

Meet The Choroid

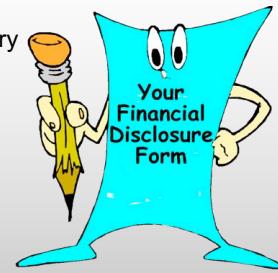
Joe Pizzimenti, OD, FAAO

allthingsoct@gmail.com

Financial Disclosures

□ Honoraria

- Review of Optometry
 - Optometric Management
- Scientific Advisory Boards
- Zeiss
 - Zeavision
 - Thrombogenics
 - Genentech



Financial Disclosures

□ Consulting Fees

- Zeiss
- Zeavision
- Maculogix

□ Proprietary Interests

- None

□ Stockholder: Zeavision



Goals for This Course

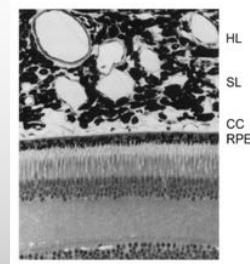
□ Functional anatomy review

- Choroid

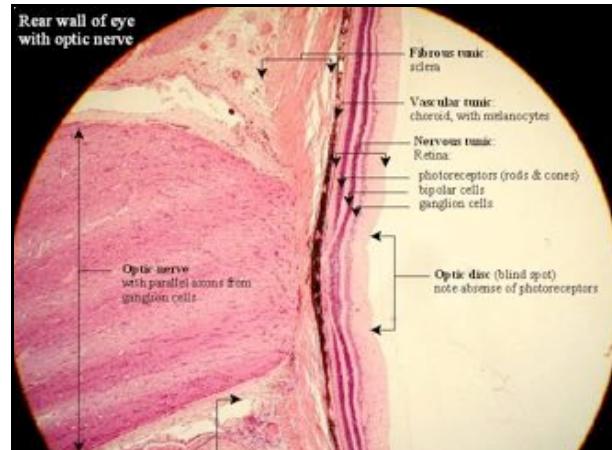
□ Choroid examination and evaluation

□ Case examples

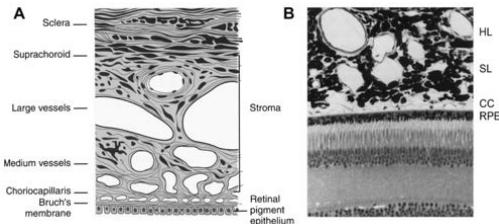
□ Interactive



Questions?

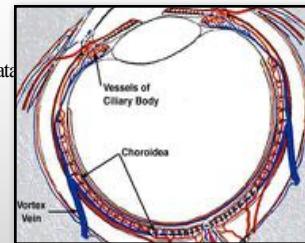


The Choroid: Structure, Function, and Evaluation

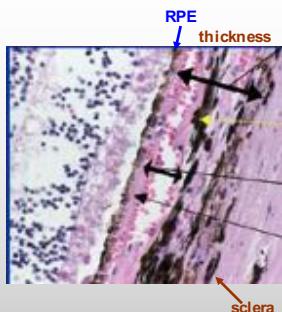


The Choroid

- Located between the sclera and the RPE
 - Extends from ora serrata to optic nerve
- Pigmented/vascular tissue **.75mm** thick
- Nourishes the RPE
 - Choriocapillaris designed to leak
- Absorbs light that passes through retina

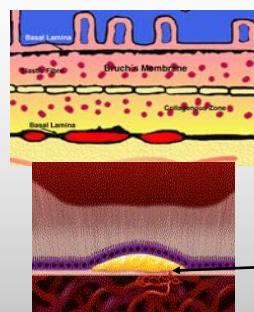


The Choroid



- Loose connective tissue
- Melanocytes
- Choriocapillaris
 - Fenestrated endothelium allows diffusion of proteins
 - S _____ regulation
 - High blood flow
 - Very little O₂ extracted, so high venous O₂

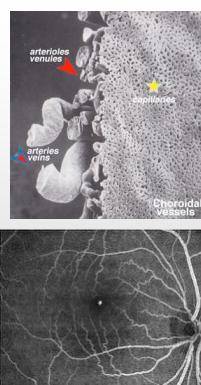
Bruch's Membrane



- Basal lamina of RPE
- Anterior collagenous layer
- Elastic layer
- Posterior collagenous layer
- Basal lamina of CC endothelium
- Contamination of Bruch's can result in d _____, CNVM

