atrial septal defect. Social history was negative for alcohol, tobacco or illicit drug use.

Presenting best-corrected visual acuity was 20/20 OD and 20/400 OS with a left afferent pupillary defect. Applanation tonometry measurements were 31 mm Hg OD and 35 mm Hg OS with pachymetry measurements of 540 μ m OD and 543 μ m OS. Gonioscopic exam revealed a bilateral plateau-like iris configuration with peripheral anterior synechiae (PAS) temporally with anterior trabecular meshwork in all other quadrants in the right eye and PAS temporally with anterior trabecular meshwork and scattered PAS in all remaining quadrants in the left eye. Slit lamp biomicroscopy was significant for asymmetric cupping of the optic nerves with a cup-to-disc ratio of 0.6 OD and 0.9 OS. There was advanced cupping of the left optic nerve and thinning of the inferior right optic nerve rim. There was no evidence of pigment on the corneal endothelium, iris transillumination defects, anterior capsular pigment or lens opacities.

INVESTIGATIONS

Humphrey visual field (HVF) 24–2 testing OD revealed a superior nasal step (figure 1). HVF 10–2 testing OS revealed central depression (figure 2). Optical coherence tomography (OCT) retinal nerve fibre layer testing revealed temporal thinning from 12 to 6 o'clock of the left optic nerve and borderline thinning superiorly of the right optic nerve. OCT ganglion cell complex analysis revealed early inferotemporal thinning in the right eye and parafoveal thinning in the left eye. UBM testing was significant for multiple ciliary body cysts in both eyes (figures 3 and 4). The anterior chamber depth measured 3.22 mm in both eyes (OU) on lens biometry with IOL Master.

TREATMENT

The patient was diagnosed with secondary chronic angle closure glaucoma secondary to ciliary body cysts in both eyes and underwent 360-degree argon laser iridoplasty OS. Despite iridoplasty OS, his angle remained closed on gonioscopy. The patient underwent LPI OS to relieve any component of pupillary block; however, it was ineffective at deepening the angle and his IOP progressively increased. He underwent cataract extraction with endocyclophotocoagulation of the superior, nasal and inferior quadrants OS to shrink the cysts.

OUTCOME AND FOLLOW-UP

In the subsequent examinations in the postoperative period of the left eye, the anterior chamber was deep and open, with IOP ranging from 9 to 18 mm Hg

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Central 24-2 Threshold Test

Fixation Monitor: Gaze/Blind Spot

Fixation Target: Central

Fixation Losses: 0/15

False POS Errors: 0 %

False NEG Errors: 11 %

Test Duration: 05:50

Fovea: 35 dB

Stimulus: III, White

Background: 31.5 ASB

Strategy: SITA-Standard

Pupil Diameter:

Visual Acuity:

RX: +1.50 DS DC X

Figure 1 Humphrey visual field 24-2 testing of the right eye revealed a superior nasal step. The visual field was reliable as all testing indices were within acceptable limits. (Image is property of the Ohio State University Eye Physicians-Surgeons, Columbus, Ohio, USA). DC, diopters of cylinder; DS, diopters of sphere; GHT, glaucoma hemifield test; MD, mean deviation; PSD, pattern standard deviation; Rx, prescription.

Central 10-2 Threshold Test

Fixation Monitor: Gaze/Blind Spot

Fixation Target: Central

Fixation Losses: 2/17

False POS Errors: 0 %

False NEG Errors: 25 %

Test Duration: 08:47

Fovea: <0 dB ■

Stimulus: III, White

Background: 31.5 ASB

Strategy: SITA-Standard

Pupil Diameter:

Visual Acuity:

RX: DS DC X

-26	-26
-16	-20 -19
-20	-12 -12 -16
-15	-20 -16 -21
-19	-25 -20 -20 -20
-35	-36 -34 -37 -35
-35	-36 -36 -37
-33	-35 -36 -24
-21	-20 -16
-17	-11

-10	-10
-1	-4 -3
-4	4 4 0
1	-4 0 -5
-3	-9 -4 -4 -4
-19	-20 -18 -21 -19
-19	-20 -20 -21
-17	-20 -20 -8
-5	-4 0
-1	5

MD -25.71 dB P < 1%

PSD 8.70 dB P < 1%

Total
Deviation Pattern
Deviation

:: < 5%

⊗ < 2%

⊗ < 1%

Figure 2 Humphrey visual field 10-2 testing of the left eye with superior and inferior central depression. Testing indices show an elevation in false positives without affecting the overall testing reliability. (Image is property of the Ohio State University Eye Physicians-Surgeons, Columbus, Ohio, USA). ASB, apostilb; DC, diopters of cylinder; DS, diopters of sphere; MD, mean deviation; PSD, pattern standard deviation; Rx, prescription.

