Differential Diagnosis of Pigmented Lesions of the Fundus

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Disclosure Statement:
Nothing to disclose

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Choroidal Melanoma with Radiation Retinopathy

Dilated Fundus Examination

- Binocular Indirect Ophthalmoscopy
  - Which lens to use?
  - 20D most common
  - 14 to 40 D lenses
- Scleral depression

Dilated Fundus Exam

- Funduscopy
  - Classic lenses: 60D, 78D, 90D
  - Super Series
    - Super 66
    - Super Field
    - Super Pupil
    - Super Vitreofundus
  - Digital High Mag or Wide Field
3 mirror contact lens fundus exam
- AC
- Posterior pole
- 2 peripheral mirrors

Extended Ophthalmoscopy
- Draw optic nerves
- Draw peripheral lesions
- Draw macular lesions

Advanced diagnostic techniques
- Fundus photography
- Fluorescein or ICG angiography
- B-scan ultrasound
- Electrodiagnostics
- Radiology
- Laboratory
- Ocular Coherence Tomography

Enhanced Depth Imaging of Spectral Domain OCT
- Provides higher resolution images of the choroid

EDI-OCT
- THIN Choroid
  - Myopia
  - Aging
  - Choroidal Atrophy
- THICK Choroid
  - Choroiditis
  - CSCR
  - Tumors
  - Hyperopia
  - Younger age

Histology of the Fundus
- Where is the pigment?
  - RPE
  - Choroid
Pigment Location in the Fundus

- **RPE**
  - Congenital-CHRPE
  - Acquired-RPE hyperplasia

- **Choroid**
  - Benign neoplasm
  - Malignant neoplasm
    - Primary
    - Metastatic

CHRPE

- Histology=enlarged RPE cells
- Flat, gray to black retinal lesion with halo
- If multiple lesions OU then question patient about family history of colon cancer***

RPE hyperplasia

- Histology= increase in number of RPE cells
- Dense black lesion in retina
- Occurs due to injury to retina

Benign Choroidal Melanoma-nevus

- Histology= accumulation of melanocytes in choroid
- Flat, slate grey lesion of choroid
- Disappears with red free light

Choroid nevus-risk factors

- Size
  - Up to 2DD typically benign
  - 2-5DD suspicious
  - >5DD highly suspicious

- Elevation
- Lipofuscin
- Feeder vessels
- Only 0.025% convert to malignant choroidal melanoma

Malignant Choroidal Melanoma

- Histology=accumulation of malignant melanocytes in choroid
- Large, mottled, elevated lesion
- White to grey-green color
- Lipofuscin

- Most common primary ocular tumor
Lipofuscin

- Yellow, orange, brown pigment granules
- Form from oxidation of protein and lipid

Autofluorescent Drusen

To Find Small Ocular Melanomas Use Helpful Hints Daily

- Thickness > 2mm
- Fluid-subretinal
- Symptoms: photopsia or metamorphopsia
- Orange pigment
- Margin of tumor touches or within 3mm of ONH

- Ultrasonographic Hollowness
- Absence of Halo
- Absence of Drusen

Malignant Melanoma

- Oncologist consult
  - Liver enzymes
  - Imaging of brain, chest, abdomen
- Retinal oncologist consult
  - Radiation
  - Enucleation
  - Monitor-if small

Metastatic Neoplasm to Choroid

Lung Carcinoma
Breast Carcinoma

OSU SVOSH 2015

Case Study

- 76 year old white male
- Ocular history
  - Cataracts
  - Reticular degeneration
- Complains of flashes, floaters
- Vision 20/20 OD and OS

Case Study

Advanced Macular Degeneration

Peripheral CNVM

ARMD

Treatment Options

- Anti-VEGF injections
- Or Observation
Case Study-Fundus photos

Choroidal Nevi

Color and Red Free

HD Spectral Domain OCT

Color and Fluorescein Angiogram
Further History

• Patient has history of melanoma of arm
• Systemic work up found no other primary tumor or metastatic tumor

• Retinal consult:
  – Choroid Nevus
  – Monitor

Suspicious Choroid Nevus

Case Study

• 45 white male complains of flickering lights OD for 1 month.
• NO floaters

• Ocular history
  – Choroid Nevus OD

Case Study

• Vision
  – OD 20/20
  – OS 20/20
• EOM full OU
• Pupils: ERRL no APD
• Confrontations full OD, OS