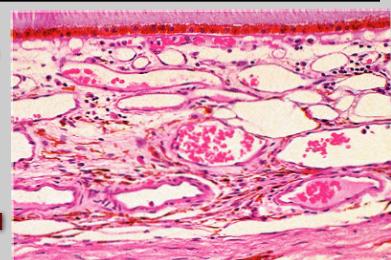
Nourishing the Retina

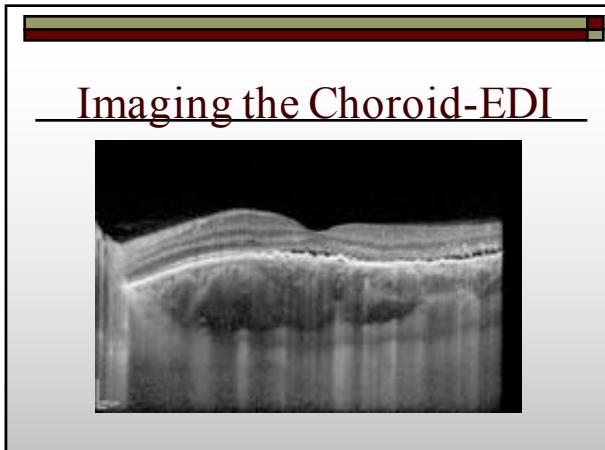
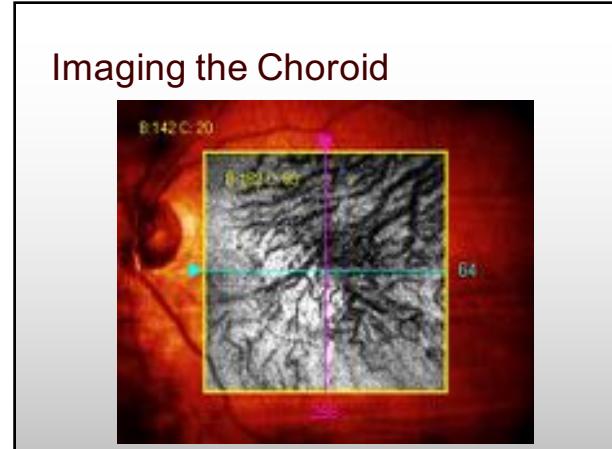
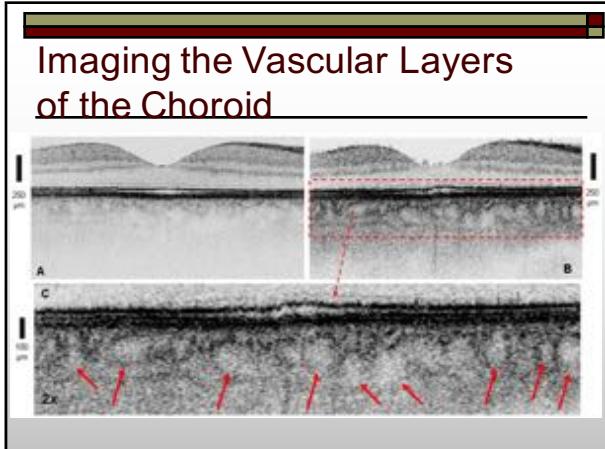
- 2 main sources of blood supply to retina:
- **Choroidal BVs**
 - Supplies outer retinal layers, including PRs
- **CRA**
 - 4 branches nourish inner retina
 - Run radially toward fovea



Choroid Microstructure

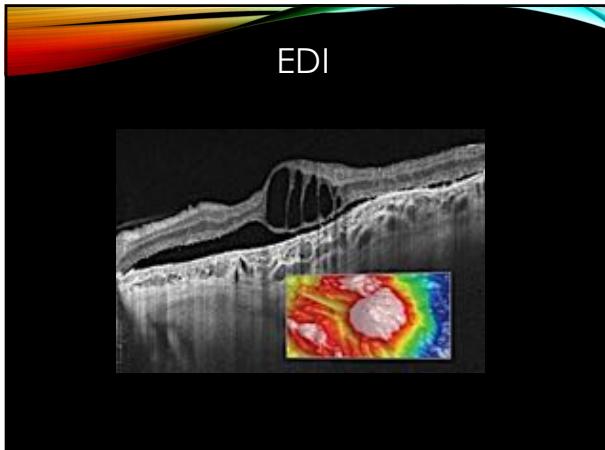
- **Choriocapillaris**
- **Sattler's layer**
- **Haller's layer**
- **Supra - choroid**





WHAT IS ENHANCED DEPTH OCT IMAGING?

- EDI-OCT
- Enhanced-depth imaging (EDI) OCT modifies the standard technique of image acquisition to better reveal the structural details of the **choroid**.