Figure 3 Ultrasound biomicroscopy image of the right eye demonstrating pseudoplateau iris configuration with deep central anterior chamber and narrow angles secondary to anterior displacement of the iris insertion due to ciliary body cysts. (Image is property of the Ohio State University Eye Physicians-Surgeons, Columbus, Ohio, USA).

on prostaglandin analogue monotherapy. The patient initially had stable IOP control in the right eye, but his IOP OD increased over time to 36 mm Hg while on topical therapy OU. A 360-degree argon laser iridoplasty OD was performed as initial management in this eye given the prior experience of ineffectiveness of LPI in the left eye. However, his angle remained narrow and IOP consistently elevated. In an attempt to preserve his excellent visual acuity and prevent further functional loss, he subsequently underwent trabeculectomy with Mitomycin-C (MMC). On his most recent examination, his best-corrected vision was 20/20 OD and counting fingers OS with IOP of 8 mm Hg OD and 18 mm Hg OS on a prostaglandin analogue at bedtime (QHS) OS with stable OCT and visual field testing OU.

DISCUSSION

Our case details a patient with asymmetric glaucomatous damage from chronic angle closure secondary to bilateral ciliary body cysts. Histologically, ciliary body cysts result from a separation of the non-pigmented and pigmented ciliary body epithelium with clear serous contents.⁵ Cysts can be primary or secondary to trauma, surgery or inflammation. The location of these cysts predisposes to a posterior pushing mechanism of angle closure. The gradual narrowing of the anterior chamber can lead to PAS formation and chronic angle closure glaucoma.

Prior case reports have further highlighted this unique clinical entity. Patients within these case reports presented both with and without symptoms of angle closure. Slit lamp and gonioscopic examinations were supplemented with ancillary testing, such as UBM, to identify the ciliary body cysts. Treatment of these patients consisted of observation and argon laser iridoplasty with reference to more direct approaches to cyst management such as nd:YAG laser iridocystotomy.^{6–8}

Kunimatsu and colleagues evaluated 232 eyes of 116 healthy patients, both volunteers and established clinical patients. In their study, 54% of subjects were found to have ciliary body cysts on UBM. The cysts were typically located in the inferior and temporal quadrants, more commonly bilateral and associated with younger patients (aged 20s and 30s). They did not find a significant relationship between cysts with regard to refractive error.¹ Shukla and colleagues conducted a retrospective analysis of patients with PPI and PI syndrome. They found that patients with PPI are more likely to be male, slightly younger than patients with PI, 'bumpy' peripheral iris appearance on gonioscopy, greater trabecular meshwork pigmentation and fewer clock hours of angle closure versus patients with PI.⁹

Clinical examination and a high index of suspicion are critical in the diagnosis of ciliary body cysts, especially in atypical presentations of narrow or closed angles in a young patient without prior

Figure 4 Ultrasound biomicroscopy image of the temporal angle and sulcus (transverse scanning) of the left eye demonstrating extensive ciliary body cysts. (Image is property of the Ohio State University Eye Physicians-Surgeons, Columbus, Ohio, USA).

trauma or surgery.¹⁰ The treatment of ciliary body cysts involves a stepwise approach, which carries a risk of subsequent pigment dispersion, inflammation and cyst recurrence.⁷ In asymptomatic patients, cysts can be observed as long as clinical examination and testing remain stable. With progressive angle narrowing, iridoplasty

or gonioplasty is a proven treatment to pull the iris away from the anterior chamber angle, resulting in opening of the angle, similar to PI syndrome. However, more definitive approaches to cyst management such as endoscopic cyclophotocoagulation as in our patient may be required to stabilise a patient's clinical course.

Contributors GPF was the primary clinician/surgeon, but all authors were involved in the patient's care. JSS and GPF were responsible for the creation of the original and revised manuscript and performed the scientific literature review. SGJ and AKS provided critical review of the manuscript and necessary revisions.

Competing interests None declared.

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Learning points

- Pseudoplateau iris (PPI) syndrome is an important cause of secondary angle closure glaucoma, especially in younger patients.
- Ciliary body cysts can be diagnosed based on gonioscopic findings and with ancillary testing such as ultrasound biomicroscopy.
- Primary treatment of PPI involves in-office laser procedures such as a peripheral iridotomy to relieve any possible component of pupillary block and gonioplasty/iridoplasty to open the anterior chamber angle.
- Cysts refractory to initial ophthalmic interventions may require laser iridocystotomy, surgical drainage and removal, endoscopic cyclophotocoagulation, or potential adjunctive filtering or shunting surgery for adequate pressure reduction and stabilisation of clinical course.

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