The Tell-Tale Iris

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Abnormal iris findings may be associated with:

- Developmental abnormalities
- Space occupying lesions
- Degenerative disease
- Vascular disease
- Infectious/Inflammatory disease
- Trauma
- Glaucoma-related pathology***
Review of Iris Anatomy

Iris: Stroma and 2 layers of epithelium

- 2 arteriole complexes
- 2 muscle complexes
Review of Iris Anatomy
Review of Iris Physiology Functions

- **Forms and regulates Pupil size**
  - Regulate retinal illumination
  - Reduction of spherical aberration in accommodation
  - Increase depth of focus

- **Vascular tissue provides nutrition for non-vascularized structures**
Clinical Examination of Iris

Slit Lamp/External

Gonioscopy

Ultrasound biomicroscopy

AS-OCT
And NOW......The Cases
Case #1: Iris Mass Lesion

- 24 year old female
- Referred for second opinion regarding peripheral iris lesion
- Mild blur OD x 1 year
- No pain, trauma, surgery, inflammation
- Medical, family, and social histories unremarkable
Case #1

• BVA: 20/25 OD, 20/20 OS
• Pupils: round, equal vigorous reactions; -APD
• SLE/Gonio: as shown
• DFE: slight cortical cataract adjacent to mass lesion
• Mass lesion evident upon dilation, with areas of transillumination evident within mass
Case #1: Ultrasound biomicroscopy
Case #1: Differential Diagnosis

- Cystic mass lesion
  - Iridociliary
  - Stromal
- Solid tumor
  - Melanoma
  - Nevus
  - Metastatic carcinoma
  - Medulloepithelioma
  - Adenoma
  - Leiomyoma
- Embedded Foreign Body
- Inflammatory Nodule
Case #1: Differential Diagnosis

- Iris stromal cyst
- Iridociliary or epithelial cyst
Case #1: Differential Diagnosis

- Solid iris tumors
  - Nevus
  - Iris/CB melanoma
  - Medulloepithelioma
  - Adenoma
  - Leiomyoma
  - Metastatic carcinoma
Case #1: Differential Diagnosis

Amelanotic Iris Nevus

Video courtesy U Iowa, Gonioscopy.org; WLM Alward, MD
Case #1: Differential Diagnosis

- Solid iris tumors
  - Nevus
  - Iris/CB melanoma
  - Medulloepithelioma
  - Adenoma
  - Leiomyoma
  - Hemangioma
  - Metastatic carcinoma
Case #1: Differential Diagnosis

- Combined iris and ciliary body melanoma
Case #1: Differential Diagnosis

Metastatic carcinoma
Case #1: Differential Diagnosis

Hemangioma

Leiomyoma
Case #1: Differential Diagnosis

Medullo-epithelioma
Case #1: Differential Diagnosis

Pigment epithelial adenoma
Case #1: After iridocyclectomy at Wills Eye Hospital, the Diagnosis is:

- Cystic mass lesion
  - Iridociliary
  - Stromal
- Solid tumor
  - Nevus
  - Melanoma
    - Metastatic carcinoma
    - Medulloepithelioma
    - Adenoma
    - Leiomyoma/Hemangioma
- Embedded Foreign Body
- Inflammatory Nodule
Iris nevi / melanoma compromise 49-72% of all solid iris tumors. (Of these, 70-90% are nevi)

Uveal melanoma is most common primary intraocular malignancy.

Iris/CB melanoma incidence 3-15% of all uveal melanomas (the rest is choroidal melanoma).
- Metastasis rate for iris/CB melanoma: 3-5%
Slit lamp diagnosis of melanoma according to J. Shields

- Lesion replaces iris stroma
  - At least 3mm diameter
  - At least 1mm thickness

OR.....3 of the following

- Photodocumentation of growth
- Prominent vascularity
- Ectropion iridis
- Secondary cataract
- Secondary glaucoma
Differential Diagnosis

Iris stromal cyst

Iris melanoma
Differential Diagnosis Caveats

Cyst

Adenoma
Mass Lesion Management

- Observation
  - Slit lamp
  - Gonioscopy
  - UBM
  - Photography

- Local Excision

- Plaque Radiation

- Enucleation
Iris Mass Lesion Pearl

- Don’t forget gonioscopy, post-dilation examination, and the superior conjunctiva
Case 2: presents for routine exam

- 23 year old male
- No complaints/concerns except for mildly blurred distance vision through SCL’s

- Medical Hx: unremarkable
- Family Hx unremarkable
- Social Hx: smoker
- LEE: 2 yrs ago elsewhere
Case 2: presents for routine exam

- VA 20/20 OD, OS
  - -4.00 – 1.25 x 180
  - -4.25 – 0.75 x 180

- Pupils sluggish: -APD
- EOM’s: full/normal
- CF: full OD/OS
- Slit lamp: nml except for .....
Case: Iris transillumination

- **GAT-IOP:** 23, 24
- **Gonio OU**
  - 4+ TM pigment $360^0$
  - *Concave approach*
  - CBB visible $360^0$
- **UBM:**
  - Confirms concave approach/backward bowing of the iris
DFE findings

Lens:

Zentmeyer (Scheie) line

Large cups in average-sized discs with Thin inferior rim OU
24-2 Visual field findings
Iris transillumination with Glaucoma

• DDx:
  – Pseudoexfoliation
  – Pigment dispersion
  – Chronic/Intermittent angle closure
  – Chronic/intermittent herpetic inflammatory glaucoma
  – Trauma (including intra-ocular surgery)
Iris transillumination with Glaucoma

- **DDx:**
  - Pseudoexfoliation
  - Pigment dispersion
  - Chronic/Intermittent angle closure
  - Chronic/intermittent herpetic inflammatory glaucoma
  - Trauma
Iris transillumination with Glaucoma

- DDx:
  - Pseudoexfoliation
  - Pigment dispersion
  - Chronic/Intermittent angle closure
  - Chronic/intermittent herpetic inflammatory glaucoma
  - Trauma
Iris transillumination with Glaucoma

Iris backbowing

Resolving PDS

Video courtesy U Iowa, Gonioscopy.org; WLM Alward, MD
Iris transillumination with Glaucoma

- Villainous PDS masqueraders
Iris transillumination with Glaucoma

• DDx:
  – Pseudoexfoliation
  – Pigment dispersion
  – **Chronic/Intermittent angle closure**
  – Chronic/intermittent herpetic inflammatory glaucoma
Iris transillumination with Glaucoma

• DDx:
  – Pseudoexfoliation
  – Pigment dispersion
  – Chronic/Intermittent angle closure
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Iris transillumination with Glaucoma

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Iris transillumination with Glaucoma

- DDx:
  - Pseudoexfoliation
  - Pigment dispersion
  - Chronic/Intermittent angle closure
  - Chronic/intermittent herpetic inflammatory glaucoma
  - Trauma
Case: And the Diagnosis Is:

- DDx:
  - Pseudoxfoliation
  - **Pigment dispersion**
  - Chronic/Intermittent angle closure
  - Chronic/intermittent herpetic inflammatory glaucoma
Case: Management Concerns

- Management Pigmentary Glaucoma/PDS
  - Pathophysiology
    - Reverse Pupillary Block
  - Characteristic findings
    - 5 clinical findings
  - Meds
    - Pilocarpine
  - LPI
  - ALT

pre-Pilo  post-Pilo
Case: Management Concerns

- Pathophysiology
  - Reverse Pupillary Block

- Characteristic findings
  - 5 clinical findings

- Meds
  - Pilocarpine

- PI

- ALT

Case: Management Concerns

- Pathophysiology
  - Reverse Pupillary Block
- Characteristic findings
  - 5 clinical findings
- Meds
  - Pilocarpine
- PI
- ALT/SLT
69 year old male bumped eye and now it is red

Also notes mild decrease in VA OS x 1 week through new SRx (from exam 2 weeks ago in which he was not dilated)

MHx: DM x 2 years, diet

FHx: unremarkable

SHx: unremarkable
Case: OD red eye

- VA 20/25 OD, 20/30 OS
- VF’s: grossly full OU
- Pupils: sluggish OU
  - No definitive APD
- EOM’s: full/normal
- Slit lamp:
  - OD normal
  - OS: as shown
Case: OD red eye

- GAT-IOP: 23 OD, 18 OS
- Gonioscopy OU:
  - open to CBB 360°
  - 2+ diffuse TM pigment
  - mildly convex approach
  - No signs of ANV OU
- DFE:
  - OD: see photo
  - OS: see photo
DDx: NVI with hyphema

- **Ischemic Vascular Disease:**
  - CVO
  - DM
  - OIS
  - CRAO
  - Various other vascular disorders

- **Iris Tumors**
  - Melanoma/Metastatic carcinoma
  - Retinoblastoma

- **Fuch’s Heterochromic Iridocyclitis**

- **Chronic uveitis**

- **Chronic RD**

- **Iris Tufts**
Case: Rubeosis Irides DDx

- **Vascular Disease:**
  - CVO
  - DM
  - OIS
  - CRAO
  - Various other vascular disorders
- Melanoma/Metastatic carcinoma
- Retinoblastoma
- Fuch’s Heterochromic Iridocyclitis
- Chronic uveitis
- Chronic RD
- Iris Tufts
Case: Rubeosis Irides DDx

- **Vascular Disease:**
  - CVO
  - DM
  - OIS
  - CRAO
  - Various other vascular disorders
- **Melanoma/Metastatic carcinoma**
- **Retinoblastoma**
- Fuch’s Heterochromic Iridocyclitis
- Chronic uveitis
- Chronic RD
- Iris Tufts
Case: Rubeosis Irides DDx

- **Vascular Disease:**
  - CVO
  - DM
  - OIS
  - CRAO
  - Various other vascular disorders
- **Melanoma/Metastatic carcinoma**
- **Retinoblastoma**
- **Fuch’s Heterochromic Iridocyclitis**
- **Chronic uveitis**
- **Chronic RD**
- **Iris Tufts**
Case: Iris Neovascularization DDx