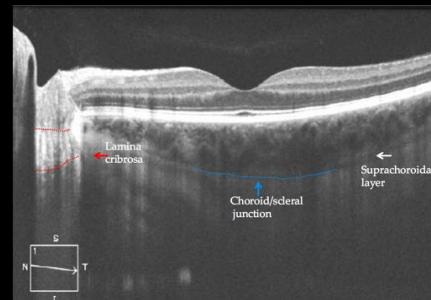
HOW IS EDI ACHIEVED?

- SD-OCT has a **coherence gate** of about 2 mm.
- Coherence gate is the **tissue depth** at which the interference image can be obtained.
- An **interference signal** can be obtained when the tissue being examined enters the coherence gate.
- However, the signal intensity attenuates in the depth direction, from superficial (retinal) to deep layers (choroid).

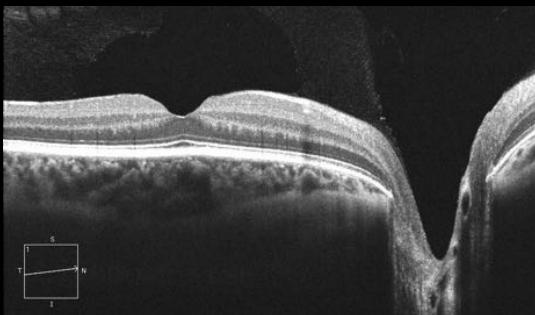
HOW IS EDI ACHIEVED?

- Consequently, to obtain high-quality images in standard SD-OCT, it is important to bring the retinal tissue (B-scan) to the upper aspect of the imaging range.
- In contrast, EDI-OCT creates an **inverted mirror image**. The reference surface of the inverted mirror image is on the **choroidal side**.

EDI SHOWS DEEPER INTRAORBITAL ON, LAMINA, C/S JXN

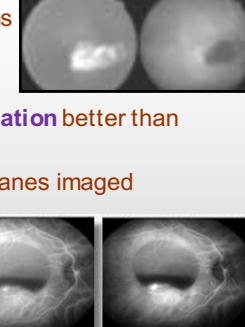


EDI



Indocyanine Green Angiography (ICGA)

- Uses digital imaging systems
- Dye properties
- “Sees” through blood
- Delineates **choroidal circulation** better than fluorescein angiography
- Boundaries of occult membranes imaged



Questions?

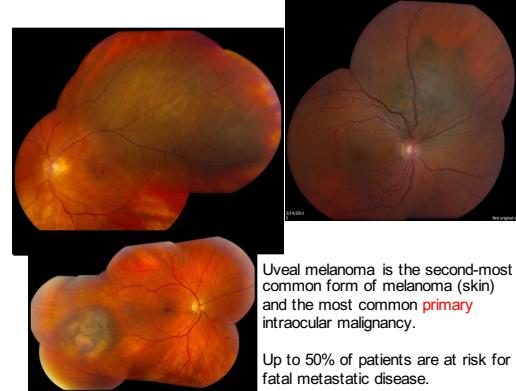


Uveal Melanoma



Many patients with uveal melanoma have no symptoms.

Their tumors are found during a "routine" eye examination.



Uveal melanoma is the second-most common form of melanoma (skin) and the most common primary intraocular malignancy.

Up to 50% of patients are at risk for fatal metastatic disease.



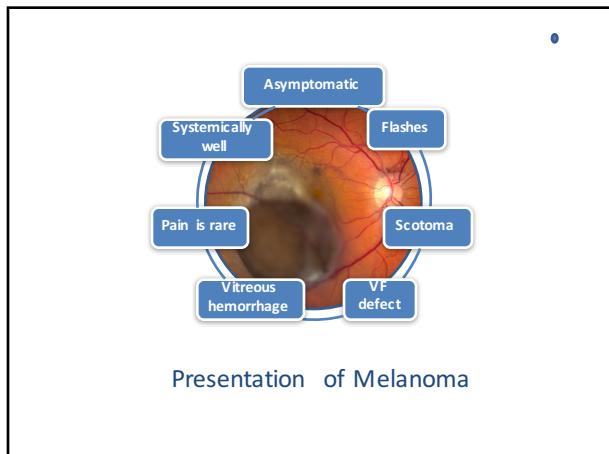
Uveal Melanoma

Choroidal melanoma has an annual incidence of 5-6 cases per million people per year.

Other than having blue or green eyes and a light complexion, studies have not identified any definite risk factors or exposures that predispose patients to developing this cancer.

Amelanotic Choroidal Melanoma





Q: Where does Choroidal melanoma come from?

