American Journal of Ophthalmology Case Reports 28 (2022) 101694

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Multiple iridociliary cysts: One entity with various clinical presentations

S.A. Kandarakis [a,*], L. Doumazos [a], P. Petrou [a], K. Droutsas [a], D. Karagiannis [b], I. Georgalas [a]

[a] 1th Opthhalmology Department, "G.Gennimatas" Hospital, National and Kapodistrian University of Athens, Athens, Greece
[b] 1st Department, Opthalmiatreio Eye Hospital of Athens, Athens, Greece

ARTICLE INFO

Keywords:
Angle closure glaucoma
Secondary glaucoma
Iridociliary cysts
CDR asymmetry
Pseudoplateau iris

ABSTRACT

Purpose: Iris cysts have a wide range of clinical manifestations ranging from totally asymptomatic to full-blown glaucoma. The aim of this report is to present three patients with multiple bilateral iridociliary cysts. Each one of them had a different stage of glaucoma and required individualized management.

Observations: All three patients presented to our clinic. Patient 1 was a 25-year old female with no symptoms and normal intraocular pressure (IOP), Patient 2 a 20-year old female with cup-disk asymmetry and nocturnal IOP spikes and Patient 3 a 69-year old male with chronic angle closure, uncontrolled IOP on maximum medical therapy and optic nerve cupping. During clinical evaluation, gonioscopic findings raised the suspicion of possible multiple bilateral iridocilary cysts, which were then verified with ultrasound biomicroscopy in all patients. Each case was managed differently; Patient 1 was opted for observation, Patient 2 was started on prostaglandin analogue and Patient 3 underwent trabeculectomy with Mitomycin-C.

Conclusions and Importance: Although iris cysts represent a relatively rare entity, patients with this anomaly can be encountered in our everyday clinical practice. They may be totally asymptomatic, where observation alone suffices, or at the other end of the spectrum, they may present with full-blown glaucoma, requiring traditional glaucoma surgeries. Considering the wide variety of clinical manifestations and that management may differ depending on the diagnosis, it is highly important for clinicians to carry out a careful examination, especially in patients with narrow angle. In cases of high suspicion for iris cysts, ancillary testing, such as Ultrasound Biomicroscopy, should be performed for the definite diagnosis as well as for the exclusion of malignancies.

1. Introduction

Although iris cysts represent a relatively rare entity, patients with this anomaly can be encountered in our everyday clinical practice.[1] Their clinical presentation may vary, depending on the location, number and anatomical characteristics of the cysts. Cysts mainly located in the periphery (iridociliary cysts) result in pseudoplateau iris (PPI) syndrome. This represents an anterior displacement of the iris, causing a narrowing or even closure of the angle.[2] Patients with PPI are commonly refractory to typical interventions, such as peripheral iridotomy (PI), rendering their diagnosis important for proper management.[3] The aim of this report is to present three patients with multiple bilateral iridociliary cysts, with each one of them having a different stage of glaucoma and requiring individualized therapeutic plan.

2. Case report

Patient 1, a 25-year-old emmetropic female with no past medical history, presented to our clinic for a regular ophthalmologic examination, without any complaints. During the examination, her visual acuity (VA) was found 20/20 OU, intraocular pressure (IOP) 10 mmHg in both eyes and pachymetry 540μm OD and 550μm OS.