- **Vascular Disease:**
  - CVO
  - DM
  - OIS
  - CRAO
  - Various other vascular disorders
- Melanoma/Metastatic carcinoma
- Retinoblastoma
- Fuch’s Heterochromic Iridocyclitis
- Chronic uveitis
- Chronic RD
- **Iris Tufts/normal variants**
  - Hemangioma
Additional Testing

Review of Labs: unremarkable

Carotid Doppler:
Right: less than 50% ICA stenosis
Left: >70% stenosis

Echocardiography
• Dilated LV and RV
• LVEF 40-50% with moderate hypokinesis
• severe MR, severe TR
And the Diagnosis Is:

- Retinal Vascular Disease:
  - DM
  - Central vein occlusion with iris neovascularization
  - OIS
  - CRAO
  - Other retinal ischemic disorders
- Iris Tumor
- Iris Microvascular Tufts
- Fuch’s Heterochromic Iridocyclitis
- Chronic uveitis
- Chronic RD
Rubeosis Irides Pearls

- Must determine whether **true neovascularization** is present or whether engorged iris vessels/iris tufts are causing rubeosis

- **Gonioscopy critical to rule out angle NVI**

- Sometimes must follow patients over time to determine nature of rubeosis
Management of True Iris Neovascularization

- **Urgent Anti-VEGF tx**
- **PRP:** time frame depends somewhat on underlying entity
- **Carotid endarterectomy**
  - Angle/glaucoma issues
- **Cardiac status considerations**
- **Monitor fellow eye**
NEXT Case

- 12 year old female
- Asymptomatic
- Medical Hx: pediatrician concerned about possible café au lait spots and referred for additional evaluation
- Family Hx unremarkable
Case: Iris nodules

- VA 20/20 OD, 20/40 OS
- VF’s: full OU
- Pupils normal: 1+ APD
- EOM’s: full/normal
- Slit lamp: iris nodules OU
- IOP: 14 OD, OS
Differential Diagnosis

- **DDX of iris nodules:**
  - Inflammatory nodules (syphilis/TB/sarcoid)
  - Lisch nodules
  - Melanoma
  - Metastatic carcinoma
  - Medulloepithelioma
  - Juvenile xanthogranuloma
  - Leiomyoma
  - Lymphoma
  - Iris Nevi
  - Iris Cysts
Case #2: Iris nodules

- **Inflammatory nodules**
  - **Sarcoid**
    - Busacca (surface)
    - Koepppe (pupil)
  - **Syphilis**
    - Gumma
    - Papule
  - **TB**
    - Miliary lesion
    - Tuberculoma

Differential Diagnosis
Differential Diagnosis

- Inflammatory nodules
  - Sarcoid
    - Busacca
    - Koepppe
  - Syphilis
    - Gumma
    - Papule
  - TB
    - Miliary lesion
    - Tuberculoma
Differential Diagnosis

- Inflammatory nodules
  - Sarcoid
    - Busacca
    - Koeppe
  - Syphilis
    - Gumma
    - Papule
  - TB
    - Miliary lesion
    - Tuberculoma
Differential Diagnosis

- Inflammatory nodules
  - Sarcoid
    - Busacca
    - Koeppe
  - Syphilis
    - Gumma
    - Papule
  - TB
    - Miliary lesion
    - Tuberculoma
Case: Iris nodules

- DDX of iris nodules:
  - Inflammatory nodules (syphilis/TB/sarcoid)
  - Lisch nodules
  - Amelanotic melanoma
  - Metastatic carcinoma
  - Medulloepithelioma
  - Juvenile xanthogranuloma
  - Leiomyoma
  - Iris nevi
  - Iris Cysts
Case: Iris nodules

- DDX of iris nodules:
  - Inflammatory nodules (syphilis/TB/sarcoid)
  - Lisch nodules
  - Amelanotic melanoma
  - Metastatic carcinoma
  - Medulloepithelioma
  - Juvenile xanthogranuloma
  - Leiomyoma
  - Iris nevi
  - Iris Cysts
Case: Iris nodules

- DDX of iris nodules:
  - Inflammatory nodules (syphilis/TB/sarcoid)
  - Lisch nodules
  - Amelanotic melanoma
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  - Juvenile xanthogranuloma
  - Leiomyoma
  - Iris Nevi
  - Iris Cysts
Case: And the Diagnosis is:

- Inflammatory nodules (syphilis/TB/sarcoid)
- **Lisch nodules**
- Amelanotic melanoma
- Metastatic carcinoma
- Medulloepithelioma
- Juvenile xanthogranuloma
- Leiomyoma
- Lymphoma
- Iris Cysts/Nevi
Iris nodules: Management

- Lisch nodules present in 92% of patients with neurofibromatosis-1 after 6 years of age
  - May aid diagnosis of NF1

- Workup/monitoring of patients with Neurofibromatosis
  - Monitor for glaucoma: Schlemm’s obstruction/CB tumor/NVI
  - Monitor for early PSC
  - Monitor for tumors that might affect visual or other neurologic function:
    - Optic nerve glioma
    - Intracranial meningioma
    - Malignant tumors, including schwannoma
Men have called me mad; but the question is not yet settled, whether madness is or is not the loftiest intelligence.

--EDGAR ALLAN POE