Case Study

• Slit lamp normal OU
• Tonometry @ 1:00pm
  – OD 14
  – OS 13

• DFE: 3-4DD choroidal lesion OD superior nasal to disc
Choroid Nevus 12-18-2013

Choroid Neoplasm 7-28-2014

Bscan

OCT

Autofluorescence

Work Up

- CT thorax
- MRI abdomen
- ESR, CRP
- CBC
- Liver enzymes
### Assessment
- Choroidal Melanoma OD

### Plan
- Tantalum ring placement and proton beam irradiation

### To Find Small Ocular Melanomas
- NO risk factors had 3% risk of growth in 5 years—benign
- 1 risk factor—38% risk of growth
- 2+ risk factors → >50% will grow—probably melanoma

### Case Study
- 84 WM
- **Chief complaint**: Loss of vision OD with a “pinwheel image” noted when he rubbed his left eye
- Pertinent medical history
  - Smoker
  - Alcoholism
  - Barret’s esophagus

### B-scan and CT scan
Metastatic Carcinoma

- Basaloid squamous cell carcinoma (BSCC): rare malignant tumor that most commonly affects the upper aerodigestive tract in the region of the larynx, pharynx, and oral cavity
- BSCC of esophagus is aggressive, invasive, and causes rapid death

Case Study

- 57 WM
- Poor peripheral vision and night vision OU
- H/O Retinitis Pigmentosa

Objective

- Vision
  - OD 20/25
  - OS 20/20
- Confrontations
  - Severe restriction OU
- Humphrey VF 120 screener
  - Constricted to 5 degrees OU

Objective

- Lens
  - Gr 1 NS OU; early PSC OD
- Vitreous
  - Syneresis OU
- Maculae
  - CME OU?
- Optic nerves
  - Mild pallor?; 0.2 OU

Fundus photos
OCT

Prior Testing
- Normal color vision
- Normal MRI of brain
- Abnormal ERG

Diagnosis
- Retinitis Pigmentosa sin pigment OU
- Legal blindness
  - Recommend blind rehab

Retinitis Pigmentosa
- Triad
  - Bone spicules
  - Arteriolar attenuation
  - Nerve pallor
- PSC cataract
- Vitreal floaters and syneresis

Case Study
- 60WM
- Amblyopia OD-refractive and strabismic
- Migraines with aura
Objective

- Vision
  - 20/30
  - 20/30
- Maculae
  - OD flat, large CR scar temporal
  - OS flat
- Optic nerves
  - OD 0.25, mild PPA
  - OS 0.3, no PPA

Differential Diagnosis

- Presumed Ocular Histoplasmosis Syndrome OU
- Old chorioretinitis OU
- CHRPE OU
- Pt lives in New England and no exposure to birds
- No significant history
- H/O colon polyps
- 1986 age 39: total colectomy

Assessment

- Chorioretinal scars OU
- CHRPE OU s/p total colectomy for FAP: Familial Adenomatous Polyposis

FAP-Familial Adenomatous Polyposis

- Inherited cancer of the colon and rectum
- Polyps form in the colon in the teenage years
- In classic FAP, colon cancer develops at an average age of 39
- Shortened the APC (adenomatous polyposis coli) gene
Management

- 4 or more CHRPE lesions in both eyes and family history of FAP
- Refer for colonoscopy

Case Study

- 57 WM complains of blur distance and near OU
- Sees wavy lines and black floaters OU
- + osteoarthritis, low back pain, diarrhea
- Previous alcohol abuse

Objective

- Vision
  - OD 20/20
  - OS 20/20
- PERRL NO APD
- Confrontations Full OD, OS
- + metamorphopsia OU on Amsler

Objective

- Slit Lamp exam
  - Unremarkable
- Tonometry
  - OD 13
  - OS 14 @ 1335 hours
- Lens
  - Trace NS OU

Fundus Photos
Lab Work up

- HLA A1, A2, B7, B27, BW6, CW1 and CW7 all positive
- HLA B5 negative

Differential Diagnoses

- Bechet’s Disease
  - History of oral and genital ulcers
  - But HLA B5 negative

Differential Diagnosis

- Inflammatory Bowel Disease
  - HLA B27 positive
  - Chronic diarrhea
  - s/p colectomy

Diagnosis/Plan

- Pigmented Paravenous Retinochoroidal Atrophy
- Monitor with dilation and serial visual fields
- RTC stat if eye pain or vision loss

Pigmented Paravenous Retinochoroidal Atrophy

Rare, reported in 4-70 yr old patients
Typically male
ERG usually normal
EOG usually abnormal
Rarely affects macula
No diagnostic lab findings
Visual Field defects
Case Study

• 45WM
• Referred for ARMD OD (possible wet) and pain OD
• Pt states “icopick” like pain behind OD 5 months ago. +blur and photophobia OD. Works 14 hr days on computer with no vacation.

