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Anterior Segment Optical Coherence Tomography in Pigment Dispersion Syndrome with and without Glaucoma

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Abstract

Purpose: To share initial experiences in the early diagnosis and management of pigment dispersion syndrome (PDS) and pigmentary glaucoma (PG) using optical coherence tomography (OCT) and to demonstrate the effectiveness of laser iridotomy in altering the iris configuration as a preventive measure against the progression of PDS to PG.

Observations: In this study, optical coherence tomography (OCT) was instrumental for the early diagnosis and monitoring of treatment efficacy in pigment dispersion syndrome (PDS) and pigmentary glaucoma (PG). Pre- and post-laser iridotomy OCT images demonstrated significant changes in iris configuration from concave to a more flattened profile, alongside a reduction in pigment dispersion. These findings highlight the effectiveness of laser iridotomy in modifying the anatomical contributors to PDS and PG and suggest its potential in preventing the progression from PDS to PG. The ability of OCT to visualize these changes emphasizes its value not only in diagnosing but also in guiding the management of these conditions, reinforcing the importance of early intervention.

Conclusion and Importance: The findings suggest that OCT is a valuable tool for the early detection of PDS and PG. Laser iridotomy proves to be an effective intervention for altering iris configuration, thereby reducing pigment dispersion and potentially preventing the progression of PDS to PG. Further studies are recommended to validate these results and explore the long-term benefits of early diagnosis and intervention in PDS/PG management. Anterior segment optical coherence tomography has the potential to assist in the diagnostic enhancement of individuals with suspected or diagnosed glaucoma, serving as an adjunct to gonioscopy. This tool is especially valuable for young suspects and patients, and can be used in patient education by displaying the shape of the chamber angle through an image.

Keywords: *Glaucoma; Pigmentary Glaucoma; AS-OCT; Secondary Glaucoma; Anterior Chamber Angle; Juvenile OpenAngle Glaucoma*

Abbreviations

OCT: Optical Coherence Tomography; AS-OCT: Anterior Segment Optical Coherence Tomography; PDS: Pigment Dispersion Syndrome; PG: Pigmentary Glaucoma; UBM: Ultrasonic Biomicroscopy; SLT: Selective Laser Trabeculoplasty

Introduction

Glaucoma is one of the leading causes of irreversible blindness in the world [1] and its importance has increased when considering the total number of blind people in the world over time [2]. It is a multifactorial disease whose main characteristic is the progressive loss of retinal ganglion cells associated with progressive loss of the visual field [3]. In most cases, this loss of visual field is associated with high intraocular pressure [4].

One of the biggest challenges in glaucoma is its early diagnosis. Optical coherence tomography (OCT) has been widely used by ophthalmologists to improve the diagnosis of glaucoma and other eye diseases. OCT makes it possible to diagnose the disease before the visible alteration to the perimetry and even providing data for diagnosis beyond ophthalmology [5].

Pigment dispersion syndrome (PDS) and pigmentary glaucoma (PG) are part of the spectrum of the same disease, being a subtype of glaucoma in which treatment with laser iridotomy may be indicated [6,7]. This condition affects both sexes, being slightly more frequent in middle-aged men [8]. Its main cause is the accumulation of pigments in the trabecular meshwork, which causes an increase in the resistance of the drainage area and the consequent increase in intraocular pressure [9].

Pigmentary glaucoma is considered a secondary glaucoma [10,11]. Classically, pigmentary glaucoma presents some typical signs: Krukemberg's spindle, hyperpigmentation of the camerular sinus, and concave iris. The typical iris shape that happens in PDS/PG is best seen by ultrasonic biomicroscopy (UBM) [12]. This detection is essential to confirm the diagnosis of this disease since it explains its pathophysiogenesis. UBM studies have enriched the knowledge about this disease, demonstrating the friction between the iris and the anterior capsule of the lens as a source of pigment dispersion [9,13,14].

