Acute Onset of Ocular Hypertension in Myopic Patient with Pigment Dispersion Syndrome Mimicking Uveitis

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Abstract

Purpose: To report the acute onset of ocular hypertension in myopic patient with pigment dispersion syndrome.

Methods: A 40-year-old male patient, suffering from unresponsive chronic open-angle glaucoma in multi drugs-therapy, underwent complete ophthalmological evaluation at our department in order to have a second opinion on his pathology.

Results: Visual acuity was 20/20 in both eyes, gonioscopy showed the presence of iris pigment in the iridocorneal angle (Grade 4 Shaffer classification) and an iris anterior concavity. The intraocular pressure (IOP), in spite of the topical therapy, was 28 mmhg; the anterior chamber was normal without Tyndall phenomenon, while the retina and optic disc were normal. The patient underwent a bilateral YAG laser peripheral iridotomy and a bilateral Selective Laser Trabeculoplasty (SLT). After a two-year- follow up, the visual acuity is stable and the IOP is set on 12 mmhg in both eyes (instilling one drop of 10 daily).

Conclusion: In the myopic patient the pigmentary glaucoma is a complication which has to be and often causes misdiagnoses.

Keywords: Myopic; Pigmentario glaucoma; IOP; Ocular hypertension

Introduction

Pigmentary glaucoma (PG) is the most common cause of glaucoma in young adults under 40 years of age [1]. This kind of glaucoma is associated with the release of pigment granules arising from the iris, subsequently dispersing throughout the anterior segment and possibly occluding the trabecular meshwork. Up to 50% of individuals affected by this kind of pigment dispersion develop glaucoma [2], and the risk of developing pigmentary glaucoma in the pigment dispersion syndrome is 10% after 5 years and 15% after 15 years. Young, myopic men are most likely to have pigmentary glaucoma (rif). The intraocular pressure (IOP) greater than 21 mmHg at initial examination is associated with an increased risk of conversion in (pigmentary glaucoma) PDG [3].

Some studies suggest that the accumulation of pigment in the trabecular outflow pathways directly increases the IOP [3,4]. Other studies suggest that pigmentary glaucoma is the result of a generalized mesodermal dysgenesis of the eye that results in abnormal development of the anterior segment [5]. The release of the iris pigment is the result of a concave sagging in the peripheral iris which brings the posterior surface into contact with the lens zonules (Figure1). The friction between the two surfaces causes the release of the iris pigment; this phenomenon is clearly visible by positioning a slit lamp beam in the anterior chamber, over the pupil: the resulting transillumination defects form a characteristic pattern of slits in the iris that corresponds to the location and radial arrangement of the zonules.

Figure 1: Shows Anterior rotation of the ciliary bodies.

Like the majority of human diseases, PG and PDS show a family aggregation that does not usually follow Mendelian patterns but is caused by an unknown number of multiple genes, usually interacting with various environmental factors. A single susceptibility locus to
PDS has been mapped on chromosome 7q35-q36 but the candidate gene has not been identified so far. The pathogenesis of PG has not been characterized yet, and it has been hypothesized that a major role is exerted by the presence of the iris concavity, which leads to irido-zonular friction and iris pigmented epithelium disruption. A new Italian study supports the hypothesis of the presence of defects in the stromal iris elastic fibres linking to the mutation of the lysyl oxidase like protein-1 (LOXL1) gene, already demonstrated in the pseudoesfoliation syndrome, which could justify the presence of the increased iris concavity frequently presented in PDS/PG [6].

Materials and Methods

The following clinical observation is an example of atypical onset of glaucoma related to pigment dispersion and reverse pupillary block. After a gonioscopy and dynamic contour tonometry (DTC), we got the right diagnosis and therefore we performed ultrabiomicroscopy (UBM) to confirm the reverse pupillary block. After that, we performed bilateral YAG laser peripheral iridotomy (LPI) and few months later the patient underwent to selective laser trabeculoplasty (SLT) in order to reduce intra ocular pressure (IOP) [7-10].

