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CASE REPORT

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Elevation of intraocular pressure due to ciliary body and iris cysts "pseudoplateau iris" – case report

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ABSTRACT

Iris and ciliary body cysts constitute a significant diagnostic problem and are difficult to diagnose in routine practice on slit lamp or gonioscopy examination. The diagnosis must be established by ultrabiomicroscopy, which is not widely available, but represents the only technique capable of evaluating the anatomical relationships of the visualized ocular structures.

Diagnosing ocular hypertension and pseudoplateau iris secondary to iridociliary cysts can be challenging for ophthalmologists, as it is rarely possible to perform a complete diagnostic workup. The authors of this article present a rare case of a patient with bilateral pseudoplateau iris and ocular hypertension associated with iridociliary cysts. In addition, various forms of intraocular pressure-regulating therapies recommended in treatment are reviewed.

KEY WORDS: ocular hypertension, pseudoplateau iris, iris and ciliary body cyst, ultrasound biomicroscopy.

INTRODUCTION

Until recently, cysts of the iris and ciliary body were recognized as a rare disease entity [1]. At present, primary cysts are increasingly detected in routine examinations – in 34.4% of patients with a shallow anterior chamber and 54.3% of the general population [2].

Iris and ciliary body cysts can be divided into primary and secondary types. Primary cysts arise from the epithelium or the stroma of the iris. Cysts originating from the stroma are unilateral, solitary, large in size, smooth-walled, and located in the inferior quadrants [2]. Primary epithelial cysts are the most common form of this pathology [3, 4], and arise spontaneously from the pigment epithelium [5, 6]. Typically, they are found in the inferior and temporal parts of the iris and the ciliary body [6, 7]. On ultrabiomicroscopy (UBM), they have thin walls [3-5, 7] and, depending on their location, show high or low echogenicity. Hyperechoic lesions are located in the inferior, temporal and inferotemporal regions, while hypoechoic cysts are found in the inferior, nasal and inferonasal quadrants [6]. Secondary cysts may develop as a result of prolonged use of miotics, injuries, tumor metastases, or may be associated with parasitic infections [3, 5, 8].

Iris and ciliary body cysts pose a diagnostic challenge, being difficult to detect in routine practice on slit lamp or gonioscopy examination [1, 5]. The diagnosis requires an UBM examination to evaluate anatomical relationships between visualized structures and determine the presence of infiltration or inflammation [2]. After cyst detection on UBM, it can be differentiated from tumors including iris melanoma, medulloepithelioma or adenoma of the iris pigment epithelium [2, 5].

Iris and ciliary body cysts can lead to corneal edema, uveitis, impaired visual acuity, focal cataract or glaucoma [2,4]. However, the vast majority have a mild course and do not require treatment [1, 2, 5, 6, 8, 9]. By displacing the iris, they can cause "pseudoplateau iris" (PPI) either with or without filtration angle closure [3, 4, 10, 11]. For the diagnosis of PPI, visualization of cysts at the iridociliary sulcus is necessary. Pseudoplateau iris is a rare condition that is frequently difficult to distinguish from plateau iris (PI), though the latter has a different underlying mechanism from PPI, and the diagnosis is based on the anterior position of the ciliary body which peripherally abuts the iris; a narrow (<10 degrees) or closed angle for at least 180 degrees, and the positioning of the anterior portion of the iris anteriorly to the scleral spur [11]. Closed-angle glaucoma can develop when the area of filtration angle closure exceeds 180 degrees [5]. This mechanism may be common among young people and children with secondary angle closure [3, 12].