It is already known that in PDS/PG there is an internal pressure gradient in the eye. This difference is expressed in the higher pressure in the anterior chamber than in the posterior chamber. The iris has a malleable tissue, and its shape modification is often observed in association with morphological and functional changes, as often occurs in the swelling of the lens, a condition in which anterior convexity of the iris occurs [15].

It is estimated that 20% of people with pigment dispersion syndrome develop pigmentary glaucoma [16]. In addition, the accommodation process causes an increase in the concavity of the iris, which is prevented by iridotomy [17].

OCT is a non-invasive, high-resolution technique that allows you to visualize the structures of the anterior segment of the eye, including the iris and camerular sinus. Although OCT is commonly used in the diagnosis of pigmentary glaucoma, its role has been to investigate the retinal nerve fiber layer and retinal ganglion cells [18-21]. In addition, with the limited use of gonioscopy [15] is probably an underdiagnosis of PDS/PG. It has been shown that the OCT parameters of the optic nerve head and retinal nerve fiber layer have significant differences between groups of people with and without glaucoma [22]. However, OCT has not yet been routinely used in the investigation of the camerular sinus. In our routine, we used the following standard of investigation of glaucoma in phakic patients: the 3 studies currently available in OCT devices:

1. Study of the temporal cameral sinus;
2. Study of retinal nerve fiber layer;
3. Study of retinal ganglion cells.

In this way, we take advantage of all the potential of the device for the benefit of the patient's assertive diagnosis. The detection of the concave iris acquires special importance when we know that in some cases of PDS/PG the deposit of iris pigment granules in the corneal endothelium may not be clinically detectable [23,24]. In the Afro-descendant population, in the PD/PG there is more pigmentation of the lens than deposition in the endothelium, without the formation of the Krukemberg 's spindle [24].

Aim of the Study

The aim of this article is to present our initial experience with the early diagnosis of PDS/PG using OCT, as well as to present the change in the pattern of OCT of the camerular sinus after treatment with laser iridotomy. This is a clinical case series report, using spectral domain OCT as the main diagnostic method.

Materials and Methods

All examinations in this study were performed on the same optical coherence tomography (OCT): CirrusTM HD-OCT Model 400 (Carl Zeiss Meditec, Jena, Germany). All patients were treated at the Clínica do Olho in Salvador, Bahia State, Brazil. They were evaluated between September 2016 and February 2023 by the same ophthalmologist.

All laser treatments were performed on the same equipment: Lightmed Lighlas Yag/SLT Deux combination system (Lightmed Corporation, CA, USA).

Clinical Case Reports

For comparing the images that we will present below, we considered the normal AS-OCT pattern in this study to be the following image.

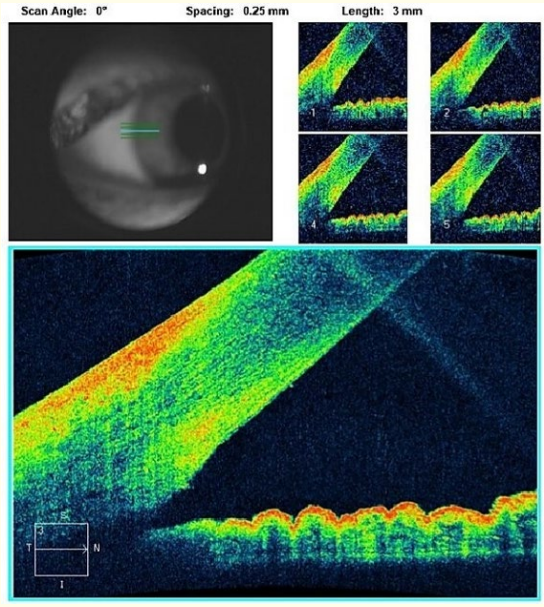

Figure 1: *Normal AS-OCT. Note that there is no concavity or anterior or posterior direction of the iris.*

The following clinical cases were the first in which we detected the iris pattern through OCT, as well as its modification after iridotomy. There are other cases in which, at the time of this publication, they are under monitoring and documentation. Our intention is to initially present a clinical case series report of three patients diagnosed with PDS or PG. We intend to publish the long-term evolution in the next articles.