Objective

• Vision
  – OD 20/20
  – OS 20/20
• PERRL NO APD
• Optic nerves healthy OU
  – OD 0.25
  – OS 0.30

Objective

• Maculae
  – OD 2+ RPE mottling; no blood, fluid, exudate
  – OS clear
• OCT
  – Normal OU
• IVFA
  – OD mottled hyperfluorescence in macula
  – OS normal

RPE mottling OD only

OCT-Stratus
Differential Diagnosis

- Dry ARMD OD only
- Solar retinopathy OD
- Pattern dystrophy—but should be OU if dystrophy
- Others??

Assessment/Plan

- Central Serous Chorioretinopathy (CSCR) OD-stable, inactive.
  - Educate. Reduce stress. Monitor Amsler
- Eye pain OD
  - RTC for VF 24-2, color vision, cover test
  - Consider sinusitis. Order imaging prn

Central Serous Chorioretinopathy

- Idiopathic, sporadic, self-limited
- Risk factors:
  - Young-middle age males
  - Type A personality
  - Steroid use

Fluorescein Angiogram and OCT

- Treatment is observation
- Longterm picture=RPE hyperplasia that can resemble ARMD

Case Study

- 61 WM
- Glasses broken
- +tobacco and alcohol use
- Daughter blind in one eye
- No medications or health problems
Objective

- Vision
  - OD 20/20
  - OS 20/20
- Maculae
  - OD small CR scar superior temporal
  - OS flat and normal
- Fundus
  - Scattered “punched out” CR scars OU

Assessment/Plan

- Presumed Ocular Histoplasmosis OU
  - Photograph and educate
  - Monitor Amsler
  - DFE in 9 months

Ocular Histoplasmosis

- 83WM
- Had laser OD 30 years ago at Dartmouth
- Vision
  - OD 20/100 eccentric fixation
  - OS 20/25

Fundus

Histoplasma capsulatum

- Fungus endemic to central US
- Warm, humid soil
- Bird and bat feces
Choroidal Osteoma
- Rare, benign ossifying tumor
- Female
- Bilateral 25% of cases

Optic Nerve Melanocytoma
- Typically benign and stable
- Monitor VF
- Watch for ONH compression and vascular occlusion
Choroid Rupture
- 57WM
- trauma OD-golf ball

Choroidal Rupture
- Concentric with nerve and ora
- Risk for CNVM
- Epiretinal membrane may form
- Treatment of CNVM
  - Laser
  - Anti-VEGF

Toxoplasmosis

Toxoplasmosis
- Most common cause of intra-ocular inflammation in the world
- Intracellular parasite

Toxoplasmosis
- Congenital
- Acquired
  - Contaminated undercooked meat
  - Ingestion/inhalation of oocysts
  - Blood transfusion/organ transplant
- If posterior uveitis
  - Pyrimethamine
  - Sulfadiazine
  - Clindamycin
  - Prednisone

Active Toxoplasmosis
Lattice Degeneration

Choroid Nevus with Drusen

Macular Scar-Sarcoid

Sarcoidosis
- Chronic, granulomatous disease
- Unknown etiology
- Exaggerated immune reaction by T-helper cells

Sarcoidosis
- Work up
  - ACE
  - Chest Xray
  - Brain MRI prn
- Treatment
  - Systemic steroids
  - Topical steroids

Retinal Coloboma
Malignant Myopia

Retinitis Pigmentosa

Malignant choroidal melanoma

Macular Degeneration

Bull’s Eye Maculopathy

CHRPE
Phenothiazine Retinopathy

Tuberculosis Chorioretinitis

Summary

- RPE
  - Congenital-CHRPE
  - Acquired-RPE hyperplasia

- Choroid
  - Benign neoplasm-nevus
  - Malignant melanoma
  - Metastatic tumor

“It is more important to know what sort of person has a disease than to know what sort of disease a person has.”

Hippocrates