CLINICAL CONSULTATION

The Clinical Consultation section is offered as an educational exercise under the direction of Etty Bitton, OD, MSc, École d'optométrie, Université de Montréal, Montreal, Quebec. It is intended to be a continuing forum wherein we as optometrists share our views and opinions with our colleagues. In this issue, Richard Trevino, OD, provided the case report and the questions that accompany it; Patricia Koester, OD, Gerald S. Komarnicky, OD, and Russell Pearlman, OD, commented on the case.

Intraocular Pressure Fluctuation in a Young Man

History

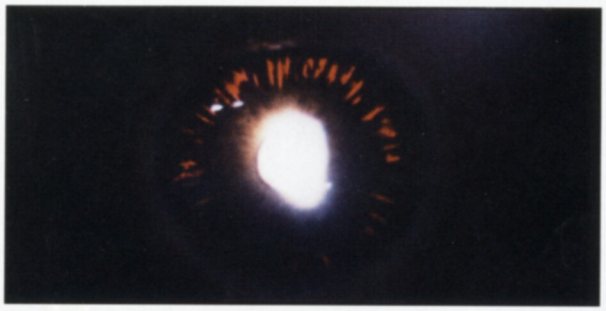
A 32-year-old man presents for a routine eye examination with complaints of blurred distance vision. He is in excellent health and reports that he had undergone an unremarkable eye examination two to three years ago. There is no family history of glaucoma or other significant ocular disease.

Findings

He is corrected to 20/20 in each eye by updating his low myopic correction. Applanation tonometry readings are 30 and 34 in the right and left eyes, respectively, at 9:00 A.M. Biomicroscopy findings include spoke-like transillumination defects in the mid-periphery of the iris and pigment deposition on the corneal endothelium of both eyes (see Figure). Gonioscopy reveals that the anterior chamber angles are wide open with heavy pigmentation of the meshwork in all quadrants of each eye. Ophthalmoscopy through dilated pupils reveals physiologic but slightly asymmetric cups. The cup-to-disc ratios are 0.4 OD and 0.5 OS, with myopic tilting OU.

Treatment

The patient is prescribed 0.25% levobunolol q.d. in the left eye only and instructed to return in two weeks. At this visit the patient reports excellent compliance with the treatment and the intraocular pressure is 20 and 21 in the right and left eyes, respectively, at 6:00 P.M. Perimetry reveals normal visual fields in each eye, although reliability for the left eye is suspect due to several fixation losses. The patient is instructed to discontinue the medication and return in two weeks. At this time tonometry readings are 26 in each eye at 8:40 A.M. Upon direct questioning the patient admits to occasional episodes of transient visual obscurations without pain or headaches. He does not associate these episodes with aerobic exercise or other activities and has no history of migraine.



Spoke-like transillumination defects in the mid-periphery of the iris

Questions

1. How do we account for this patient's widely fluctuating intraocular pressure and apparent lack of response to treatment?
2. Does this patient's condition require treatment? How should this patient be managed?

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This patient presents a text-book example of pigmentary glaucoma. He is a 32-year-old myopic man with mid-peripheral iris spoking, endothelial pigment deposits, pigmentary banding in the trabecular meshwork, and fluctuating pressures which remain above normal. The blurred vision and transient visual obscurations are attributable to the pigment storms associated with pigmentary glaucoma which elevate pressures and result in corneal epithelial edema.

The widely fluctuating diurnal pressure is a result of the mechanical rubbing between the posterior iris and

the anterior zonular bands, causing disruption of iris pigment epithelium that leads to the liberation of pigment granules. The accumulation of pigment granules in the trabecular meshwork leads to a decrease in outflow, thus increasing the IOP.

Beta-adrenergic antagonists, such as levobunolol, are considered the first-line medical treatment of the glaucomas due to their 20–30% pressure-lowering potential. The uniocular administration of levobunolol has been shown to have contralateral pressure-lowering effect in the untreated eye, but the determination of a base line is necessary before accurate pressure-lowering effects can be assessed. If the IOP reading of 26 is taken as base line then the levobunolol was effective in lowering the pressure from 34 to 21. It should be noted that beta blockers, when used in secondary glaucomas, have the potential to exasperate the condition by decreasing outflow.

Preventive treatment of pigmentary glaucoma should be initiated immediately with the goal to reduce iridozonular contact, thus decreasing pigment release. Pilocarpine (Ocusert) will accomplish this goal while minimizing the miotic side effects that are intolerable to younger patients, such as pupillary miosis, induced myopia, accommodative spasm, and possible retinal detachment. Pilocarpine reduces IOP and increases pupillary block which lifts the iris from the zonules. Before and after treatment is initiated, a thorough peripheral retinal evaluation is indicated. Patients should be monitored every three months for the first year to determine stability, and then every six months.

A new treatment option on the horizon may involve peripheral iridotomy to change the iris configuration with accommodation. Long-term clinical trials have not proven effective and a patient profile to determine who would benefit from the treatment has not been identified.