Case Report 1

A 54-year-old brown patient came to us in December 2019 to obtain a second opinion on his glaucoma treatment, which was done with Bimatoprost eye drops. The patient reported a positive history of diabetes since the age of 50, controlled with the use of Metformin. From family history, it was reported a mother with "suspected glaucoma" and a deceased brother with a history of blindness associated with uveitis. After thorough clinical evaluation and measurement of the visual field, Daily Intraocular Pressure Curve and OCT glaucoma, we suspected pigment dispersion syndrome.

In the OCT evaluation, we were struck by the posteriorly directed iris associated with the open angle more than usual in grade IV. Subsequent gonioscopy showed an open angle, grade IV, anterior concavity and trabecular nigra pigmentation. On biomicroscopy, we can see Krukenberg's spindle, as seen in the following image (biomicroscopy of right eye).

Figure 2: *Biomicroscopy photograph of the anterior segment of the right eye of the patient in which we can identify the Krukemberg spindle.*

We then instituted the following therapy: Laser iridotomy in both eyes to eliminate the pressure gradient between the anterior and posterior chambers, eliminating the source of pigment dispersion. After one month, we performed the replacement therapy of prostaglandin (Bimatoprost) by selective laser trabeculoplasty (SLT), which served the correction of the patient's red eye complaint.

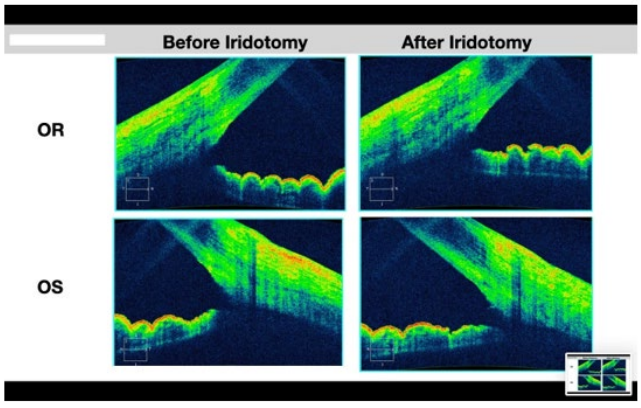

Figure 3: *Assembly with AS-OCT images of the patient in which we can easily visualize the reduction of the posterior direction of the iris and its rectification after iridotomy, as well as the elimination or reduction of its concave shape.*

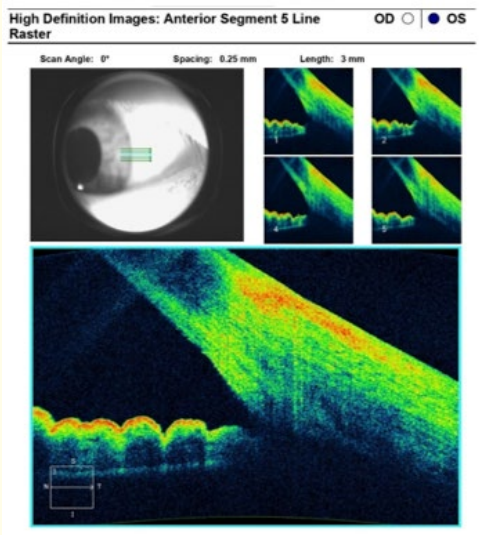

Figure 3.1: *Update of the examination of the anterior segment of the left eye of the patient, in August-2022, demonstrating that, in this case, the iris rectification process continues over time.*

Case Report 2

A 43-year-old black patient, highly myopic, whose father has a history of glaucoma, came to us in March 2020 for a routine consultation. During anterior and posterior segment biomicroscopy, we identified an "extremely wide" anterior chamber and Krukemberg's spindle in both eyes and papillary excavations estimated at 0.9 in the right eye and 0.85 in the left eye. Intraocular pressure (IOP) 16/14 mmHg (OD/OS). We did a minimal complementary investigation with OCT and visual field; the images of the previous segment are as follows.