A: Choroidal Nevus

- Ch Nevus is the most common intraocular tumor
- Proliferation of choroidal melanocytes
- Present in ~ 7.9% of Caucasians
- Growth is rare after puberty?



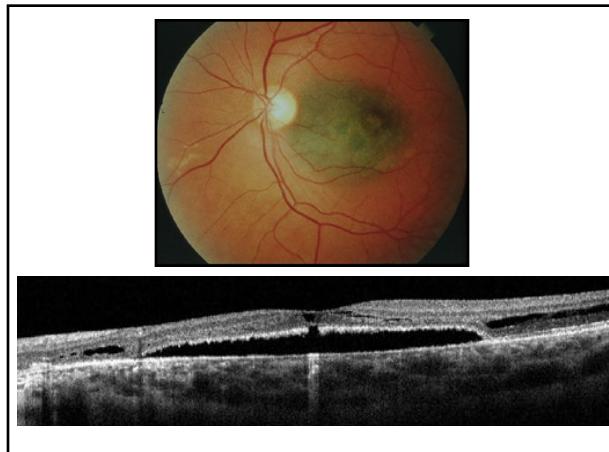
As we age:

- Nevi increase in number and thickness
- Pigment changes
- Metaplasia
- Drusen/lipofuscin

Choroidal Nevus?

Choroidal Nevus?

- Nevi < 2 mm in thickness (A-scan)
- No known relationship to sunlight exposure
- Indistinct borders
- May undergo malignant change into melanoma



Choroidal Nevus Workup and Management

- Baseline fundus photography, FAF
- OCT if location permits, OCTA?
- A/B-Scan
- IVFA?
- Yearly dilated fundus examination
 - Or more frequent

Risk Factors for Melanoma

- Caucasian
- Light colored eyes (blue)
- Fair skin
- Propensity to burn when exposed to UV light
- Cutaneous nevi or freckles
- Iris nevi
- Welders




Follow or Co-manage?

To Find Small Ocular Melanoma Using Helpful Hints Daily

- T= thickness ($>2\text{mm}$)
- F= subretinal fluid
- S= symptoms
- O= orange pigment
- M= margin touches disk

DOCUMENTED GROWTH - MEANS EVERYTHING

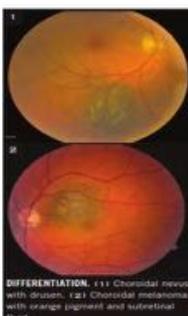
Using Helpful Hints Daily = Ultrasound hollow, halo absent, drusen absent

7/29/2016

Nevus to melanoma

"To Find Small Ocular Melanoma Using Helpful Hints Daily" (TFSOM-UHHD)

T → thickness greater than 2 mm,
 F → fluid subretinally
 S → symptoms
 O → Orange pigment present,
 M → margin with in 3 mm of the optic disc
 UH → USG hollowness (versus solid/flat)
 H → halo
 D → drusen absent



DIFFERENTIATION. (1) Choroidal nevus with drusen. (2) Choroidal melanoma with orange pigment and subretinal fluid.

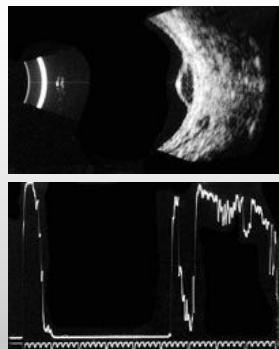
OcularmelanomaCalculator.com



Echography of Small Choroidal Melanoma

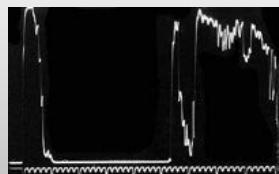
B-Scan Echogram

Assess topographic features, including tumor shape, surface contour and boundaries



A-Scan Echogram

Internal structure, reflectivity, tumor height (elevation)



EDI-OCT

ARCH OPHTHALMOL/VOL 130 (NO. 7), JULY 2012

CLINICAL SCIENCES

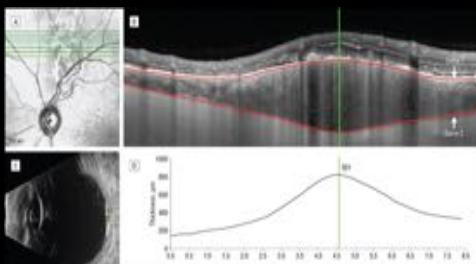
Enhanced Depth Imaging Optical Coherence Tomography of Small Choroidal Melanoma

Comparison With Choroidal Nevus

Carol L. Shields, MD; Swathi Kaliki, MD; Duangkate Rojanaporn, MD; Sandor R. Ferenczy, CRA; Jerry A. Shields, MD

- Mean small melanoma thickness was 1025 microns on EDI-OCT compared to 2300 microns on ultrasonography.