On Slit-Lamp (S/L) examination the anterior chamber (AC) depth was moderate. A lumpy bumpy appearance was noted at the periphery of the iris. Therefore, a gonioscopy was performed, revealing a moderately pigmented angle, as well as multiple localized convexities of the regular flat iris, which extended 360° (Fig. 1A). This morphology led to a sectoral occlusion of the angle, mimicking a plateau iris configuration in both eyes; i.e. pseudoplateau iris. No other abnormalities were found on S/L exam. Furthermore, no glaucomatous damage was observed on dilated fundus exam, while optic disk size, color and neuroretinal rim appeared within normal limits. Additional evaluation with visual field (VF), Optical Coherence Tomography- Retinal Nerve Fiber Layer (OCT-RNFL) and diurnal IOP measurement failed to reveal any remarkable findings. In an effort to further investigate the aforementioned gonioscopic findings, an anterior-segment OCT (AS-OCT) and Ultrasound

* Corresponding author. "G. Gennimatas" Hospital Mesogeion Ave. 154, Athens, GR, 15669, Greece.
E-mail address: s.kandarakis@gmail.com (S.A. Kandarakis).

https://doi.org/10.1016/j.ajoc.2022.101694
Received 1 April 2022; Received in revised form 23 August 2022; Accepted 28 August 2022
Available online 2 September 2022

Biomicroscopy (UBM) were ordered. AS-OCT revealed sectoral variable bulging of the iris, which extended 360° in both eyes, confirming gonioscopy (Fig. 1C). High resolution UBM scanned the iridociliary body both in transverse and radial sections, revealing bilateral, multiple circumferential iridociliary cysts that extended 360° (Fig. 1B). Cysts were more prominent and larger in size in the inferior and temporal sectors of both eyes. Based on both the clinical evaluation and imaging findings the diagnosis of multiple iridociliary cysts with pseudoplateau iris configuration was made. The patient was opted for close observation, initially every three months, with a potential gradual progression to every six months, as long as no worsening was observed.

Patient 2, a 20-year-old emmetropic female with no other past medical history, was referred to our clinic after being found with a cup-to-disk ratio (CDR) asymmetry. The clinical examination performed, revealed a VA of 20/20 OU, an IOP of 14 mmHg OU and a pachymetry measurement of 530μm OD and 543 OS. On S/L examination the AC depth of the patient was moderate. Nevertheless, an observable bump could be seen at the periphery of the iris, which was made clearer on making a slit beam at an angle on her right eye. This finding was then confirmed on gonioscopy, which showed that a large part of the peripheral iris was pushed forward, causing the closure of the angle (Fig. 2A). The rest of the periphery seemed to have smaller protrusions of the iris. Dilated fundus exam confirmed the CDR asymmetry, with OD estimated at 0.40 and OS at 0.15 (Fig. 2OD/OS), while no other significant findings were noted. A new IOP measurement was then taken after dilation of the pupil, showing an increase (IOP: 22 mmHg OD). VF examination was normal with a visual field index of 100%, while OCT-RNFL showed thinning of the superior and inferior quadrants of the right eye. Lastly, diurnal IOP measurement revealed elevated IOP levels of 25 mmHg OD during nocturnal hours. Further evaluation of the angle using AS-OCT, confirmed the aforementioned protrusion of the iris with the concurrent closure of the angle, which was found on gonioscopy. Given that UBM revealed an anechogenic image of the cysts, suggestive of a fluid content, the diagnosis of multiple bilateral iridociliary cysts was made (Fig. 2B). The patient's findings were indicative of ocular hypertension secondary to intermittent angle closure. As an initial approach, treatment options that were discussed with the patient, were topical medical therapy or peripheral laser iridoplasty. The patient opted for topical medical therapy and was initiated with pilocarpine 2%. Since this medication was not well tolerated, it was switched to a prostaglandin analogue QD. After 6 months under therapy, the maximum IOP measured was 16 mmHg OD, while VF and OCT-RNFL remained stable.