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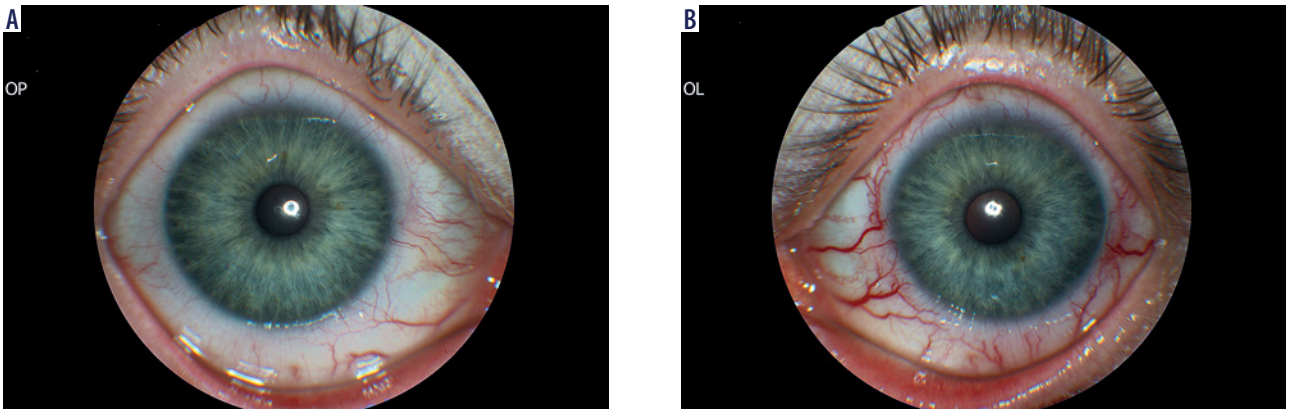


Figure 1. A) Photograph of the anterior segment of the right eye. B) Photograph of the anterior segment of the left eye

Diagnostic issues related to iris and ciliary body cysts are an interesting topic that is rarely discussed in the literature. To highlight the condition, the authors present an ophthalmic case report of a 41-year-old female patient with a diagnosis of bilateral cysts of the iris and ciliary body.

CASE REPORT

The case report concerns a female patient who had undergone multiple ophthalmic consultations for glaucoma and intracranial hypertension associated with ocular hypertension, and a characteristic appearance of the bulbar conjunctiva in both eyes, with dilated, tortuous, corkscrew-shaped conjunctival vessels resembling a "medusa head" (Figure 1A, B).

The patient's medical history showed the following comorbidities: stage 3 endometriosis, diverticular disease, ovarian cysts, and breast fibroids. The woman's family history was significant for cancer (grandmother: colorectal cancer, grandfather: prostate cancer) and rheumatoid arthritis (nephew). Laboratory tests confirmed the presence of the HLA B-27 antigen, and the patient's ophthalmic examination revealed a bulge in the iris of the left eye at 3:30 o'clock together with paralimbally dilated corkscrew-shaped episcleral blood vessels. Ocular oncological diagnostic evaluation consisted of slit-lamp examination, gonioscopy, OCT of the anterior segment of the eye and, most importantly, UBM, which ruled out iris malignancy.

Predominant clinical manifestations included frequent headaches and conjunctival congestion of both eyes, more prominent in the left eye. A Doppler ultrasound showed an increase in systolic velocity in the ophthalmic artery of the left eye and disturbed venous flow in the central retinal vein (CRV) and central retinal artery (CRA), more prominent on the left, with an increase in vascular resistance in the CRA and PCAs (posterior ciliary arteries). A contrast-enhanced magnetic resonance imaging (MRI) examination confirmed segmental dilatation of the sphenoparietal sinus on the left, and ruled out thrombotic and postinflammatory lesions in the cavernous sinus and the orbital region. Subsequent ophthalmic follow-up examinations revealed periodic significant increases in intraocular pressure (IOP), with mean values ranging from approximately 27.6 mmHg to 30 mmHg in the right and left eyes. A follow-up Doppler ultrasound confirmed previously identified abnormalities in the retrobulbar circulation. Subsequent cerebrovascular arteriography ruled out intracranial vascular pathologies. A later UBM examination showed a shallow anterior chamber with a narrow, beak-like, slit-shaped filtration angle, open around the entire circumference. Numerous small and sparse medium-sized cysts were identified between the ciliary processes in both eyes. The largest iris cyst was visualized in the posterior chamber of the right eye at 11:00 o'clock (Figure 2).

Because of persistently elevated IOP levels, conservative treatment (Monoprost) was initiated, but without achieving a desirable reduction in IOP. Subsequently, timolol was added to therapy with good results. A UBM examination performed after two years showed a further increase in the number of cysts, varying in size, abutting the base of the iris around the entire circumference. The filtration angle had an irregular appearance and it was narrow over most of the circumference, closed in some sections. At the time of the evaluation, topical treatment (β-blocker) was effective: IOP in the right eye was 16.8 mmHg, and in the left eye 17.2 mmHg (measured with PASCAL Dynamic Contour Tonometer in the Topcon DC-3 system). A concurrent Doppler ultrasound showed normalized blood flow velocity, though with persistently elevated blood flow resistance (RI 1.0). Ophthalmological evaluation for glaucoma revealed no abnormalities (Figures 3A, B, 4).