Enhanced Depth OCT (EDI) of a small melanoma

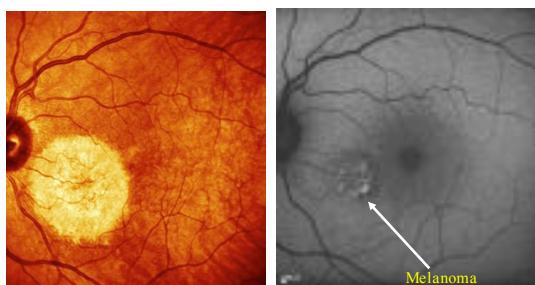


Shields, 2012

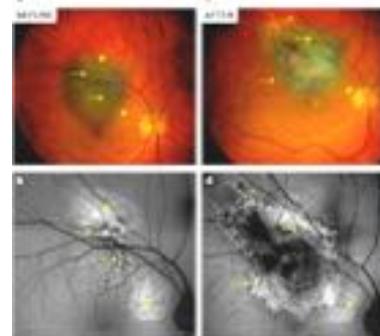
Orange Pigment = Lipofuscin



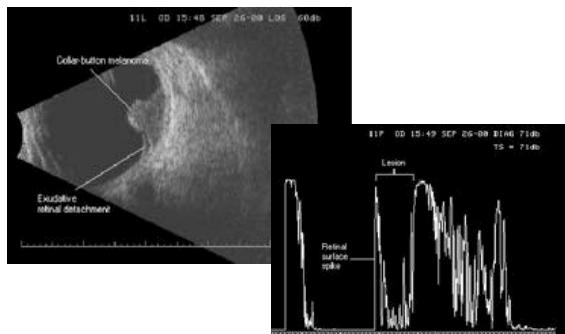
Fundus Autofluorescence (FAF) of a Small Choroidal Mass



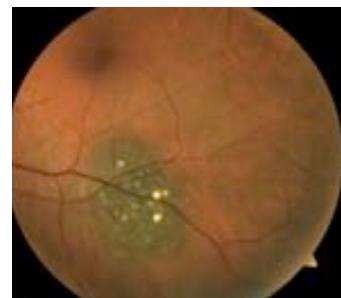
Fundus Autofluorescence (FAF) in Choroidal Melanoma



Echography of Large Melanoma



Nevus w/Drusen = Chronicity



Halo Nevus = Chronicity



No Drusen, No halo



No Drusen, no Halo



Questions?



Differential Diagnoses

- Common, benign lesion
- Focal area in which RPE cells are taller and more densely packed with melanosomes



Congenital Hypertrophy of the Retinal Pigment Epithelium

Familial Adenomatous Polyposis (FAP) & CHRPE

- AD inheritance
- Adenomatous polyps throughout rectum & colon
- Starts to develop in adolescence (15-40 yrs)
- If untreated, all pts will develop colorectal CA



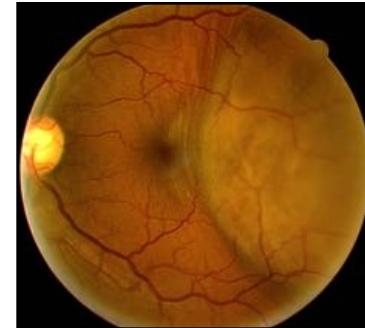
>80% of patients with FAP have atypical CHRPE lesions

Metastatic Tumors to the Choroid

- Breast cancer is the most common CA type to metastasize to the eye - followed by lung CA
- 85% of patients with breast CA metastases will have a known history of breast CA
- Breast CA metastases tend to be bilateral and multifocal (multiple)
- 40% of these patients have a brain metastasis



Metastasis from lung to choroid



Metastatic Prostate Cancer

METS

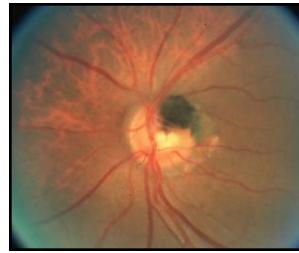
- Benign melanocytic uveal tumor

Melanocytoma

- Composed of large, plump nevus cells that are heavily pigmented
- Can present in all age groups and races, though more common in
 - African-Americans
 - Females