Patient 3, a 69-year-old myopic male was referred to our clinic for surgical management of his glaucoma. The patient was under maximal medical therapy (MMT) OU and the clinical examination showed the following findings; his best corrected VA was 20/100 OD and 20/60 OS, pachymetry measurement was 515μm OD and 529μm OS and his IOP was 28 mmHg OD and 20 mmHg OS. He was pseudophakic with posterior chamber intraocular lens OD and phakic OS. On S/L examination his AC was very shallow, while gonioscopy showed extensive iridotrabecular contact (ITC) of more than 270° (Fig. 3A). When dynamic gonioscopy was performed, the angle failed to open, suggesting the presence of peripheral anterior synechiae (PAS). Upon dilation, a large 4 clock hours cyst could be seen in his right eye. Furthermore, the optic disks had an extensive cupping of 0.8 OD and 0.7 OS, while VF revealed an almost tunnel vision OD and a superior arcuate defect OS. OCT-RNFL was also pathologic, presenting a global RNFL and an inferior thinning in the right and left eye respectively. AS-OCT image was captured after dilation, showing the extension of the large cyst beyond the pupillary margin on his right eye. UBM confirmed the existence of this cyst, revealing, additionally, multiple cysts bilaterally (Fig. 3C). Therefore, the diagnosis of chronic angle closure glaucoma with extensive PAS was confirmed and the patient underwent trabeculectomy with Mitomycin-C (MMC) on his right eye (Fig. 3B). Regarding his left eye, further management for the near future was proposed, including close IOP monitoring and a possible 360-degree argon laser peripheral iridoplasty or phacoemulsification with gonio-synechiolysis. At his follow-up visit, six months post-op, the IOP measured was 10 mmHg OD without any topical medication and 16 mmHg OS on MMT. Both OCT-RNFL and VF testing OU were stable.

3. Discussion

Iris cysts are broadly categorized into primary and secondary cysts.[4] Primary cysts are mainly located on the posterior pigment epithelium of the iris.[5] They may be found at the following three zones; at the pupillary margin (central), between the iris root and the ciliary body (midzonal) and most commonly at the iridociliary sulcus (peripheral).[4,5] In some cases, cysts are located on the iris stroma, or are dislodged.[4,5] Primary cysts usually appear during childhood or early adulthood and follow a non-progressive, benign course.[4] It has been, though, reported that some primary cysts, exhibit high recurrence rates after removal and are linked to complications, such as corneal decompensation, amblyopia and strabismus.[6,7] Secondary cysts commonly arise from trauma, surgery, tumor or inflammation and are associated with a higher rate of recurrence and more complications compared to primary cysts.[4,8] One possible pathogenetic mechanism, that may be responsible for the formation of primary cysts, occurs during embryogenesis. In the course of eye development, the zonules may apply traction forces on the ciliary

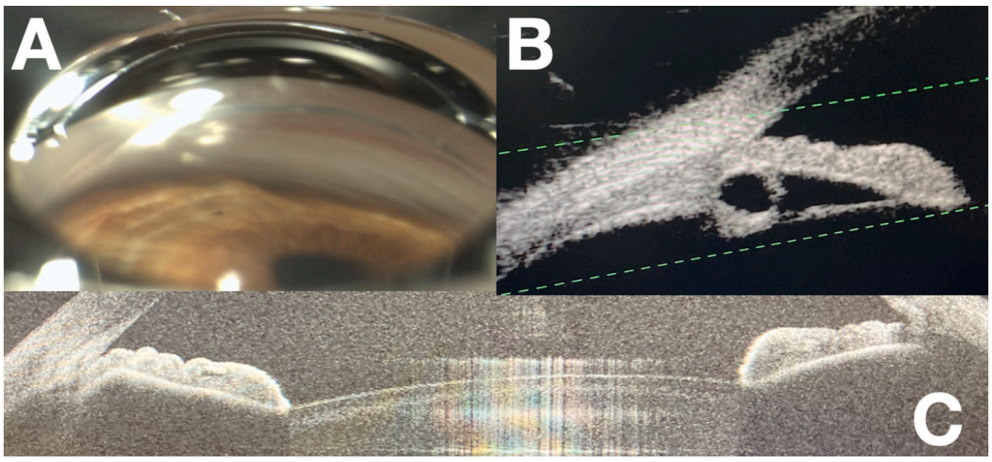

Fig. 1. **A**: Gonioscopic findings of Patient 1 showing the "lumpy-bumpy" appearance of the iris. **B**: UBM image showing the clear contents of the cyst pushing the iris forward. **C**: AS-OCT showing the bowing of the iris but failing to reveal posterior structures.