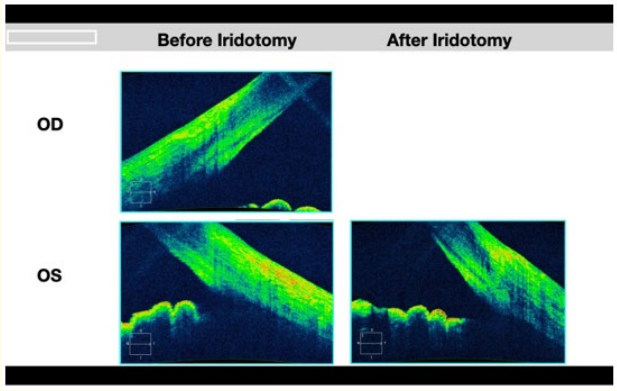

Figure 4: *Assembly with AS-OCT images of the patient in which we can easily visualize the reduction of the posterior direction of the iris and its rectification after iridotomy in the left eye, as well as the elimination of its concave shape.*

We do not yet have images of the right eye after iridotomy as of this publication. The iris is so posterior that it was not possible to capture its full image in the standard capture positioning prior to iridotomy in the right eye.

Case Report 3

A 28-year-old white patient was referred by an ophthalmologist colleague for treatment with selective laser trabeculoplasty (SLT). The treatment of this patient was done with laser iridotomy in March 2020 in the right eye and in January 2021 in the left eye, in addition to SLT in both eyes in March 2020. The peculiarity of this individual, in addition to the modification of the iris pattern, is that we can clearly perceive the progression of the ganglion cell lesion in the comparison with the 2020 exam with the 2016 exam, prior to the iridotomy treatment, as shown in figure 6 and 7. Prior to laser treatment, this patient had been treated exclusively with Latanoprost (Xalatan) eye drops and a fixed combination of Timolol and Dorzolamide (Drusolol).

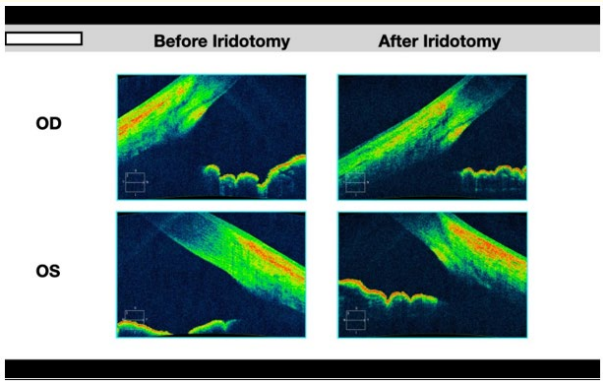

Figure 5: *In the images above, we can easily see the reduction of the posterior direction of the iris and its rectification after iridotomy, as well as the elimination or reduction of its concave shape.*