Melanocytoma

- Patients are usually asymptomatic
- Adjacent to or within the optic nerve
- Black in color with feathery margins
- Visual field defect may be present
- APD may be present

**Combined Hamartoma of the Retina and RPE**

- Grey pigmentation with superficial gliosis
 - Secondary retinal wrinkling and vessel tortuosity
 - Lesions can be juxtapapillary, peripapillary or within the posterior pole
- Absence of retinal detachment, hemorrhage, exudation or vitreous inflammation

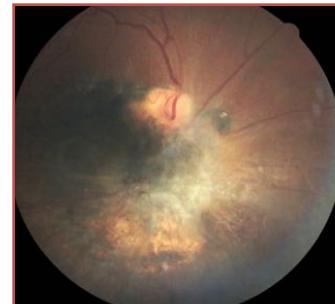
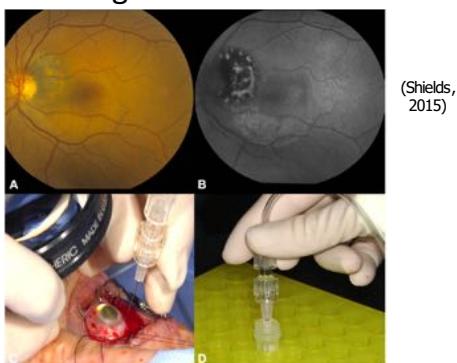
**Genetic Testing on Small Melanoma**

Figure 2. Diagnostic techniques of small melanoma management. A, Early detection of tiny choroidal melanoma using risk factors of subretinal fluid, orange pigment (B) confirmed on fundus auto-fluorescence, symptoms, and tumor touching the optic disc. C, Fine-needle aspiration biopsy and (D) urine sample as preservative for genetic testing.

- Enucleation
 - Radioactive plaques
 - Proton beam radiotherapy
- Most widely accepted

Treatment & Management**Treatment & Management**

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Co-manage Uveal Melanoma with:

Retina/Ocular Oncology
PCP
General Oncology



Collaborative Ocular Melanoma Study

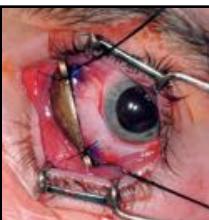
- Organized and funded in 1985 to address issues related to management of choroidal melanoma.
- Main Outcome: overall survival of patient following treatment
- > 4000 patients. 65% pts eligible

<u>Small</u> melanomas	< 2.5 mm in height
<u>Medium</u> melanomas	2.5 – 10.0 mm
<u>Large</u> melanomas	> 10.0 mm

- Secondary outcomes: metastasis-free survival, years of useful vision

Brachytherapy for Uveal Melanoma

Plaque left in place for 4 days to provide 8,000 centigray of radiation to entire tumor. The remainder of the body receives a small amount of radiation, about the equivalent of a chest x-ray.



Treatment Side Effects

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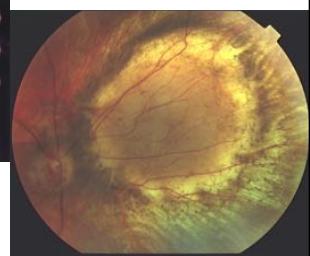
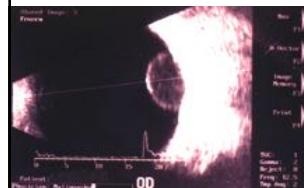
- Main side effect of focal ocular treatment is...
- Radiation retinopathy!
- NVD / NVE
- Exudative changes
- Macular edema
- Occurs several weeks to months after therapy



Choroidal melanoma-pre-Radiotherapy



Melanoma pre-Tx echography (left), post-radiotherapy (right)

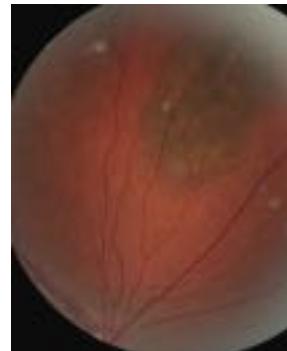


Radiation Retinopathy: exudate, NVD



Choroidal Melanoma Pre-Tx

Acknowledgement: Sherrol Reynolds, OD, FAAO



S/P Radiotherapy



Management of RR

- Avastin/Lucentis/Eylea
- Laser
- Silicone oil at time of Brachytherapy
 - attenuates radiation dose, and may protect against radiation retinopathy



Melanoma Metastasis

Risk factors for metastasis from the choroid

- Thickness > 2 mm
- Symptoms – Flashes, floaters, loss of vision
- Proximity to the optic nerve
- Documented growth



Shields CL, Shields JA. Risk factors for metastasis of small choroidal melanocytic lesions. Ophthalmology 1995.