DISCUSSION

Since ocular hypertension associated with iris and ciliary body cysts is a rare occurrence, and the differentiation and diagnosis may pose difficulties, no universally recognized treatment algorithm exists.

Kunimatsu *et al.* found no evidence for a link between an increased prevalence of cysts and sex or refractive defects of vision. Of the 116 subjects studied, iris cysts were present in 63 individuals (54.3%). Men and women accounted for 60.8% and 49.2% of the study group, respectively. Iris cysts were more common in patients in the 2nd and 3rd decades of life (73.1% and 80%, respectively). The size of the cyst was found to decrease with age [6, 8]. Monocular and binocular cysts were detected in 16.4% and

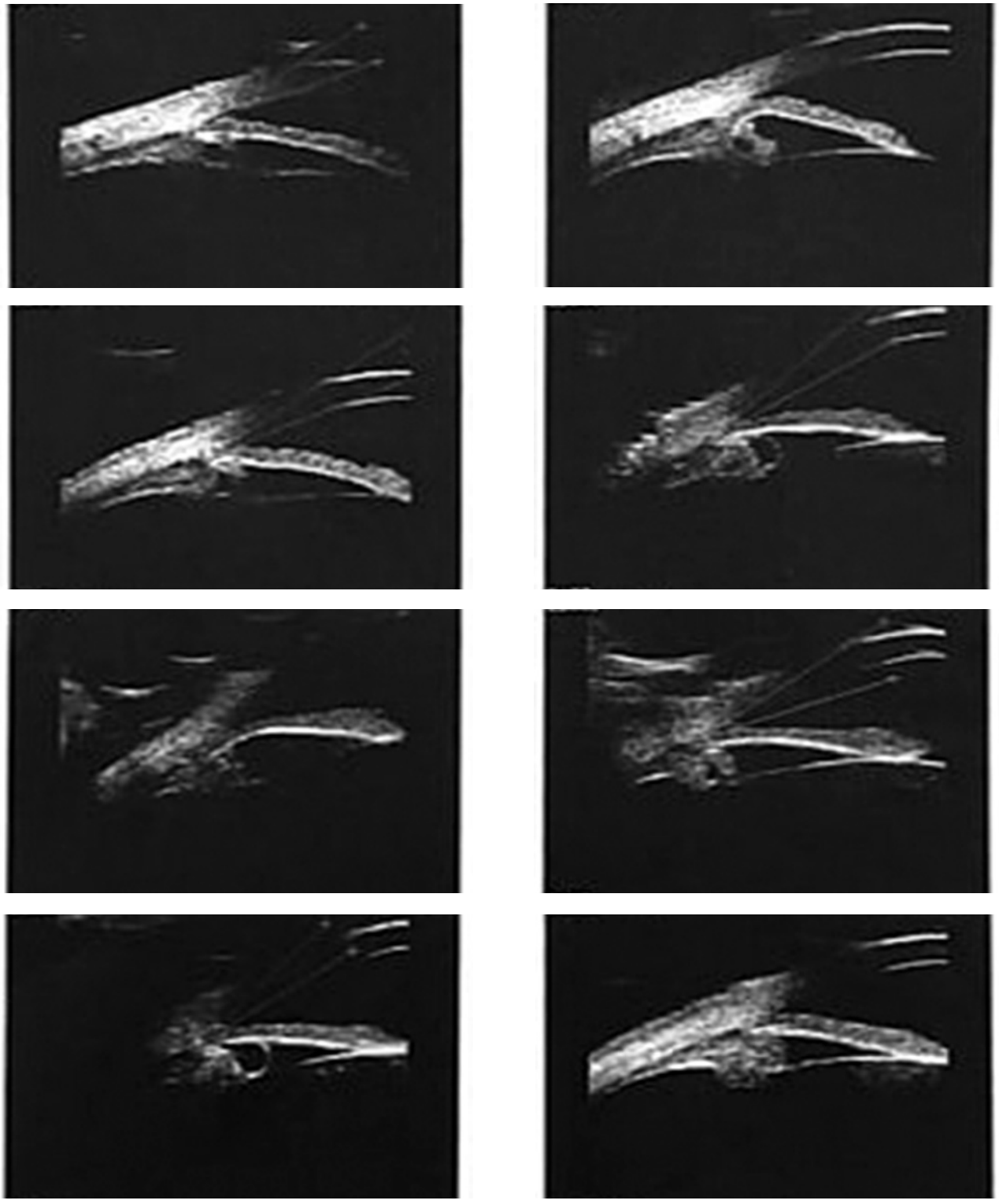


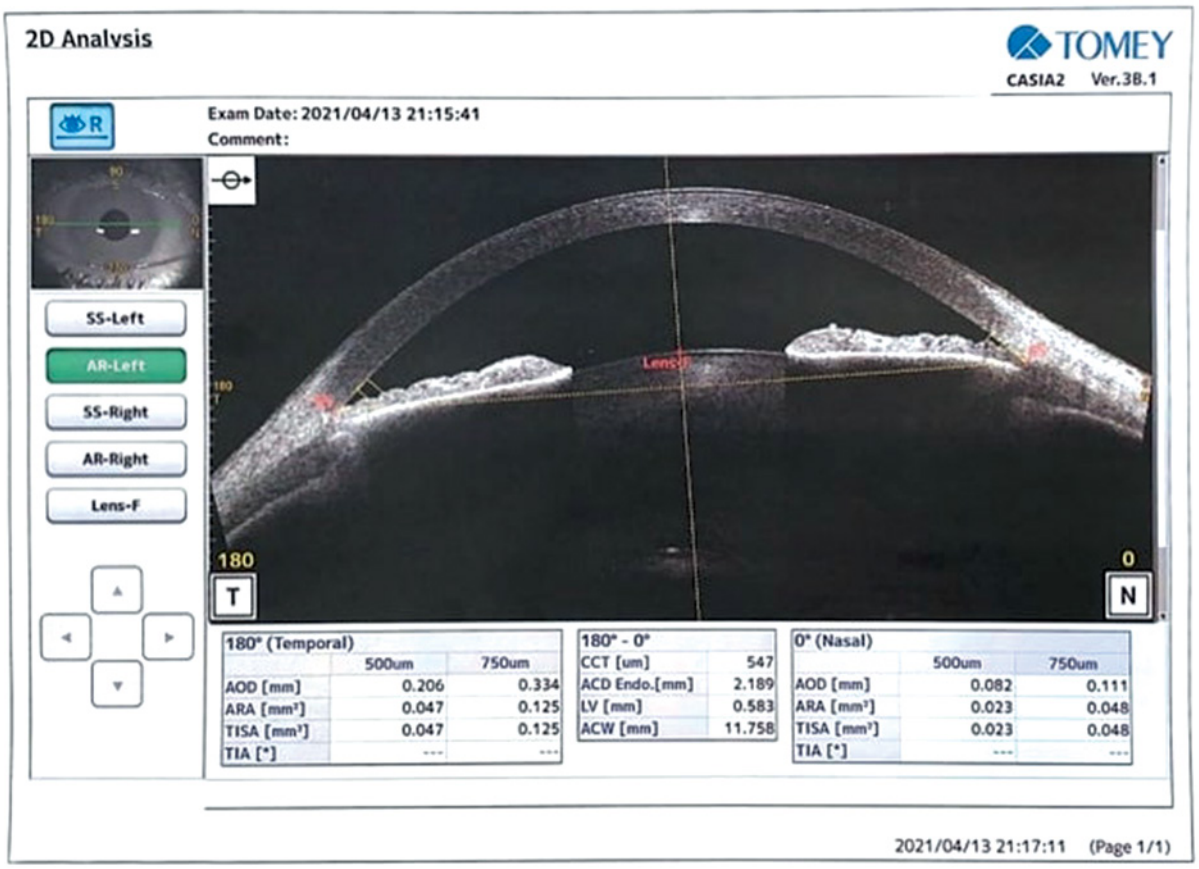
Figure 2. A) Filtration angle and surface of the iris and ciliary body visualized by UBM of the right and left eye

44.8% of cases, respectively. The location and size of the cysts were similar [6].

