THE RED ECLIPSE: RETINAL ARTERIAL MACROANEURYSMS WITH MACULAR INVOLVEMENT

Kevin Phan, OD

Course Outline
I. Course Objectives
   A. Recognize the clinical features of retinal arterial macroaneurysms on a dilated fundus exam and macular OCT.
   B. Discuss management and treatment options currently available for retinal arterial macroaneurysms.
   C. Identify when a referral is necessary for further management with a retinal specialist.

II. Case Presentation (95-year-old Caucasian Male)
   A. Initial Visit
      1. Chief Complaint: Patient reports a large red spot in the center of his vision in his right eye only for the past week. Vision has progressively gotten worse and the size of the red spot has increased since onset.
      2. Pertinent Exam Findings
         i. VA: (OD = 20/800 PHNI; OS = 20/40 PH: 20/30-2)
         ii. Pupil: PERRL (-) APD OU
         iii. Anterior Segment findings and IOP unremarkable OU
         iv. Posterior segment findings
            - Large intraretinal heme within the macula as seen on fundus photo and macular OCT
      3. Differential Diagnosis
         i. Valsalva Retinopathy
         ii. Retinal Arterial Macroaneurysm
         iii. Coat’s Disease
      4. Plan
         i. Referred out to retina specialist due to RAM involving macula and concurrent inferior hemi-retinal vein occlusion
   B. Follow-up # 1 (3 months)
      1. Chief Complaint: Received 2 Anti-VEGF injections with retinal specialist; scheduled for 3rd injection later in the month. Vision has improved overtime with treatment.
      2. Pertinent Exam Findings
         i. VA: (OD = 20/150 with EV PHNI; OS = 20/25)
         ii. Pupil: PERRL (-) APD OU
         iii. Anterior Segment findings and IOP unremarkable OU
         iv. Posterior segment findings
            - Dehemoglobinized fluid within the macula as seen on fundus photo and macular OCT
   C. Follow-up # 2 (3 months)
      1. Chief Complaint: Declined 3 injection due to pain from injection process. Vision has improved overtime and still followed by retina specialist.
      2. Pertinent Exam Findings
         i. VA: (OD = 20/60+2  PH: 20/50-2 ; OS = 20/25)
         ii. Pupil: PERRL (-) APD OU
         iii. Anterior Segment findings and IOP unremarkable OU
         iv. Posterior segment findings
- Residual dehemoglobinized material within the macula as seen on macular OCT

III. Diagnosis and Discussion
   A. Background
      1. Retinal arterial macroaneurysm (RAM) is caused by acquired focal dilations of an arteriole.
      2. These dilations weaken the arteriolar wall and can allow blood to rupture through multiple layers of the retina.
      3. They have the tendency to occur within the temporal arcades of the retina at areas of bifurcation or arteriovenous crossing.
   B. Epidemiology
      1. The incidence of retinal arterial macroaneurysms are rare (1 in 4500).
      2. Typically appear in elderly patients over the age of 60 with a history of systemic vascular disease.
   C. Diagnostic Tools
      1. Optical Coherence Tomography (OCT)
         - Can visualize the RAM and retinal layers involved to a certain extent
      2. Fluorescein Angiography (FA)
         - Shows rapid filling during early arteriole phase, but can vary due to clot formation or scarring from blockage of the lumen by thrombosis
      3. Indocyanine Green Imaging
         - Used if hemorrhage is too obscuring for OCT or FA.

IV. Treatment and Management
   A. Observation
      1. Most cases of RAM have spontaneous regression and can be followed closely.
      2. Cases that threaten the macula or within the macular region should consider additional treatment options to obtain the best visual prognosis for the patient.
   B. Laser photocoagulation
      1. Traditional form of treatment
      2. Complications from laser include enlargement of laser scar, choroidal neovascularization and subretinal fibrosis
   C. Anti-VEGF injections
      1. Newest form of treatment
      2. Studies have shown that patients treated with IV bevacizumab recovered VA and had reduced central retinal thickness faster than those who were under observation only.