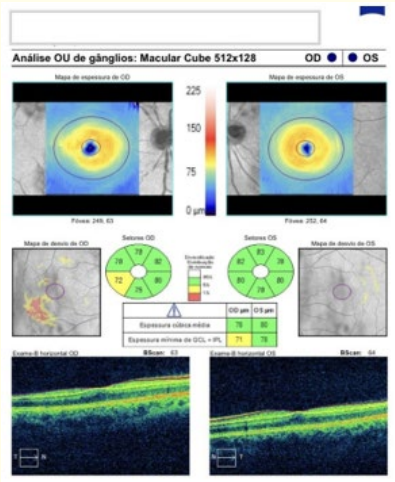

Figure 6: *Ganglion cell study in September 2016. Compare with image 7.*

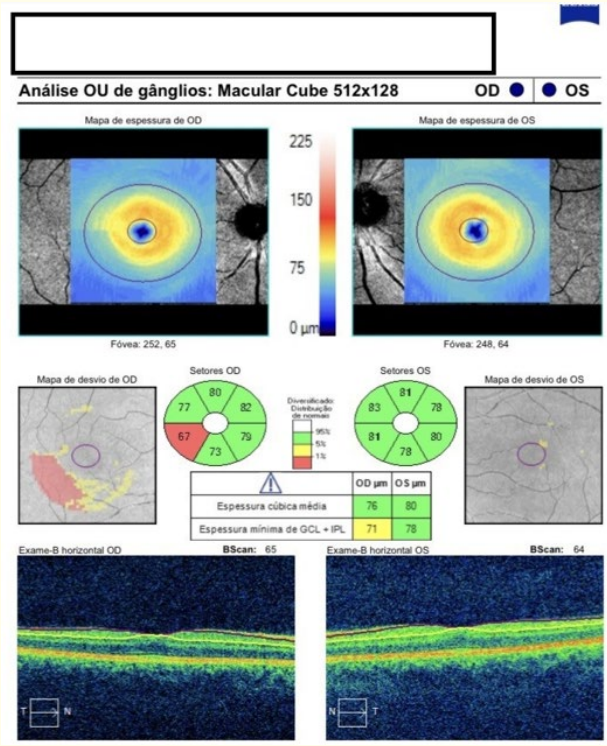

Figure 7: *Ganglion cell study in January 2020. Observe the progression of the lesion in the right eye during the exclusive treatment with eye drops.*

Discussion

In 1994, Costa and Spaeth were the first to demonstrate that Peripheral iridotomy can promote a change in the configuration of the peripheral iris and reduce pigment dispersion in patients with pigmentary glaucoma. In the same article, authors report that this treatment was proposed in 1991 at the meeting of the American Glaucoma Society. However, at the time, optical coherence tomography (OCT) technology did not yet exist [25].

OCT is a non-invasive, high-resolution technique that allows the visualization of the structures of the anterior segment of the eye, including the iris and trabecular meshwork. This study described a series of three clinical cases (five eyes) of PDS/PG in which we used the laser iridotomy procedure to break the internal pressure gradient. This mechanism has been proposed as the main cause of pigmentary dispersion, caused by the friction of the iris near the lens [26]. Previous studies using UBM have shown changes in the shape of the iris in people with PDS/GP [14].

In addition to gonioscopy, both UBM and OCT can be used for complementary evaluation of the anterior segment of the eye [27]. In this study, we used OCT as a tool to evaluate the shape of the iris before and after iridotomy, due to the practicality of the technique and its availability in the medical service responsible for the evaluations.

Although the parameters of the retinal nerve fiber layer are considered to have the greatest potential for differentiation between people with glaucoma and healthy people [22], the shape of the iris may have implications for the genesis of glaucoma, such as that observed in PDS/PG, but it is not a usual parameter for the diagnosis of glaucoma, to date. As shown in the previous images, the rectification of the iris occurred sometime after the laser iridotomy, a fact demonstrated by the OCT images of the anterior segment.

OCT has revolutionized diagnosis in ophthalmology and its use has grown continuously worldwide. This technology has also been widely used for complementary research in glaucoma [28]. Classically, ultrasonic biomicroscopy (UBM) has been used to investigate the anterior segment of the eye in patients with PDS/PG. UBM makes the concave shape of the iris evident in these patients [29]. However, it is a more complex technique with less availability within the Ophthalmology service.

As is already known in the public domain, in PDS/PG there is an internal pressure gradient which leads to variation in the shape of the iris and constitutes its pathophysiological mechanism. More specifically, the mechanism of action of pigmentary glaucoma is the friction of the iris on the anterior capsule of the lens, which causes the release of pigments towards the trabecular meshwork, as well as other structures such as the lens and retina [26].

As with UBM, the use of anterior segment OCT can help in the identification of pigmentary glaucoma early with the advantage of greater practicality. This allows the treatment of the disease in a more specific way, through the application of laser iridotomy and elimination of the difference in the pressure gradient of the anterior and posterior chambers of the eye, eliminating irido-crystalline friction. It is also important to highlight the high rate of retinal detachment and in these patients, 12%, which makes it even more important to identify the diagnosis and eliminate its pathophysiological mechanism [30].