No association has been found between primary iris and ciliary body cysts and systemic conditions [10]. A statistical correlation has been identified between the number and size of iridociliary cysts and the presence of elevated IOP [4, 9, 11, 12]. The likely mechanisms leading to IOP elevation include stimulation of the ciliary body by the cysts to produce fluid, accumulation of substances produced by the cysts in the trabecular meshwork and concomitant blockage of ventricular fluid outflow, and anterior displacement of the iris causing a progressive narrowing of the angle along with an increase in cyst size [9].

There are two theories explaining the pathogenesis of iridociliary cysts, though neither of them is comprehensive [6]. According to the traction theory, the pull on the lens liga-

A



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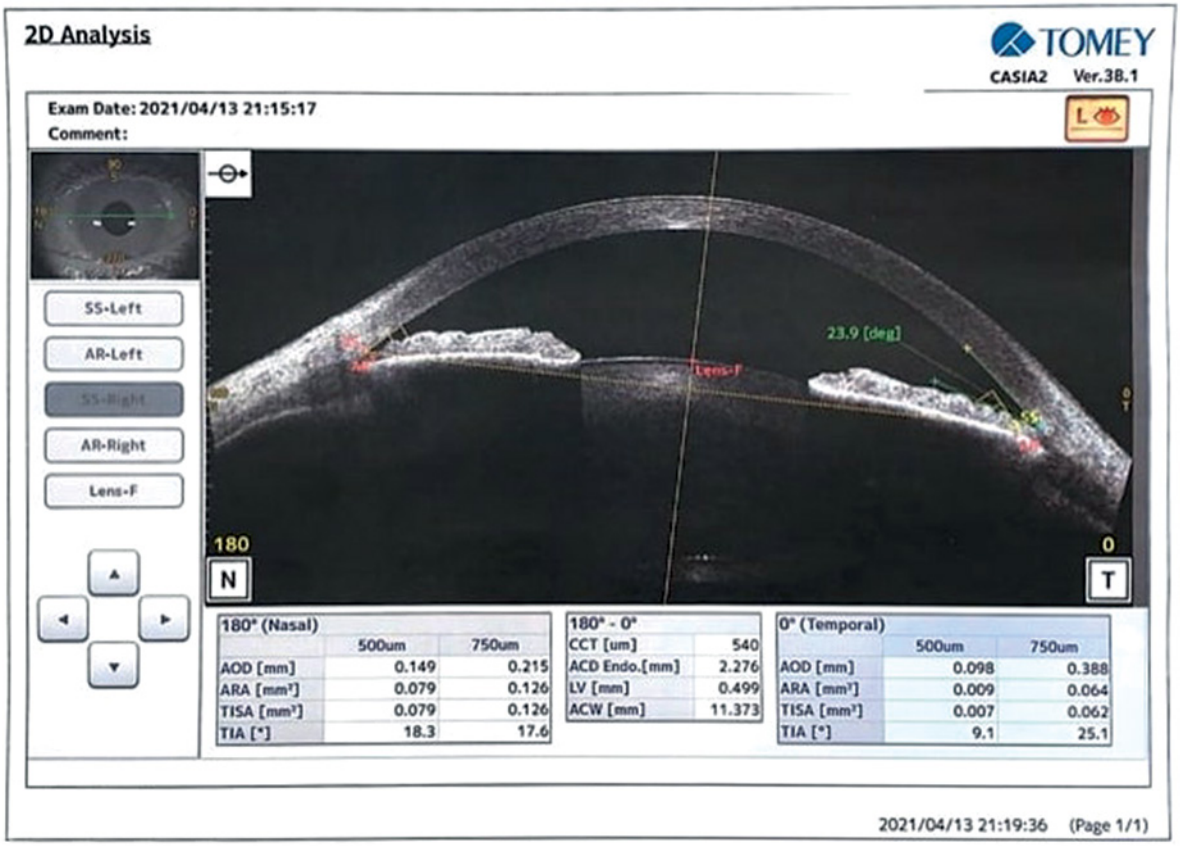
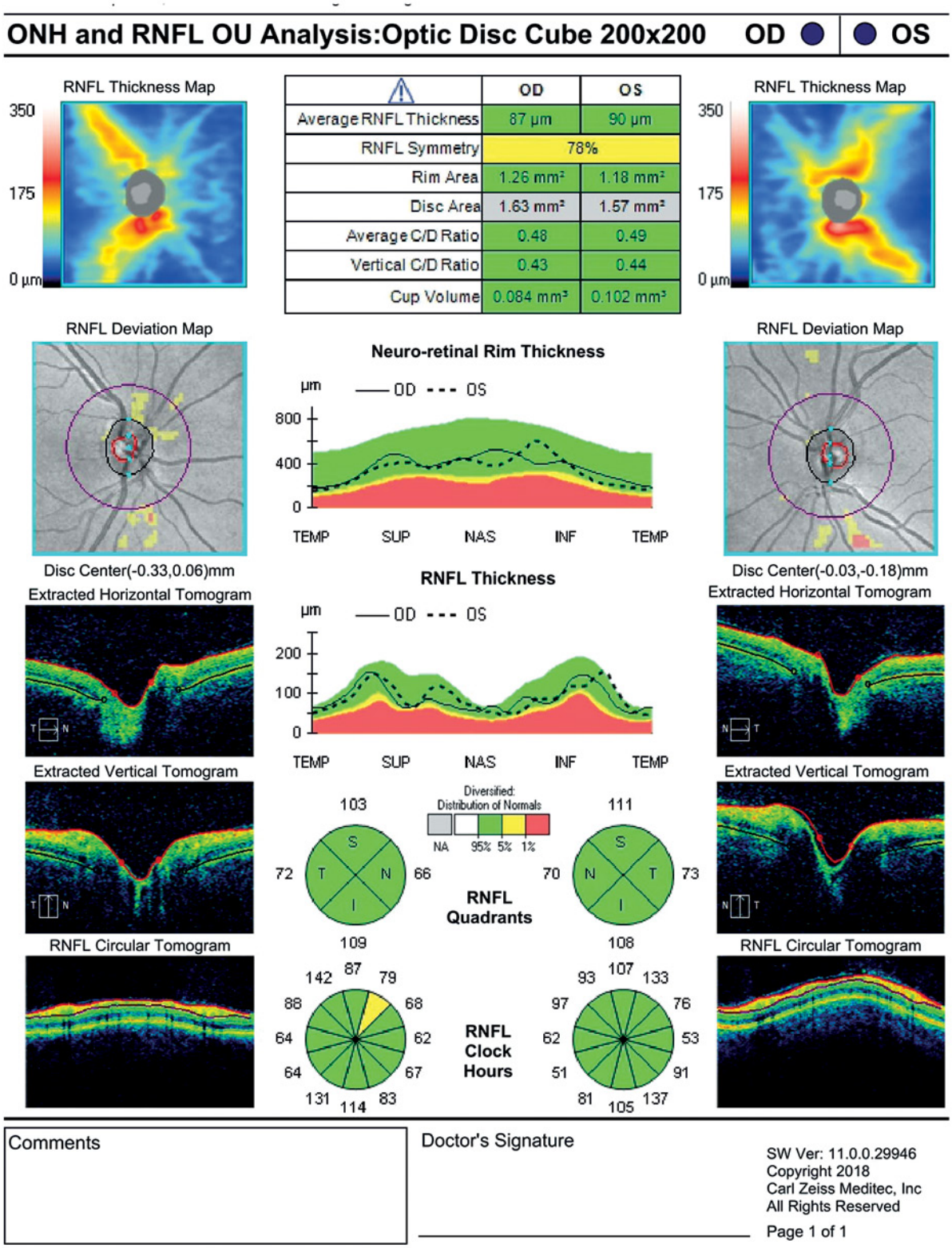