METASTASIS

BOX 48.1 Sites for Metastatic Uveal Melanoma

■ Liver 93%
■ Lungs 24%
■ Bone 16%
■ Skin 11%
■ Lymph nodes 10%
■ Brain 5%
■ Fellow eye 0%

Multiple sites involved in about half the cases.
In an atypical case consider a second primary tumor.

Ocular Melanoma Quiz

- What is the 10-year mortality rate for patients diagnosed with a large uveal melanoma?
 - 1 %
 - 7%
 - 9%
 - 50%

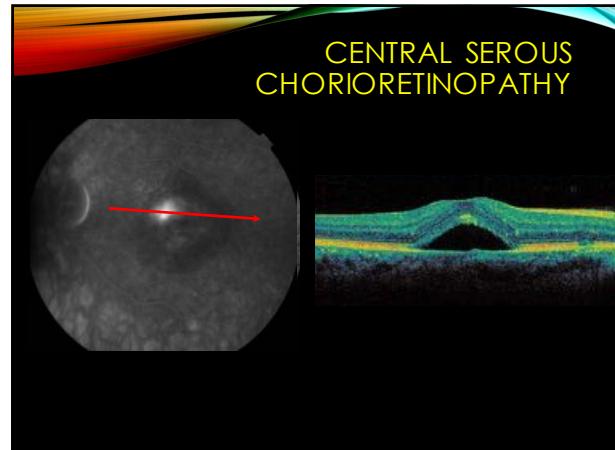
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Questions

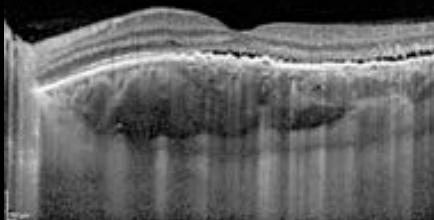


CENTRAL SEROUS CHORIORETINOPATHY



OCT W/EDI IN CSC

PACHYCHOROID AND SUBRETINAL FLUID IN CSC



CSC MANAGEMENT

- Due to the high likelihood of spontaneous resolution, first line therapy for first time CSC remains risk factor modification (reduce stress, d/c steroids) and observation.

CSC MANAGEMENT

- For CSC that persists or returns:
 - In eyes with focal lesions not involving the fovea, focal argon laser treatment may be suitable.
 - In eyes **with foveal involvement**, photodynamic therapy or micropulse diode laser would spare central vision.
 - As our understanding of the mechanism of CSC grows, new therapies, such as a **spironolactone** or **eplerenone**, may prove to be beneficial.
- Pichi F, Carrai P, Ciardella A, Behar-Cohen F, Nucci P. "Comparison of two **mineralcorticosteroids receptor antagonists** for the treatment of central serous chorioretinopathy." *Int Ophthalmol* (2016)

Common Causes of CNV

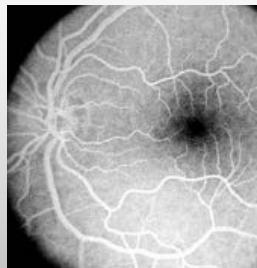
Exudative AMD

- Ocular Histoplasmosis
- High Myopia
- Angioid Streaks
- Choroidal Rupture
- Chronic CSC (less common)



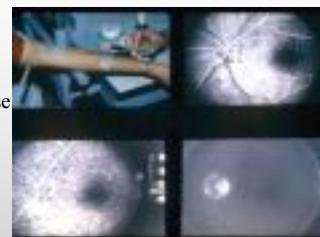
Fluorescein Angiography (FA)

- FA answers the question: is the blood-retinal barrier intact?**

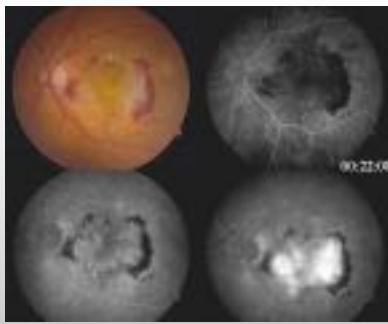


The Fluorescein Angiogram

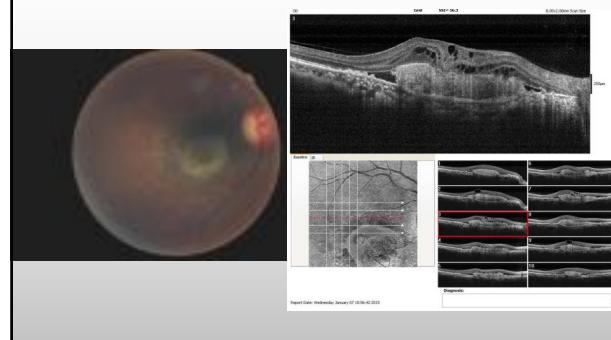
- Stages
 - Choroidal phase**
 - Arterial phase
 - Laminar venous phase
 - Venous phase
 - Recirculatory phase
 - Late phase