In our understanding, the study of the camerular sinus by OCT has been undervalued. Many physicians consider the findings of gonioscopy to be the only source of information about the camerular sinus. Although Gonioscopy is the gold standard in the analysis of the drainage system, OCT brings us complementary information about this system [31].

The use of AS-OCT routinely in patients suspected of having glaucoma or diagnosed with glaucoma could better guide the ophthalmologist in identifying the type of glaucoma, since it is a rapid complementary test with availability in OCT equipment, but often forgotten. An important fact is that OCT detects the angle closure more often than gonioscopy [32].

Moreover, the signs of pigmentary glaucoma may reduce over time; very elderly patients may lose the Krukemberg spindle progressively. However, the anterior concavity and posterior direction of the iris are still present, unless this patient has undergone cataract surgery or received laser iridotomy treatment.

From another perspective, the identification of the main causal factor of glaucoma and the treatment of its underlying cause will make the therapy better targeted. Based on the cases presented in this study, we understand that it is possible to prevent the use of multiple drugs to control intraocular pressure in patients with PG. To reach this conclusion, long-term prospective studies are needed in order to follow patients treated with prophylactic iridotomy, compared with those who started their treatment with eye drops. Once PDS/PG is diagnosed, laser iridotomy treatment can be instituted, with the aim of eliminating the source of pigment dispersion, since this therapeutic resource is accessible to physicians and patients almost all over the world.

In this study, we did not include the measurement of the distance between the trabecular meshwork and the iris due to the limitations of the device used and because it was an observational study. We consider that making these measurements would bring more complications than usefulness in identifying the shape and direction of the iris, since the concave shape can be easily seen in cases of PDS/PG. We also consider it unnecessary to make limbo-to-limbo images to illustrate the finding.

In our opinion, the identification of pigment dispersion syndrome and the early performance of laser iridotomy allows fewer patients to become ill with Pigmentary Glaucoma, since its pathophysiological cycle is broken. We estimate that investing in diagnosis in PDS/PG prevents individual and public expenditures on the treatment of this disease. Early identification of PDS can prevent the formation of PG and retinal detachment and its consequent effects on the individual and society.

OCT can help ophthalmologists identify cases of pigment dispersion syndrome and pigmentary glaucoma early. We intend to make new publications in order to share the evolution of the iris shape over time in patients treated with laser, of whom we have the record of anterior segment OCT.

The results of this study suggest that laser iridotomy may be an effective technique to correct iris deformity in patients with PDS/PG by reducing the internal pressure gradient of the eye and improving aqueous humor drainage. In addition, OCT was useful to evaluate the efficacy of the procedure in eliminating the internal pressure gradient in the eyes affected by PDS/PG and in monitoring the patients' response to treatment over time.

Although the study described only three cases, the results are encouraging and suggest that laser iridotomy may be an effective technique in cases of PDS/PG. However, further studies are needed to evaluate the efficacy and safety of the technique in a larger number of patients.

In summary, the use of OCT to verify the result of laser iridotomy is a valuable tool in the management of PDS/PG. In addition, the study describes successful cases in the rectification of the iris after iridotomy, using OCT as a tool to record the anatomical change.

Conclusion

In this article, we present a series of clinical cases in which we used OCT as a way to identify PDS/PG, as well as the result of iridotomy on the anatomy of the anterior segment. The concave iris sign is an important sign for the diagnosis of this type of glaucoma. The AS-OCT provides important information in the diagnosis of PDS/PG. In our opinion, this investigation should become standard and join the study of the optic disc and macula in the investigation of glaucoma.

Conflicts of Interest

The following authors have no financial disclosures: Honassys R. Rocha Silva, Renato Galao Cerquinho Leca and Luciano Rabello Netto Cirillo.

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