Two Pits in a Pod: Using EDI-OCT to evaluate the lamina cribrosa in a patient with open-angle glaucoma and multiple optic pits

Abstract:
EDI-OCT imaging is used to evaluate glaucoma by examining the thinning and posterior displacement of the lamina cribrosa. This novel multimodal technique is valuable when optic nerve evaluation is complicated by multiple optic pits.

I. Case History:

Patient demographics: 65-year-old Caucasian male

Chief complaint: 65 y/o Caucasian male presented as a new patient to the Optometry clinic for a consult eye exam secondary to hydroxychloroquine therapy for Rheumatoid Arthritis.

Ocular history:
*Multiple optic pits OS
*Advanced open-angle glaucoma OS
*Open-angle glaucoma suspect OD
*Hydroxychloroquine therapy since October 2015
*HX of ectropion surgery
*HX of ocular injuries from fiber glass and gasoline (between 1970-1980) → subsequent finding of the optic pits upon evaluation from fiber glass injury

Medical history:
*Hypertension
*PTSD
*Rheumatoid arthritis
*Current day smoker
*Chronic alcoholism in remission x 27 years

Medications:
*Sertraline
*Prazosin
*Lisinopril
*Sulfasalazine
*Hydroxychloroquine sulfate 200mg BID

Ocular: *Brimonidone 0.1% OS
*Travatan Z OS

Other salient information:
Family HX of glaucoma → mother + father
II. Pertinent findings:

Clinical:
*BCVA 20/20 OD and 20/30+ OS
*Pupils and EOMS: normal
*Amsler grid defects: OD → metamorphopsia to the right of the fixation
   OS → **absolutely scotoma inferior to fixation target**
*Confrontation Fields: OD → WNL, FTFC
   OS → **absolutely scotoma inferior to fixation target** (nose);
   periphery full
*Slit lamp exam: unremarkable; (-)Krukenberg spindle (-)PXF material (-)Iris Transillumination
*IOP Timeline:

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*Optic Nerves: OD 0.6R, healthy rim tissue with distinct margins, no pallor
   OS 0.9V/0.8H → NO rim temporally + **2 optic pits** (1 larger inferior/temporal and 1 smaller temporally), distinct margins nasally
   (+)peripapillary scarring temporally; NO active fluid
*Macula: hard drusen OU + pigment migration OD
*Periphery OD: pigmented operculated hole inferior/temporal; clear OS

Ophthalmic Imaging:
*Fundus photos: **2 Optic pits evident OS**
*Baseline RNFL OCT:
   OD: Robust RNFL 360
   OS: **profound thinning temporally** (25um of RNFL versus norm of 71um);
   All other quadrants robust RNFL OS
*Baseline Posterior Pole OCT:
   OD: Normal foveal contour + drusen subfoveally, band 2-3 disruption inferior to fovea
   (-)SRF (-)IRF
   OS: Normal foveal contour + drusen subfoveally, band 2 disruption parapapillary
   (-)SRF (-)IRF
*Baseline Fundus Autofluorescence (FAF):
   OD: small area of hyperfluorescence inferior to fovea
   OS: large area of hypofluorescence adjacent to ONH temporally, with leading edge of hyperfluorescence
*Baseline HVF 10-2:
   OD: Reliable; no defects
   OS: Reliable; **dense scotoma inferior/temporal + ½ of inferior/nasal field + 1/3 of superior/temporal field**
**Baseline HVF 24-2:**
OD: Reliable, no glaucomatous defects
OS: Reliable; dense paracentral scotoma inferior/nasal

### III. Differential diagnosis:

**Primary/leading:** Advanced open-angle glaucoma (OAG) OS in the setting of congenital optic pits and OAG suspect OD

**Secondary:**
*Uncontrolled POAG OU with persistently elevated intraocular pressures in an already compromised lamina cribrosa, resulting in acquired optic pit formation OS
*Optic nerve head colobomas

### IV. Diagnosis and discussion:

*The patient was diagnosed with multiple optic pits of the left eye (OS) based on the clinical fundus examination, SD-OCT, and visual field testing. Fundus autofluorescence (FAF) imaging showed an extended area of retinal nerve fiber layer (RNFL) loss in the temporal peripapillary area OS. The patient was also diagnosed with open-angle glaucoma (OAG) OS in the presence of strong family history, elevated intraocular pressures OU, visual field defects OS, and profound loss of RNFL OS. The patient is an open-angle glaucoma suspect in the right eye (OD) based on strong family history, moderate cup-to-disc ratio, and elevated intraocular pressure.

*Optic pits can be congenital or acquired. This case begs the question, which came first, the open-angle glaucoma or the optic disc pits. The patient’s ocular history does not reveal a definitive answer, and it is difficult to clinically diagnose congenital versus acquired optic pits.