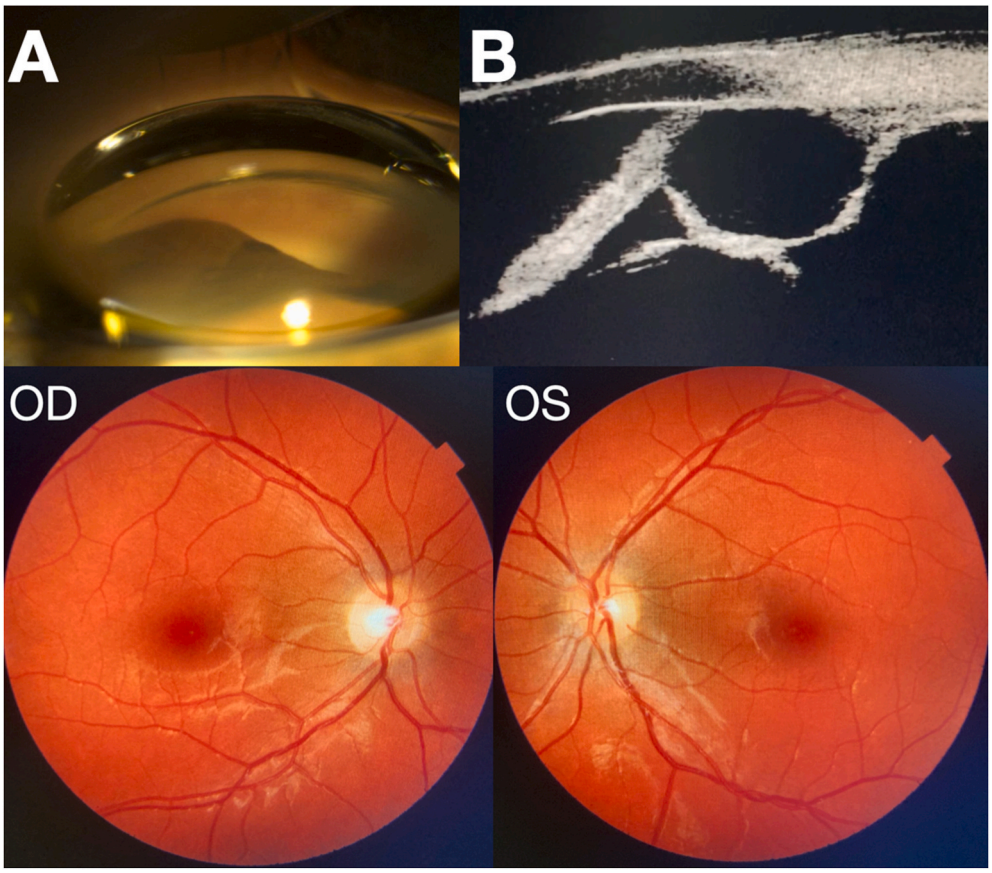

Fig. 2. **A**: Gonioscopic image of Patient 2 showing the angle closure at the location of the cyst. **B**: UBM image of the same cyst. **OD/OS**: Bilateral color fundus photos revealing cup-to-disk ratio asymmetry. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

