Essential Iris Atrophy

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The patient is a 27 year old female with complaints of:

- “Black spots” occasionally OD for the past 3-4 wks
- Sharp pain OD for 1 wk which became worse the day before she presented to our clinic
- An irregular pupil for the past 5 yrs
- Decreased vision OD
Patient History

- POHx
  - (-) Trauma
  - (+) Myopic Astigmatism
  - (+) Strabismus OD as a child

- PMHx
  - (-) DM
  - (-) HTN
  - (-) Thyroid Dz
  - (-) Cancer
  - (-) HIV
Patient History (cont.)

- **PFHx**
  - (-) DM
  - (-) HTN
  - (+) Cancer - Maternal Grandmother
  - (-) Glaucoma
  - (-) Cataracts
  - (+) Strabismus – Father, Brother

- **Present Medications**
  - Tylenol and Codeine
  - Excedrin

- **Allergies**
  - NKDA
Initial Exam

- VA (without correction)
  - OD: 20/25-2
  - OS: 20/20-2

- Motility
  - WNL OU

- CVF
  - Full OU

- Pupils
  - OD: Irregular with 1+ reaction
  - OS: 5 mm with 2+ reaction
  - No RAPD

- IOP
  - OD: 44 mmHg
  - OS: 14 mmHg
Slit Lamp Exam

- Lids & Adnexa
  - OD: Some swelling
  - OS: WNL

- Conjunctiva & Sclera
  - White & Quiet OU

- Cornea
  - Clear OU

- A/C
  - OD: Trace-1+ Cell
  - OS: Deep & Quiet

- Iris
  - OD: Irregular
  - OS: Flat & Round

- Lens
  - Clear OU
Gonioscopy & Fundus Exam

- Gonioscopy showed 360 degrees of peripheral anterior synechias OD
- The dilated fundus exam was essentially normal except the C:D ratio was slightly greater in the right eye
- The dilated fundus exam also revealed that the blood vessels in the right eye were only full during systole
Impression & Plan

- The doctors diagnosed the patient with essential iris atrophy related to ICE syndrome.
- The patient was given two rounds of Alphagan, Lumigan, Diamox, and Cosopt which reduced the IOP OD to 17mmHg.
- The patient was prescribed Alphagan, Diamox, and Timolol and was scheduled for close follow-up due to the likelihood that she would need a shunt implanted.
Photographs

Photographs taken at age 34 after implantation of a Baerveldt shunt

Photographs taken by Lori J. Wood COMT
What is Essential Iris Atrophy?

- Essential iris atrophy is one type of ICE (iridocorneal endothelial) syndrome. Other forms of ICE syndrome include Chandler syndrome and Cogan-Reese syndrome.
- It is typically a unilateral, progressive condition that affects the corneal endothelium.
- The condition does not appear to be related to any type of systemic disease.
- Patients may present with a pupil that slowly becomes “out of place” or distorted (corectopia) and atrophy of the iris resulting in holes. Peripheral anterior synechiae (PAS) often accompanies the disorder.
- Glaucoma develops in about 50% of cases.
Demographics

- Patients that present with essential iris atrophy are typically young to middle-aged, Caucasian females.
- Essential iris atrophy has been documented in children although this is rare.
Main Ocular Structures Involved

- Cornea
  - Specifically the endothelium

- Iris
  - Membrane formation of degenerative endothelial cells which may cause traction on the iris

- Anterior Chamber Angle
  - Can become obstructed by PAS
Pathophysiology

- All ICE syndromes are characterized by the degeneration of the endothelial layer of the cornea. Essential iris atrophy displays the greatest degree of endothelial degeneration among ICE syndromes.

- Endothelial cells transform into “epithelial-like cells” and form a membrane which may adhere-to, and pull-on the iris. This causes holes to form in the iris and synechiae to develop. The synechiae can block the angle and result in secondary angle closure glaucoma.

- Formation of the membrane between the endothelium and the iris can damage the sodium-potassium pump of the endothelium and cause corneal edema.
Signs & Symptoms

- **Signs**
  - Beaten bronze appearance of the endothelium
  - Corneal edema
  - Iris atrophy and formation of holes
  - A pupil that is “out of place” (corectopia)
  - Peripheral anterior synechiae usually resulting in secondary angle closure

- **Symptoms**
  - Blurred vision
  - Possible monocular diplopia due to light entering through multiple holes in the iris (polycoria)
  - Other symptoms, like pain, may result from angle closure glaucoma
Important elements of the ophthalmic exam

- Visual Acuity
  - It is always important to know if vision has been affected
- Pupil Function
  - To see if the pupil is responsive and observe for corectopia
- Slit Lamp Exam
  - The cornea should be evaluated for edema and beaten metal appearance
  - The iris should be evaluated for synechiae and atrophy or holes
- IOP
  - It is important to see if pressure is elevated in the affected eye
- Gonioscopy
  - Allows for evaluation of PAS and better visualization of blocked angle
Treatment

- Treatment typically focuses on controlling the secondary glaucoma caused by essential iris atrophy. This can be accomplished either by using medications or through surgical procedures.

- If corneal edema persists, penetrating keratoplasty (PKP) is often indicated.

- There are also procedures such as deep lamellar endothelial keratoplasty (DLEK) and Descemet's stripping with endothelial keratoplasty (DSEK) that can be used to remove and replace only the defective layer of the cornea. These procedures have a lower rejection rate and recovery time than PKP.
Prognosis

- The prognosis is good for many patients
- The progression of the disorder is unpredictable
- Patients typically do not become completely visually impaired as the condition is most times unilateral
References