CNV in Wet AMD

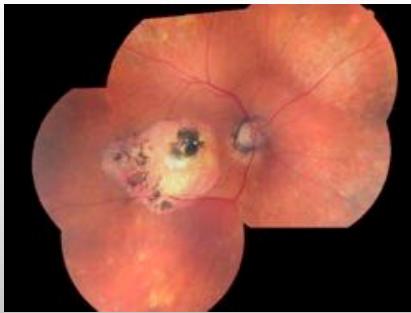


FV Scar

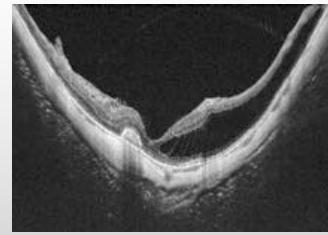


Causes of CNV

- OHS



48 y/o WM, -12.00D

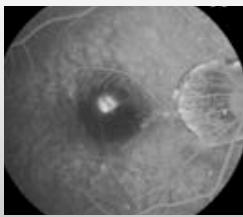
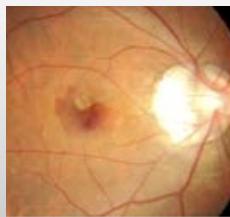


Concave fundus, CNV, schisis

Causes of CNV

- High Myopia in a
52 y/o WM

- CNV w/heme



Choroidal Rupture



ANGIOID STREAKS



- Note Angiod Streaks radiating from the optic discs and macular laser scarring

Differential Dx. of Angiod Streaks: PEPSI

Diagnosis

Friedreich's neuropathy

Key Clinical Features

retardant, "plucked chicken" skin
hypertension

weak peripheral pulses

gastrointestinal bleeding

blue sclera

joint hyperextensibility

fragile, elastic skin

excessive bruising

osteakolateral calcification

bony erosion and abnormal formation

osteoarthritis

hearing loss, vertigo, tinnitus

slurred speech, difficulty swallowing

hemoglobin SS (most frequently)

anemia

neurofibromatosis

Ehlers-Danlos syndrome

Paget's disease

Sickle cell disease

Muscular dystrophy

CNV Variants

Polypoidal Choroidal Vasculopathy (PCV)

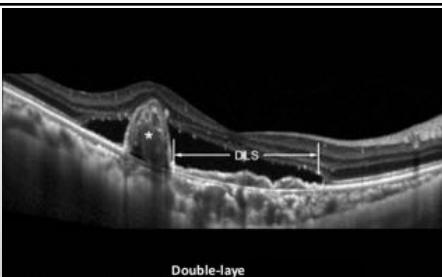
Retinal Angiomatous Proliferation (RAP)



PCV

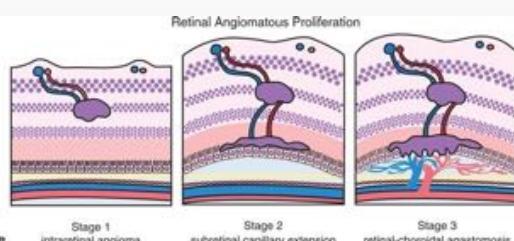


PCV

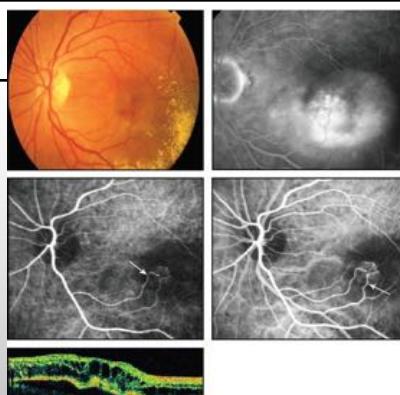


Double-layer
RPE & another highly reflective layer underneath (correlates with BVN)

RAP



RAP



Conclusions

- The choroid is among the eye's most important, yet ignored tissues.
- It is a high-flow vascular structure that provides nourishment to the outer retina.
- Clinicians should be familiar with the various conditions that affect the choroid, as these can lead to blindness or even death.

