Abstract

The evolution of treatment and management of congenital glaucoma and the implications in long term management of adults with a history of congenital glaucoma will be reviewed.

I. Case History

- Patient demographics
  BM, 60 year old Caucasian male with history of congenital glaucoma
- Chief complaint
  Blur at distance and near with current glasses
- Ocular, medical history
  1. Congenital Glaucoma diagnosed around 1 year of age.
     Goniotomy surgery OU shortly after in early 1954, by Dr. Barkan, and Dr. Ferguson, at St. Mary's in San Francisco. Repeat goniotomy OS shortly after.
  2. Eye surgeries in adulthood
     1989 Penetrating Keratoplasty OS
     2011 OD CE combined with Descemet's Stripping Endothelial Keratoplasty
     2012 OS CE
  3. Strabismic Amblyopia OS (LHT, LXT)
     patching OD at 3-4 years old

II. Pertinent findings

- Clinical
  Entering Rx & VA:
  OD: +2.50 -3.25 x180  20/30-2
  OS: +2.00 -3.50 x137   20/200+1
  Refraction & VA:
  OD: +2.50 -2.50 x180  20/30+1
  OS: +2.00 -3.50 x137   20/80
  Pertinent Anterior Segment findings:
  Clear graft, no edema, no neovascularization, no keratic precipitates, no signs of rejection OU
  OS: Visible Haab’s striae on host cornea
  IOP: 15/17 mmHg
  Posterior segment findings:
  OD: c/d 0.30 round, macula flat, evenly pigmented
  OS: c/d 0.50 round, macula flat, evenly pigmented
  (ONH appearance stable since 1982, fundus photographs)
  Peripheral retina flat and attached 360 OU
  All else unremarkable
  OCT- GCC:
  OD: borderline thinning superiorly and inferiorly
  OS: thinning inferiorly and temporally, borderline thinning superiorly, nasally
III. Differential diagnosis

- Primary/leading
  Primary congenital glaucoma
- Others
  Cloudy cornea at birth:
    - Birth trauma
    - Intrauterine rubella
    - Metabolic disorders
    - Congenital hereditary endothelial dystrophy
  Large cornea:
    - Megalocornea
    - Very high myopia
  Lacrimation:
    - Delayed canalization of nasolacrimal duct
  Secondary infantile glaucoma:
    - Tumors (i.e. retinoblastoma, juvenile xanthogranuloma)
    - Persistent hyperplastic primary vitreous
    - Retinopathy of prematurity
    - Intraocular inflammation
    - Trauma
    - Ectopia lentis

For corneal findings seen on our patient’s past anterior segment photography prior to corneal transplants:
  Haab’s striae before BM’s corneal surgeries:
    - Posterior Polymorphous Dystrophy
    - Descemet’s folds due to numerous conditions, including:
      Endophthalmitis
      Uveitis

IV. Diagnosis and discussion

- Elaborate on the condition
  Primary congenital glaucoma develops from intrauterine period to 3 months (congenital), 3 months to 3 years (infantile), 3 to 35 years (juvenile) due to abnormal development in the anterior chamber angle with faulty cleavage and abnormal insertion of ciliary muscle, where the iris and ciliary body do not recede posteriorly, and thus instead insert anteriorly into the trabecular meshwork (trabeculodysgenesis)
    - Obstruction to aqueous outflow causes elevation in intraocular pressure
    - Epidemiology: 1 in 10,000 to 1 in 30,000 births
    - Majority of cases are sporadic
    - 40% occur at birth, 85% by 1 year of age
    - Bilateral in 70%, multifactorial inheritance
    - Common signs include elevated IOP, increased corneal diameter (>12mm), blepharospasm, photophobia, tearing, corneal edema, optic nerve cupping, Haab’s striae, flat iris insertion, peripheral iris hypoplasia
Prognosis:
- Occasional spontaneous remission may occur, but most infants become blind unless successful early surgical intervention is performed
- Once intraocular pressure is controlled the cupping in infants may regress

- Expound on unique features
  Glaucoma in infants presents with the following unique features:
  - Stretching of the globe
  - Media opacities, anisometropia, and high astigmatism often lead to amblyopia (seen in BM)
  - Corneal opacity caused by epithelial corneal edema initially, followed by permanent stromal edema, then breaks in Descemet’s membrane (the Haab’s striae seen in BM in earlier photos)

V. Treatment, management

- Treatment and response to treatment
  Primary congenital glaucoma treatment:
  If cornea is clear:
  - Goniotomy – an ab interno surgery
    Requires a clear cornea for good visualization
    Horizontal incision at midpoint of superficial layers of TM
    Introduced by Barkan in 1938
    Requires good visibility, excellent surgical technique
    Success rate is as high as 85%
    May need to be repeated
    Poor results if corneal diameter is 14mm or greater since Schlemm’s canal is destroyed in these eyes
  - Trabeculotomy ab externo
    Preferred if repeated goniotomy failed or if cornea clouded
    Trabeculotome inserted into Schlemm’s canal through a partial thickness scleral flap, and is rotated into the anterior chamber
  If Cornea too edematous and opacified:
  - Trabeculectomy ab externo
  - Cobined trabeculotomy and trabeculectomy

Cyclophotocoagulation in intractable cases
More recently developed procedures:
- Glaucoma Drainage Implants
- Sclerotomy
- Trabectome
- Endolaser
- Transcleral diode laser cyclophotocoagulation

Medical therapy usually unsuccessful
BM underwent 3 total goniotomy surgeries as an infant.
BM currently not on any treatment, monitoring for any change in corneal stability, and possible development of secondary glaucoma
VI. Conclusion

Clinical pearls, take away points:

1. Children with primary congenital glaucoma grow up to be adults who need periodic re-examination for life, with emphasis on monitoring for development of complications such as endothelial dystrophies, elevation in IOP, among others.

2. In irregular corneas even a relatively insignificant change in refractive error can make a significant difference to patient.

3. Importance of fundus photography – able to compare to 1982 optic nerve head photos to establish stability.