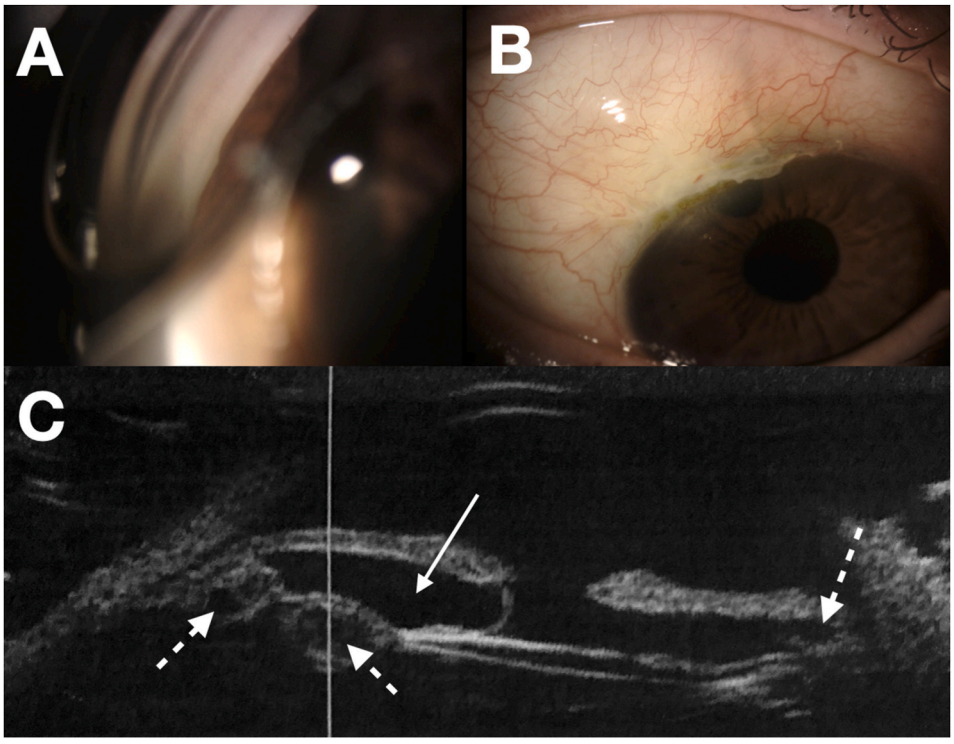

Fig. 3. **A**: Gonioscopic image of Patient 3 with extensive angle closure with peripheral anterior synechiae. **B**: Trabeculectomy with filtering bleb six months post-op. **C**: UBM image revealing the extent and the location of cysts. Note, one large cyst in posterior iris (solid arrow) and multiple, smaller cysts in the ciliary body (dashed arrows).

epithelium, causing the inner and outer layers of the optic cup to pull apart.[4] This mechanism could explain why the inferior and temporal sectors are more frequently affected, as the apposition of the two epithelial layers seems to be embryologically weak on those locations.[1] In fact, our patients had greater size and number of cysts in the inferior and temporal sectors as well. As already mentioned, these cysts can push the iris root anteriorly resulting in a PPI configuration, with or without angle closure. This may consequently cause a rise in the IOP.[2,9,10]

Ultrasound Biomicroscopy (UBM) is an invaluable imaging tool, which provides a resolution of 20–50 μm and a penetrating tissue depth of 4mm.[1,4,11] These features allow a better visualization and evaluation of the size, structure, location as well as the extent of involvement of the cysts.[1,4,11] Therefore, UBM can be used to diagnose iridociliary cysts mimicking iris plateau configuration. This condition is known as pseudoplateau iris and is clinically difficult to distinguish.[1,11] Moreover, UBM may be the only effective tool in differentiating iris cysts from pathologic uveal neoplasms.[11]

Iridociliary cysts have already been emphasized in previous case reports, and describe patients both with and without angle closure symptoms. In these cases, UBM examination confirmed the diagnosis and was recognized as an irreplaceable tool, ancillary to S/L and gonioscopy.[3,9–12] Patients with multiple iridociliary cysts extending 360° in both eyes, have been relatively rare in literature. This is a unique case series highlighting three different case reports with this rare pathology, similar findings in S/L, gonioscopy and UBM, nevertheless requiring completely different management based on the severity of glaucoma.

