Title: Spontaneously Resolved Congenital Glaucoma—A Rare, but Likely Diagnosis for Atypical Corneal Findings in an Adult Patient

Abstract: An adult with normal intraocular pressures, no history of glaucoma treatment, and no evidence of anterior segment dysgenesis presents with bilateral corneal findings suggestive of congenital glaucoma. This case discusses suspected spontaneously resolved congenital glaucoma.

I. Case History:
   a. 49 year old African American male
   b. Chief complaint: Blurred vision OU, worsening in the last 3-5 years
   c. Denies history of eye surgery, confirmed with patient’s father
   d. History of trauma to the eye as a child, hit with a rock in one eye
      i. Ocular Meds: history of unknown drop or ointment as a child
      ii. An unknown drop or ointment was recommended a few years prior. No longer using
      iii. No known history of birth trauma/forceps delivery
   e. No known family history of eye disease/glaucoma
   f. Medical History: AIDS, Hypercholesterolemia
   g. Medications: Truvada, Reyataz, Norvir, Pravastatin, Aspirin

II. Pertinent Findings:
   a. Uncorrected VA
      i. OD: 20/80 -1, PH 20/70+1
      ii. OS: 20/200, PH 20/150
   b. Refraction
      i. OD: -1.00 DS, VA: 20/70+2
      ii. OS: -1.50 -1.00 x180, VA: 20/100
   c. Pupils: Normal, no RAPD
   d. EOMs: Full and smooth OU
   e. Cover test: >45 prism diopter CAXT at distance and near
   f. Corneal findings:
I. OD: 1+ stromal edema, multiple horizontal, vertical, and circumferential band-like lesions at the level of the endothelium. Decreased TBUT

II. OS: 2+ stromal edema, multiple horizontal, vertical, and circumferential band-like lesions at the level of the endothelium. Stromal scar inferotemporal to pupil. Decreased TBUT

g. Corneal diameter, measured with caliper (HxV)
   i. OD: 14.5mm x 13.75mm
   ii. OS: 14.25mm x 13.5 mm

h. Anterior chamber: Deep & quiet OD, OS

i. Intraocular Pressure (IOP)
   i. OD: 14mmHg
   ii. OS: 15 mmHg

j. Iris: Normal OD, OS. (-)transillumination defects, (-)atrophy

k. Lens: WNL OD, OS

l. Posterior segment findings
   i. Optic nerves: Moderate C/D OU. OD: .65/.7, OS: .5/.55 (H/V)
   ii. Macula: flat, intact OU

m. Humphrey Visual Field 24-2, Sita Fast
   i. OD: Reliable. No glaucomatous pattern
   ii. OS: Reliable. General depression on total deviation. Possible start of inferior nasal defect

n. OCT RNFL: Thinning superiorly OU

o. Pachymetry: OD: 661μm, OS: 667μm

p. Axial length: OD: 26.5mm, OS: 25.42mm

q. Corneal curvature: OD: 38.89/41.36 D, OS: 43.60/44.53 D

r. Anterior chamber depth: OD: 3.91mm, OS: 3.69mm

s. Gonioscopy: Difficult views due to edema. Appears to be open to at least scleral spur in all quadrants with no evidence of angle dysgenesis OU

III. Differential Diagnosis

a. Haab’s striae
   i. As a result of buphthalmos
   ii. As a result of birth trauma (forceps delivery)

b. Posterior Polymorphous Dystrophy (PPMD)

c. Congenital Hereditary Endothelial Dystrophy (CHED)

d. Simple Megalocornea

IV. Diagnosis and Discussion

a. Haab’s striae from buphthalmos, presumably caused by spontaneously resolved congenital glaucoma
   i. Presence of megalocornea, longer than average axial length, and deep anterior chambers suggest buphthalmos from increase IOP at a young age. Corneal enlargement generally occurs before age 3 (1)
ii. In infants and young children, elevated IOP may cause enlargement of the globe (buphthalmos), limbal tissue (deep anterior chamber), and cornea (leading to circumferential and horizontal ruptures of Descemet’s membrane, or Haab’s striae) (2).

iii. In most cases, surgery is necessary in order to control intraocular pressure to prevent damage to the optic nerve (3). However, this patient is certain that he never had eye surgery.

iv. Primary congenital glaucoma (PCG) is an isolated abnormality of the anterior chamber drainage angle (goniodysgenesis) (4). Although gonioscopy views of this patient are compromised due to corneal edema, there are no signs of angle dysgenesis.

v. Patient is currently a glaucoma suspect due to moderate C/D ratios OU and RNFL thinning on OCT. Uncertain whether this progression is a remnant of childhood disease, or rather an active glaucomatous process.