Figure 3. **A)** OCT image of the anterior segment of the right eye. **B)** OCT image of the anterior segment of the left eye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RNFL Symmetry 78%
Rim Area 1.26 mm² 1.18 mm²
Disc Area 1.63 mm² 1.57 mm²
Average C/D Ratio 0.48 0.49
Vertical C/D Ratio 0.43 0.44
Cup Volume 0.084 mm³ 0.102 mm³
RNFL Deviation Map
Neuro-retinal Rim Thickness
µm
OD OS
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400
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TEMP SUP NAS INF TEMP
Disc Center(-0.33,0.06)mm
Extracted Horizontal Tomogram
RNFL Thickness
200
100
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NA 95% 5% 1%
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RNFL Quadrants
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Figure 4. ?????

ments causes separation of the pigmented and nonpigmented epithelia of the ciliary body, resulting in cyst formation. The other theory suggests that cysts are formed through the proliferation of the pigmented and nonpigmented ciliary epithelia and the iris pigment epithelium. The former theory is supported by the observation that both the size and number of cysts decrease with age, which can be attributed to the weakening of the ligaments holding the lens. Another argument is that the cysts are commonly located in the temporal and inferior sectors, where the epithelial junction in the ciliary body is embryologically the weakest [6, 8].

In their 2018 paper, Tiago Ribeiro Schmalfuss, Egidio Picetti, and Helena Messinger Pakter reviewed various available treatment modalities. A total of 19 published articles were included in the review, of which 13 described cases of binocular cysts, and 7 concerned monocular cysts [5, 12-17, 19, 20]. The group of patients with PPI caused by iris and ciliary body cysts comprised 8 cases: 6 cases of iris and ciliary body cysts in both eyes and 2 cases with unilateral lesions. YAG laser iridotomy was performed in all the patients. Open configuration of the filtration angle was achieved in just 2 cases (unilateral cysts) [5, 13]. In all other cases, neither an opening of the filtration angle nor a decrease in the IOP was noted. One patient additionally underwent laser iridoplasty, initially achieving IOP normalization, iris repositioning, and cyst atrophy. After 6 months, though, the cysts recurred and the IOP rose again [5, 14]. In yet another case, after the failure of iridotomy and iridoplasty, phacoemulsification was performed. The patient achieved IOP control and angle opening [5, 15]. Another patient underwent iridotomy and argon laser peripheral iridoplasty, but no change in angle conformation was achieved. Since the patient reported no symptoms, and there was no glaucoma-related damage with only a slight increase in the IOP, it was decided that the patient would be followed up without any other interventions [5, 16]. Another patient, presenting with bilateral cysts of the iris and ciliary body, elevated IOP and open angle, was treated with Nd:YAG laser and iridocystotomy followed by selective laser trabeculoplasty. No further treatment was needed [5, 17]. The authors of this article hypothesize that all laser iridotomy procedures were performed as the first-choice treatment because UBM had not been conducted before to determine the mechanism of angle closure [5]. In patients with iris and ciliary body cysts, Nd:YAG laser iridotomy is not regarded as an effective treatment method [3, 5, 18].

The inefficacy of Nd:YAG laser iridotomy was also reported by Crowston *et al*. who described argon laser peripheral iridoplasty or Nd:YAG laser iridocystotomy as effective treatment options [3, 11]. YAG laser cystotomy may cause pigment dispersion and, consequently, cyst recurrence or inflammation [3, 10, 11]. Choudhari *et al.* describe argon laser iridoplasty as a safer and more effective treatment option [10]. Ravi *et al*. also report that argon laser therapy is a more successful therapeutic modality [18].

In contrast, Baba *et al*. report on the case of a 38-year-old woman who achieved no decrease in the IOP after argon laser iridotomy. IOP normalization was achieved after the initiation of pilocarpine treatment [12]. Topical antiglaucoma agents such as timolol or pilocarpine also lead to a reduction in intraocular pressure [18]. In the reviewed papers, 4 patients received timolol after failed YAG laser iridotomy, achieving a good therapeutic effect [1, 5, 14, 19, 20], while another patient was prescribed pilocarpine, which also led to IOP normalization [5, 12].

CONCLUSIONS

Iris and ciliary body cysts are a far more common condition than the detection rates might suggest [1, 2, 4, 6, 9]. In patients diagnosed with ocular hypertension and a narrow filtration angle, further diagnostic workup for pseudoplateau iris, secondary to iridociliary cysts, is particularly important. Gonioscopy is an essential initial diagnostic modality in patients with ocular hypertension, but UBM remains the primary method of ophthalmic examination [5]. Pseudoplateau iris resulting in elevated intraocular pressure can be treated by various modalities including topical medicines (β-blockers and pilocarpine), laser treatment (YAG iridotomy, argon iridoplasty, iridocystotomy), and surgical methods (phacoemulsification of the lens). However, these methods do not always yield good long-term outcomes because of an increase in the number and size of the cysts in both structures discussed.

DISCLOSURE

The authors declare no conflict of interest.

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