→ Congenital optic pits are a defect of the lamina cribrosa with a herniation of nerve tissue into the pits. The developmental process for the lamina cribrosa suggests that congenital optic disc pits form because of poor migration of mesodermal cells from adjacent sclera and poor differentiation of the mesodermal cells into the lamina cribrosa.

→ Optic pits can also be acquired from focal lamina cribrosa defects. When there is elevated intraocular pressure, there is a posterior bowing of the lamina cribrosa, posterior migration of the laminar insertion and thinning of the lamina cribrosa. The glaucomatous damage will occur in a localized area, resulting in acquired optic pits. When there are focal lamina cribrosa defects, the laminar beams in the adjacent areas have greater mechanical stress and strain than before, becoming more susceptible to further RNFL loss.

Regardless of congenital or acquired optic pits, they represent a localized area of susceptibility of the lamina cribrosa to the damaging effects of elevated IOP. The multiple optic pits and glaucoma puts the patient at greater risk for quicker global progression of RNFL loss, with subsequent visual field defects.

*Previously, the lamina cribrosa was only evident ex-vivo. However, with new advances in technology such as EDI-OCT, it is now possible to examine thinning, posterior displacement, and focal defects of the lamina cribrosa in-vivo. The lamina cribrosa is important to consider in glaucoma. Abnormalities of the lamina cribrosa are thought to contribute to or in some cases initiate the blockage of orthograde and retrograde axonal transport within the retinal ganglion
cell axons that lead to glaucomatous vision loss. Studies have revealed that the lamina cribrosa is thinner in glaucomatous eyes versus nonglaucomatous eyes. Posterior deformation of the anterior lamina cribrosa is considered one of the key manifestations of glaucomatous optic neuropathy. Using EDI-OCT technology, it is possible to compare the posterior displacement of the anterior lamina cribrosa with an elevated intraocular pressure, as well as the anterior movement of the lamina cribrosa once the intraocular pressure is under control. Experimental studies have shown that lamina cribrosa displacement occurs at an early stage of glaucoma, and that this may precede the retinal nerve fiber layer change that is detected on the SD-OCT.

*Additionally, it has been demonstrated that associated visual loss is more likely to be severe and in paracentral locations, with varying size and density of the scotoma, in eyes with both optic disc pits and open-angle glaucoma. In one particular study, peripheral optic disc pits in an older Caucasian population were associated with fixation threatening open-angle glaucoma; this is important to keep in mind when addressing the case in discussion since this patient has a dense scotoma OS evident on HVF 10-2.

V. Treatment & Management:

*The patient is currently being treated with ocular hypotensive medications in the left eye only. Patient presented to the VA in April 2016 on 1 drop of Travatan Z at bedtime and 1 drop of Brimonidine 0.1% twice a day. No changes have been made to the current treatment regimen thus far. However, patient is scheduled for a follow-up exam in the next few weeks, and with consistently elevated intraocular pressures in both eyes, the treatment regimen will be adjusted and drops will be initiated for the right eye.

*The patient will continue to be monitored with visual field testing, 24-2 and 10-2, SD-OCT imaging will be acquired to track the progression of the retinal nerve fiber layer, and EDI-OCT will be acquired prior to and after initiating drops for the right eye, as well as acquiring images of the left to track progression or changes to the lamina cribrosa.

*Complications of optic pits include retinoschisis and maculopathy, so patient dispensed home amslcr to assess changes to vision in either eye.

VI. Conclusion & Clinical Pearls:

*Eye care providers have a set of standard testing that is necessary when managing and treating glaucoma and glaucoma suspects; these consist of visual field testing, pachymetry, gonioscopy, optical coherence tomography and intraocular pressure measurements. Newer modes of technology such as EDI-OCT make it possible to take glaucoma management and treatment to a more detailed level. In this way, it may be beneficial to start measuring the lamina cribrosa depth in those patients who are suspected of glaucoma, as well as those patients who have glaucoma. It may be possible to monitor for a change in the location of the lamina cribrosa, either anterior or posteriorly displaced, prior to and after treating the patient with ocular hypotensive medications.

*Lowering intraocular pressure remains the best way to treat glaucoma. With EDI-OCT, there is further evidence to confirm that in-vivo, the lamina cribrosa is affected by the elevation in intraocular pressure. Pressure-induced displacement of the lamina cribrosa impairs the axoplasmic flow that supports the ganglion cell axons; this in turn leads to retinal ganglion cell
death and glaucomatous defects on visual field testing. There have been studies to show that the lamina cribrosa does rebound once the intraocular pressure is under control. EDI-OCT has potential to become an early diagnostic tool for glaucoma instead of waiting until there is progression evident on the RNFL scan or on the visual field.

*In conclusion, EDI-OCT is advantageous for evaluation of the lamina cribrosa in a patient that has glaucoma complicated by multiple optic pits. Optic nerve head evaluation is difficult in the presence of pit excavation, so the posterior displacement and thinning of the lamina cribrosa made visible with EDI-OCT is useful.

References:


