18 year old African American male presenting with advanced Juvenile Open Angle Glaucoma

Abstract

An 18 year old African American male presented secondary to referral for elevated intraocular pressure and marked vision loss in the left eye. He was diagnosed with JOAG and ultimately treated with bilateral trabeculectomies.

1. Case history
   a. Patient demographics: 18 year old African American male
   b. Chief complaint: blurry vision in the left eye for past 6 months
   c. Ocular/medical history: acne, asthma
   d. Family history: glaucoma (grandparent)
   e. Medications: tazorac topical cream, multivitamin tablet, history of steroid inhaler use

2. Pertinent findings
   a. Clinical:
      i. VA sc OD: 20/30-1, OS: hand motion
      ii. Pupils: PERRL OU, 3+ APD OS
      iii. CVF OD: inferior altitudinal defect, OS: unable
      iv. IOP by applanation OD: 40 mmHg, OS: 41 mmHg
      v. Pachymetry OD: 592 microns, OS: 592 microns
      vi. C/D ratio OD: 0.85 (-) heme, OS: 0.95 (-) heme
      vii. Gonioscopy OD: grade IV, 1+ pigment, OS: grade IV, 1+ pigment
      viii. OCT RNFL thickness OD: 54, superior and inferior thinning; OS: 66, superior and inferior thinning
   b. Physical: none
   c. Radiology: MRI of brain/orbits showed no mass, 2 T2 hyperintense foci adjacent to frontal horn of right lateral ventricle; repeat in 6 mos

3. Differential diagnoses
   a. Primary: Juvenile open angle glaucoma, advanced OU
   b. Others:
      i. Optic atrophy
      ii. Chronic retinal disease

4. Diagnosis and discussion
   a. Uncommon subset of POAG, (<3 per 100,000)
   b. Initial diagnosis between 4-40 with characteristics including elevated IOP ≥21 mmHg by Goldmann applanation tonometry at the initial hospital visit, open angle configuration on gonioscopy, glaucomatous optic neuropathy (neural rim thinning, focal notching or a vertical cup-to-disc ratio >0.6) and/or glaucomatous VF defects
   c. Male predilection
   d. Myopia associated
   e. Autosomal dominant inheritance pattern
i. Controversial and numerous gene loci found to be associated
ii. Myocillin mutations more associated with advanced cases especially with advanced field loss
f. Older age of diagnosis was associated with worse visual acuity; children with JOAG had a higher percentage of poor visual acuity vs good/fair
g. Many patients present with significant asymmetry or unilateral disease
h. JOAG patients have been shown to respond better to surgical intervention however, younger patients with robust immune systems pose additional challenges with trabeculectomy; use of mitomycin-c with surgery is recommended
i. Unique features of present case
   i. Extremely advanced OAG in a young patient with significant field and vision loss at presentation

5. Treatment, management
   a. Initial: travatan Z qhs OU, simbrinza TID OU
      1. Lowered IOP to 25 OU but due to advanced vision and field loss further intervention was needed quickly
   b. 3 weeks later: bilateral trabeculectomy with mitomycin-c
      1. Successful, IOP lowered to OD 11, OS 13 3 weeks post-op
   c. Referral to Low Vision specialist
      1. Patient received glasses and was asked to return for 3 month follow up but has yet to return

6. Conclusion
   a. Glaucoma management in younger patients is more challenging, and less researched, than adult onset
   b. Proper patient and parent education is very important when treating this condition
   c. Children should have comprehensive eye exams every 1-2 years, especially with family history of glaucoma
   d. Low vision specialists can and should be involved in the treatment of a young patient with significantly reduced visual field and visual acuity.
   e. Glaucoma awareness overall is low

Bibliography


