Importance of Objective Documentations in Primary Open Angle Glaucoma with Hereditary Pigmentary Retinopathy Management

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Abstract:

Hereditary pigmentary retinopathy causes 360 VF constrictions; therefore it is important to use objective testing if primary open angle glaucoma is a concurrent condition. This case discusses the work-up and management of such a case.

I. Case History
   a. 81 year old African American male
   b. Chief Complaints
      i. No vision complaint today
      ii. Patient presented for possible selective laser trabeculoplasty (SLT) due to questionable compliance
   c. Ocular History
      1. Primary Open Angle Glaucoma (POAG) both eyes (OU) based on ONH appearance
      2. Target: 13-15
      4. Pach: OD 518/ OS 518
      5. No family history of glaucoma (Glc)
      i. H/O Hereditary Pigmentary Retinopathy (HPR) OU
         1. Denies nyctalopia
      ii. H/O blunt trauma to left eye (OS) 1990’s, no trauma right eye (OD)
   d. Medical History
      i. Alcohol abuse
      ii. Cocaine dependent
      iii. Thoracic Aortic Aneurysm
      iv. Hypertension
      v. BPH w/o Urinary Obstruct
e. Medications
   i. Amlodipine Besylate 10mg daily
   ii. Aspirin 81mg ec daily
   iii. Latanoprost 0.005% QHS OU
   iv. Tamsulosin HCl 0.4mg daily
   v. Timolol maleate soln 0.5% QAM OU
   vi. Vardenafil HCl 20mg PRN
f. Others
   i. Questionable ophthalmic medicine compliance due to refill schedule on file.

II. Pertinent findings
   a. Clinical
      i. BCVA 20/25 OD, OS, OU (reduced due to cataracts OU)
   ii. Anterior Segment
      1. Angles: wide open, no angle recession
      2. No pseudoexfoliation syndrome (PXF), no pigmentary dispersion syndrome
      3. Otherwise unremarkable
   iii. IOP
      1. OD 13/ OS 14
      2. With current Latanoprost QHS OU and Timolol maleate soln 0.5% QAM OU
   iv. DFE
      1. OU: trace anterior cortical cataracts, 1+ nuclear sclerosis
      2. Cup to Disc Ratio (C/D)
         a. OD 0.75/0.75 with superior thinning
         b. OS 0.85/0.85 very thin superior & inferior rims
      3. Macula: trace epiretinal membrane OU
      4. A/V: normal calibers OU
      5. Posterior Pole:
         a. Retinal pigment epithelium (RPE) atrophy and pigment clumping extending nasal and inferior from disc following arcade
         b. Semicircular circumferential area of RPE mottling temporal to macula
      6. Periphery
         OD: large chorioretinal scar with areas of hypopigmentation within, ~3 disc diameter (DD) in size, at 9:00
         OS: chorioretinal atrophy 360
   b. Imaging
         1. VF progression OS>OD
         2. Impression: constriction 360 OU, sparing the central 20 degrees, denser superiorly.
ii. Fundus photos: 2015, '13
   1. Impression: optic nerve head (ONH) OD superior thinning contributes to the inferior VF defects and inferior thinning on OCT RNFL.

III. Differential diagnosis
a. Primary diagnosis:
   i. POAG OU based on C/D ratio and VF defects with HPR
b. Other:
   i. Physiological cupping with VF defects from HPR OU
   ii. Physiological cupping with Functional VF defects OU
   iii. Physiological cupping with Retinitis Pigmentosa (RP) OU

IV. Diagnosis and discussion
a. POAG is a chronic, progressive disease that most often presents with nerve damage, RNFL defects and VF loss.
b. POAG is a difficult diagnosis when presented with conditions affecting VF. VF is an important clinical test that can evaluate the progression of glaucoma along with clinical judgment of ONH. In the case of this 81 Y.O. patient with HPR causing a constricted field 360 OU, VF has no benefit in POAG management.
c. The HPR is symmetric between OD and OS, denser along the inferior retina corresponding to a denser VF defect superiorly. HPR should not progress, if there are progression, photos should correlate with progression.
d. ONH appearance remains stable at 0.75 OD with superior thinning and 0.85 OS with thinning superior and inferior poles. ONH OD and OS thinning correlates with OCT and VF.
e. ONH OS thinning superiorly and inferior rims correspond to OCT RNFL and VF defects. The inferior thinning OS correlates with the denser inferior VF OS defects versus the inferior VF OD defects.
f. When VF is not as beneficial. POAG management is based entirely on clinical evaluation of the nerve through fundus photos, ONH photos, OCT – RNFL yearly and decrease inter-observation by scheduling patient with the same practitioner yearly.
g. Differential diagnosis
   i. Physiological cupping with VF defects from HPR OU can be ruled out due to thinning on ONH with clinical evaluation and OCT RNFL.
   ii. Physiological cupping with Functional VF defects OU can be ruled out because the VFs were repeatable.
   iii. Physiological cupping with RP OU can be ruled out since the HPR is noted at age 76 and the patient has no complaint of night vision loss. Vision had also been stable for the past 9 years. However, the patient may have a rare case of RP, but further investigation is needed to confirmed diagnosis. In addition, there are thinning on ONH with clinical evaluation and OCT RNFL.

V. Treatment, management
a. POAG OU
i. Although there was questionable compliance with medications usage over the past few years, there was no progression in ONH appearance, and IOP within target, no additional therapy or SLT was recommended.

ii. Patient educated to continued present management: Latanoprost QHS OU and Timolol soln 0.5% QAM OU.

iii. Continue to monitor with yearly OCT-RNFL, photos, intraocular pressures (IOP) and dilated fundus exam

iv. After consideration of stable ONH, good IOP with current meds, patient’s demographics, pachymetry, the patient has a higher risk for glaucoma progression.

v. The patient was advised by the glaucoma specialist to return to clinic in 6 months for a follow up.

b. HPR OU
   i. Longstanding cause of VF constriction OU
   ii. Continue to monitor with VF and photos, to compare and progression of HPR vs progression of POAG.
   iii. Continue to monitor with VF and photos at this time.

VI. Conclusion
   a. The purpose of glaucoma management is preventing functional vision loss. Important clinical tests to monitor progression include VF, OCT RNFL, IOP, clinical observation of ONH. In patients where one or multiple tests are not beneficial, clinician must rely purely on clinical observation, experience and other auxiliary tests to manage this disease.

b. In this specific case, there was minimal benefit with VF loss to monitor POAG progression due to the extensive VF constriction from HPR.

c. Clinical pearls
   i. Although AOA guideline suggests follow up at least every 6 months, I would follow this patient closer due to the multiple risk factors for progression.

ii. In this specific patient case ONH photos needs to be taken at every visit to monitor POAG, in addition to yearly RNFL OCT.

iii. It would be beneficial to schedule patient with the same practitioner to limit inter-observer variability.
References