A 40-year-old man came to Humanitas Research Hospital-Glaucoma Center in the December 2012 because of his multidrugs unresponsive, open-angle, chronic glaucoma [11]. The patient's past medical history was negative for any systemic or ophthalmic disease. He had undergone a bilateral photorefractive keratectomy (PRK) for 5 diopters myopia ten years before (2002), and in October 2012 he scheduled ophthalmological evaluation because of high pain and blurred vision in the right eye. [12-15] The diagnosis was hypertensive uveitis (Anterior chamber tyndal 1+, IOP 24 mmHg, vitreo and retina normal), and the therapy prescribed was: Timolol 0.5% (1 drop, Dorzolamide, Dexamethazone, Cyclopentolate eye. The patient was also requested to undergo to a rheumatological visit (which resulted to be negative). 1 month (November 2012) after the prescribed therapy, the clinical situation in the right eye was not resolved (anterior chamber Tyndal: 1+, IOP 34 mmHg). In November 2012, due to an herpetic ocular trabeculitis hypothesis, the ophthalmologist added the prescription of ganciclovir ophthalmic gel and Acyclovir 200 mg. The computerized Visual Field was bilaterally normal (Figure 4: EASYFIELD–Oculus Inc.-Arlington WA, USA.) [16-18]

Figure 2: Anterior concavity of the iris or posterior convexity.

In December 2012 the patient came to our department searching for a second opinion (Figure 3).

He underwent a new complete ophtalmological evaluation, a gonioscopy, a Goldmann applanation and Pascal applanation tonometry, a phenylephrine test, the OCT of retina and optic nerve (Figure 5), the Ultrabiomicroscopy (UBM) of the anterior chamber and of the iris angle (Figures 1 and 2) [19,20].

After these exams, he also underwent a bilateral YAG laser peripheral iridotomy (January 2013) and a bilateral Selective Laser Trabeculoplasty (SLT) in April 2013.
**Results**

The ophthalmological evaluation showed a clear cornea and uncorrected best visual activity of 20/20. Krukenberg spindle CA: wide (Von Herick 4), flare 0, cells 0.

The IOP was 12 mmHg in both eyes with Goldmann tonometer (GAT), but the measurements with Pascal tonometer (DCT) were 27.5 mmHg in right eye and 26.3 mmHg in the left one.

The phenylephrine eye drops test was positive after 30 minutes showing (with the slit lamp observation, an anterior chamber pigmentation) Tyndall increase from 0 up to 2+

The gonioscopy showed an iridocorneal angle Shaffer 4 grade, pigment deposit was 3+, and also qualitative evaluation? Gonioscopy? Showed an iris anterior concavity (Figure 4).

The therapy administrated was set as follows: Bimatoprost 0.1% eye drops qD (once daily) OU (Lumigan 0.1%), Timolol 0.5% + Brimonidine eyedrops (twice daily) BID OU, Pilocarpine 2%. One week after the SLT, the IOP measured with DCT was: 21.2 mmHg in the right eye and 20.1 mmHg in the left eye. One month later, the IOP measured with DCT was 15.1 mmHg in both eyes.

Ten days after, the IOP was 16.2 mmHg in the right eye and 17.1 mmHg in the left one, measured with DCT.

Currently, 4 years after LPI and SLT, the topic therapy is: Lumigan 0.1 eyedrops qD OU and Combigan eyedrops BID OU and the IOP is in good compensation (18.2 in right eye and 17.9 in left eye, measured with DTC.) Doubling Frequence Thescology perimetry (Carl Zeiss Meditec, Dublin CA USA) and Nerve Fiber Layer OCT are unremarkable in both eyes.

**Discussion**

The decrease of visual acuity, the sight of colored halos around light sources (due to a mild corneal edema) and slight eyelid soreness are symptoms and clinical signs of ocular hypertension. This clinical manifestation can be accompanied with the Tyndall effect. It is very important to describe the grade of flare in the anterior chamber and, moreover, to specify whether the Tyndall is due to blood cells, inflammatory cells or melanin pigment. Sometimes the pigment dispersion syndrome can occur with a massive presence of melanin pigment in the anterior chamber due to a massive release of pigment from the posterior face of the iris rubbing against zonular fibers.