vi. The patient has no known history of birth trauma or forceps delivery. Haab’s striae from birth trauma is most often unilateral (4). Another cause of descemet’s tears is acute corneal hydrops in keratoconus, which the patient does not have. Other reported cases of spontaneously resolved congenital glaucoma.

vii. Other cases of spontaneously resolved congenital glaucoma have been described. First reported case in 1975 (3).

viii. Nagao et al. describe the ocular findings in a series of children (14 eyes of 9 patients) with spontaneous resolution of PCG. Large corneas were noticed in all affected eyes. All nine patients had deep anterior chambers. Haab’s striae were present in 10/14 eyes. Mean corneal diameter was 13.55mm. All eyes with spontaneous resolution showed the presence of mild to moderate filtration angle abnormalities (3).

ix. Sanghi et al presents a case in which the patient had unilateral megalocornea with haab’s striae, but normal angle structures and normal IOP (5). This more accurately describes our patient, however it is possible that with better gonioscopy views, subtle abnormalities would be apparent.

x. Hsu et al (6) presents two cases in which one had anterior segment dysgenesis and the other did not. Both had Haab’s striae, megalocornea, and hyperopia. One would think that buphthalmos would result in high myopia, but that is not always the case—as is seen in our patient.

xi. A proposed mechanism for spontaneous resolution is the continual postnatal development of the iridocorneal angle. It is also possible the the growth of the eye resulted in reversal of an angle abnormality (4,6).

b. Haab’s Striae vs. Posterior Polymorphous Dystrophy (PPMD) vs. CHED

i. In a comparison between Haab’s Striae and PPMD, Cibis and Tripathi highlight 10 different cases. Out of the five cases with Haab’s striae, 3 were caused by congenital glaucoma, and all three had corneal diameters.
larger than 12.5 mm. None of the five cases with PPMD had enlarged corneal diameters (7).

ii. PPMD bands are described as having irregular and scalloped borders, while the bands in Haab’s striae have smoother outlines (7). The appearance of our patient’s bands are not scalloped.

iii. PPMD typically presents with three main appearances: grouped or aligned vesicles, broad linear bands with irregular scalloped edges that present as parallel pairs, or geographic placoid lesions (8). Our patient does not have any vesicle, vacuole, or geographic type lesions.

iv. PPMD is typically nonprogressive. Visual impairment is uncommon and usually not significant, but there is potential for severe visual loss from edema and glaucoma from iridocorneal adhesions (8).

v. In Congenital hereditary endothelial dystrophy, the patient will most often have opacified, edematous corneas before two years of age (8). The railroad track appearing lesion that our patient has are not consistent with CHED, and the degree of edema is not significant enough to be CHED.

c. Megalocornea without glaucoma (Simple Megalocornea)
   i. Megalocornea refers to corneas with greater than 13mm horizontal diameter without Descemet’s breaks or scarring. Classically x-linked. Can be associated with Marfan’s syndrome and Neuhauser syndrome (with mental retardation) (4).

V. Treatment/Management
   a. Can consider Muro 128 ointment for temporary relief of blur
      i. Per patient history, this was most likely tried in the past
      ii. Not a good long-term solution
   b. Plan for combined phacoemulsification/DSAEK surgery, OS first
      i. Started patient on Restasis BID OU in order to optimize the ocular surface
      ii. Will do laser peripheral iridotomy (LPI) OS prior to surgery to minimize risk of pupillary block from the air-bubble in the anterior chamber
   c. Regular follow-up to monitor for glaucoma
      i. IOP must be adequately maintained pre/post-surgery
      ii. Regular IOP checks
      iii. Repeat VF and OCT as necessary

VI. Conclusion
   a. Upon first glance at the patient’s cornea, buphthalmos from spontaneously resolved congenital glaucoma may seem like an unlikely diagnosis; however, it is difficult to ignore the accompanying findings of bilateral megalocornea, long axial lengths, and deep anterior chambers
   b. Although only in isolated case studies, spontaneously resolved congenital glaucoma has been described in the literature
c. With this diagnosis in mind, and given the patient’s currently normal IOPs, it is possible that the patient’s early and temporarily high intraocular pressure contributed to the presence of moderate cupping and RNFL thinning. However, the patient will continue to be monitored for a possible active glaucomatous process.

d. Fortunately, in this case, the lack of a definitive diagnosis is unlikely to negatively affect the patient’s visual outcome. The resulting corneal edema will be treated in the same manner, no matter the underlying cause.

e. The patient’s diagnosis may have been more easily determined with a better, more detailed patient and family history, and possibly genetic testing.

VII. References