Patient 1 presented with a partial angle closure and did not exhibit any IOP changes, while the other two patients had a greater degree of angle closure and consequently, higher IOP. PPI configuration, although present in all three patients, resulted in IOP spikes during nocturnal hours only in Patient 2. Baba et al. presented a symptomatic case with high nocturnal IOP levels (67 mmHg), CDR asymmetry, but without any VF defects.[13] Accordingly, Patient 2 had similar clinical features, being though asymptomatic, probably due to lower maximum IOP spikes. Moreover, a case report by Swogger et al. described a patient with PAS in gonioscopy, elevated IOP, cupping of optic nerves and VF defects, that were found to be secondary to the closure of the angle by multiple iridociliary cysts.[3] These findings come in accordance with those of Patient 3. Similar characteristics with Patient 2 and 3 were further reported by Baile et al. in a case report of a patient with bilateral multiple iridociliary cysts. A large cyst was visible on S/L examination, closing the angle at that location, a feature similar to that observed in Patient 3. In addition, similarly to Patient 2, the case's IOP was within normal levels, but got elevated upon dilation.[14] Multiple iridociliary cysts leading to glaucoma, were previously reported 26 years ago by Azura-Blanco et al. in a case series of three patients. Their presence was confirmed with UBM and the authors associated them with plateau iris configuration.[12] All prementioned reports confirm that iris cysts, despite representing one pathophysiologic entity, may have multiple clinical manifestations.

Therapeutically, several management strategies have been proposed, with a step-up approach being most commonly preferred.[4,15] Pilocarpine 2% drops have been shown to improve IOP levels in patients with pseudoplateau iris configuration. However, pilocarpine may be difficult to tolerate in the long term, owing to its possible side-effects.[11–13] Peripheral laser iridotomy has also been attempted, failing, though, to achieve a desirable outcome. In fact, in most cases, it was neither effective in lowering IOP, nor in opening the angle, since angle closure in PPI configuration is not caused by pupillary blockage. Therefore, intermittent angle-closure attacks could not be prevented.[11–13] In addition, the risk of provoking IOP spikes, inflammation and pigment dispersion make this option unfavorable.[11,16,17] Similarly, laser iridocystostomy, apart from not being practical in multiple cysts, was proven ineffective in most cases, since it carries the same risks as peripheral iridotomies.[17] Furthermore, argon laser peripheral iridoplasty has shown contradictory results; in one case IOP was improved,[16] while in another no change in IOP and angle opening was observed.[3,17] In cases where IOP remains off target, traditional filtration surgeries have been proposed.[3,11] Regarding the management of our patients, given the lack of symptoms, Patient 1 was opted for regular observation. Patient 2 was initiated with pilocarpine therapy, which however caused severe headaches. After changing her therapy to a prostaglandin analogue, no complaints arose, while the IOP was well controlled. Finally, Patient 3 was eligible for neither argon laser peripheral iridoplasty nor iridocystotomy. Actually, these methods would prove ineffective, probably even resulting in an increase in IOP, since Patient 3 presented with more than 270° ITC and PAS. Consequently, he underwent trabeculectomy, achieving low IOP levels.

4. Conclusion

Iris cysts have a vast range of clinical presentations, ranging from totally asymptomatic needing only observation, to full-blown glaucoma requiring major glaucoma surgery. Considering the wide variety of clinical manifestations and that management may differ depending on the diagnosis, it is highly important for clinicians to carry out a careful examination, especially in patients with narrow angle. In cases of high suspicion for iris cysts, ancillary testing, such as Ultrasound Biomicroscopy, should be performed for the definite diagnosis as well as for the exclusion of malignancies.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

Contact with the editorial office

The Corresponding Author declared on the title page of the manuscript is: Stylianos Artemios Kandarakis.

Declaration of competing interest

The following authors have no financial disclosures: **SK, LD, PP, KD, DK, IG.**

Acknowledgements

None.

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