In the patient who we present in this report, the pigment dispersion and the pigmentary Tyndall have been confused with inflammatory cells, resulting in wrong diagnosis of uveitis. Hence, the erroneous management of the patient with therapy and investigation was not appropriate. Phenylephrine eye drops test, creating wide mydriasis, increases the zonula-iris contact, rising the release of pigment in both posterior and anterior chambers. This condition is repeatable even in conditions of increased systemic secretion of adrenaline, for example in the case of submaximal or prolonged physical exercise (rif.) [22,23].

When the iris pigment enters massively the anterior chamber, it deposits on the endothelium, due to the convective motions of the AC, centrally and vertically, creating the Krukenberg spindle, undetected in this case. When glaucoma is suspected and especially when the patient has ocular hypertension, a gonioscopy is mandatory, in our opinion.

In the case presented, the gonioscopy, which was never performed in the first three months after the onset of visual disturbances, was decisive for understanding the cause of the pigment dispersion in AC, which was misdiagnosed as uveitis.

This simple test gave us two main indications: 1) Presence of iris convexity directed posteriorly, 2) Quantity of pigmentation on the trabecular meshwork (TM).

The melanin granules scattered in the anterior chamber are deposited onto the endothelium of TM, making the aqueous outflow difficult towards the Schlemm canal. These melanin granules are released after rubbing between the zonular fibers and posterior iris. The iris can fold posteriorly, creating a valve mechanism in which the IOP is higher in the anterior chamber than in the posterior chamber, causing the "reverse pupillary block".

This condition is well evident when performing an UBM, which can show:

1) Anterior rotation of the ciliary bodies (Figure 1)
2) Anterior concavity of the iris or posterior convexity (Figures 1 and 2)
3) Microcysts in the ciliary bodies (Figure 2)
4) Contact between the iris and the zonular fibers. (Figures 1 and 2)

Another reason which made this clinical case challenging was related to corneal biomechanics: the patient, with previous mild myopic defect, had undergone bilateral PRK ten years before the onset of ocular symptoms.

In case of previous PRK, the Goldmann tonometry and the pneumotonometer must be replaced by DC or Pascal tonometer, which is not influenced by the thickness of the cornea or corneal biomechanics changes after PRK. We used the Pascal tonometer (DCT) (Ziemer Ophthalmic System AG, Switzerland.) which showed an ocular pressure different from the Goldmann applanation tonometer. Both the Goldmann tonometry and pneumotonometry showed a large underestimation of his the patient's ocular pressure. The use of a
piezoelectric sensor which measures the value of the IOP regardless of biomechanical corneal thickness is mandatory for diagnosis of hypertension in pupillary reversal block in the patient who has undergone refractive surgery [2]. After the gonioscopy and dynamic contour tonometry (DTC), we were referred to the right diagnosis, also helped by the ultrabiomicroscopy (UBM) which confirmed the reverse pupillary block. After that, we performed bilateral YAG laser peripheral iridotomy (LPI); this procedure reduces the iris-zonular fibers rubbing [24] and helps to remove the reverse pupillary block and pigment production, which are both responsible for ocular hypertension [3].

Few months later the patient underwent selective laser trabeculoplasty (SLT): this procedure retrieves macropores in the trabecular meshwork.

The subsequent pigments phagocytosis allows the removal of the melanin in excess on TM and finally the increase of intra ocular pressure (IOP) reduction.

Topical therapy focuses primarily on the prescription of a prostamide, which increases the opening of the trabecular meshwork and conveys the aqueous humor through the uveo-scleral outflow pathway.

The use of topical corticosteroid eye drops possibly worsens the eye condition, elevating the IOP in this myopic steroid-responder patient.

**Conclusion**

In this case we can highlight different errors of management of this unresponsive glaucoma patient: the patient’ ophthalmological past history was not sufficiently considered; the misinterpretation of the anterior chamber dispersive syndrome hypothesized as anterior uveitis; the IOP should have been checked even with the DTC or Pascal tonometer.

The gonioscopy, which in case of glaucoma or uveitis should always be performed, together with UBM, helped us to address the right aethiologic diagnosis and, finally, to resolve the pupillary block.

**References**