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This young adult would appear to have pigmentary dispersion syndrome (PDS) as characterized by his mid-peripheral iris atrophy and pigmentary loss. His loose iris pigment has been carried by the aqueous convection currents through the anterior chamber and deposited onto the corneal endothelium (Krukenberg's spindle) and onto the trabecular meshwork.

I assume that the investigator ruled out other abnormalities, such as uveitis and uveal cysts that could result in a shower of cellular material, and had differentiated this iris atrophy from pseudoexfoliation syndrome, based on the age of the patient and the location of the iris atrophy.

The tentative diagnosis, based on elevated IOPs and cup asymmetry, was that the patient had pigmentary glaucoma. There was no mention of a proven field loss. Therapy was initiated only to result in an apparent lack of response to treatment.

The question is: Is this a case of pigmentary glaucoma?

PDS occurs equally in myopic males and females and is not, in itself, a high-risk factor for the development of glaucoma. Pigmentary glaucoma had at one time been thought to be a consequence of PDS and assumed that the pigment deposited on the trabecular meshwork obstructed the aqueous outflow, causing an increase in IOP. This, however, has been shown not to be the case as dense pigmentation of the trabecular meshwork can occur in the absence of elevated IOP.

Pigmentary glaucoma occurs more often in young adult myopic males, is seen earlier in higher degrees of myopia, may be autosomal dominant, and is independent of iris color. Pigmentary glaucoma has a low prevalence even in patients with PDS. While its incidence decreases with age, its severity increases. The pigment is only a contributing factor to an underlying primary open-angle glaucoma (POAG) in predisposed individuals

Evaluating IOP is important, but drawing too many conclusions from a single tonometric test is dangerous and can lead to a high rate of false positive and false negative findings. Patients with pigmentary glaucoma do not always respond well to treatment and can display wide fluctuations in IOP.

Patients with PDS should not be hastily diagnosed with pigmentary dispersion glaucoma (PDG). Rather, they should be evaluated for the risk of developing glaucoma by monitoring IOP, iris transillumination, stereo photos of the disk, threshold visual fields, and assessment of the trabeculum through gonioscopy. The

astute clinician would document that the patient had been advised of any significance of his clinical presentation and the need for ongoing evaluations.

If a diagnosis of PDG is made, management of the secondary glaucoma requires treating the underlying disease by following the guidelines for managing POAG. Since this case involves a young adult, beta blockers or levobunolol would be a reasonable first line of attack.

If therapeutic intervention does not achieve the desired goal of controlling IOP and preventing field loss, then argon laser trabeculoplasty or filtration surgery could be alternative therapies especially in older patients.

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The exam findings are consistent with PDS. PDS patients usually have a "larger iris and a mid-peripheral posterior iris concavity that increases with accommodation."[1] This leads to increased iris-lens/zonule contact creating pigment dispersal from the posterior iris, with deposition throughout the anterior chamber typically on the lens, upon the angle structures, and in a vertically oriented pattern on the corneal endothelium (Krukenberg's spindle).[2,3] As in this case, these patients often present with radial iris transillumination defects as well as increased trabecular pigment.[2] It is the obstruction of outflow through the meshwork and resultant meshwork changes[4] that can lead to elevated pressures resulting in PDG. Included in the differential of PDS are trauma, uveitis, intraocular melanoma, and exfoliation syndrome which are excluded here, given the history and examination results.

Therefore, the most likely cause of this patient's increased pressure is PDS. PDG is diagnosed as any other open-angle glaucoma via increased IOP, optic disc changes, and visual field defects. PDG is often associated with younger, mild-to-moderately myopic males and is rare in blacks.[2] It is not uncommon to find widely fluctuating pressures in the PDS and PDG patient.[2] With these IOP changes, the patient may also complain of halos around lights or transient blurring of vision as in this case.[3] Often these symptoms are associated with pupil dilation or after exercise.[5] Although this patient's symptoms were not associated with exercise, it may be of value to check his IOP after exercise to evaluate a potential pattern.[3]

A similar treatment strategy as for POAG can be pursued.[5] Since pressures have been measured with varying results with apparently normal visual fields, I feel that only close monitoring may be needed. Given increasing and consistently elevated pressures, increased symptoms, or changes in the visual field status, therapy should be initiated and followed as with any other glaucoma patient. A notable response to pilocarpine has been reported since it decreases the physical contact between the zonules and the iris.[3] Given that these patients are often younger, miotics are usually not tolerated very well.[5] Of additional concern is the use of latanoprost with its established effects on melanocyte activity.

Until further research is available, initial therapy with a beta blocker or even combination with other pressure-lowing medications is probably a better first-line treatment. It has been reported that laser iridotomy may be effective in these patients given the anatomical characteristics and the resulting so called "reverse flap" pressure gradient that actually elevates the iris from its otherwise abnormally posterior position. As the patient ages, a natural anterior repositioning of the iris may occur, thus relieving the initial precipitating factors of the disease. Regardless, the patient should be educated as to the nature of his condition and its natural course.

References

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