V. Clinical Pearls
   A. Most cases of retinal arterial macroaneurysms have spontaneous regression and have a good visual prognosis when outside the macular region.
   B. Severe presentations of retinal arterial macroaneurysm that involve the macula may require intervention and should be sent out for treatment for the best visual prognosis.
   C. Treatment of underlying systemic vascular condition is essential for management of any retinal arterial macroaneurysm.
Adult-Onset Foveomacular Vitelliform Dystrophy: A Clinical Course

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Virtual 2020 Northwest Residents Conference

Disclosures

The Presenter and Organizers for
“Adult-Onset Foveomacular Vitelliform Dystrophy: A Clinical Course”
By Dr. Yen T. Sou

have no financial relationship with any company or products mentioned in this presentation

Learning Objectives

- Identify and describe the stages of adult-onset foveomacular vitelliform dystrophy (AFVD)
- Diagnose AFVD through careful ophthalmoscopy in combination with advanced technology
  - optical coherence tomography and angiography (OCT/A)
  - fundus autofluorescence (FAF)
  - fluorescein angiography (FA)
  - electrophysiological testing (EOG and ERG)
- Discuss management of and future considerations in AFVD

68-year-old Caucasian male

- CC (early 2020): stable reduced vision OD/OS
- Clinical examination:
  - Low compound myopia and astigmatism OU
  - Mild meibomian gland stasis OU
  - 1 nuclear sclerosis OU
  - Fundus shows central pigmentation and atrophy at the fovea OU, (-) hemorrhage or elevation
  - C/D 0.3r OU, unremarkable periphery OU
  - OCT: retinal pigment epithelial (RPE) disruption and geographic atrophy in a bull’s eye pattern OU, (-) fluid
  - H/o abnormal electro-oculogram (EOG) OU in setting of normal full-field electroretinogram (fERG) OU in 2014
  - Followed since 2005, patient was 53-years-old at initial visit

Color fundus montage OD: central foveal lesion with pigment spots at outer edges of lesion (2014)

Color fundus montage OS: central foveal lesion with pigment spots at outer edges of lesion (2014)
**Fundus autofluorescence (FAF)**

**OD/OS: central foveal hyperautofluorescent material** (2014)

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**BCVA Fundus and Autofluorescence (AF)**

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**Demarcated yellow hypopigmented area at fovea OU, w/o drusen, hemorrhage, or subretinal fluid** (2012)

**Central pigment and yellow deposits at fovea OU**

**Heterogeneous hyperreflective dome-like subretinal material at fovea OU**

**Variable acuity**

**Vitelliform lesions with pigment changes OU, hypofluorescent lesions OU**

**Hyperreflective subretinal and subRPE material OU, mild enlargement of noise**

**Increase in size and height of hyperreflective material OU, ellipsoid zone (EZ) and outer retinal largely preserved**

**Mild fluctuations in vitelliform deposits OU**

**Collapse of hyperreflective subretinal material OU compared to 2017**

**Complete resorption of vitelliform lesions**
January 2020

Color fundus photo OD/OS: appearance of central atrophy with pigment clumping in a bull's eye pattern

January 2020

FAF OD/OS: appearance of central hypo- and hyperautofluorescent material in a bull's eye pattern

Pattern Dystrophy

- Dystrophies affecting the retinal pigment epithelium (RPE) includes:
  - *Adult-onset foveomacular vitelliform dystrophy*
  - Butterfly-shaped pigment dystrophy
  - Reticular dystrophy of the retinal pigment epithelium
  - Pseudo-Stargardt pattern dystrophy (multifocal pattern dystrophy simulating Stargardt disease/fundus flavimaculatus)
  - Fundus pulverulentus

Adult-Onset Foveomacular Vitelliform Dystrophy (AFVD)

- Most common form of pattern dystrophy and first described by Gass in 1974
- Late onset: usually diagnosed after the age of 40, avg. age in the 60s
- Symptoms: asymptomatic or mildly blurred vision and/or with metamorphopsia with slow progression
- Clinical findings are highly variable depending on its stage
  - Classical central solitary yellow lesions that gradually increase then decrease in size over years leading to atrophy

Pathophysiology

- Exact underlying mechanism still not well understood
- Presumed vitelliform material accumulation is result of abnormal physiology in the PR-RPE complex
- Photoreceptors degenerate, shedding of outer segment is reduced, RPE cells can clear the vitelliform deposits
- The lesion eventually atrophies accompanied by VA loss
- Possible vascular impairment from reduced vascular flow density at all layers on OCTA
- Characterized by subfoveal choroidal thickening compared with normal eyes and eyes of patients with ARMD
Clinical findings

- BCVA ranges from 20/25 – 20/400
- Mean VA of 20/50 in later stages
- Findings vary depending on the stage
- Usually normal EOG in setting of normal ERG
- Abnormal EOG possible
- Subnormal full-field ERG and suppressed multifocal ERG possible
- Color vision and visual fields are often unremarkable at initial stages
- Complications include geographic atrophy, macular hole and choroidal neovascularization

Genetics

- Inheritance pattern of AFVD is uncertain
- Some suggest autosomal dominant pattern
- Many affected have no family history
- According to several studies, most of AFVD cases do not have a mutation in PRPH2, BEST1, IMPG1/2
- A minority of patients have mutations in these genes that can cause vitelliform macular dystrophy
  - PRPH2 - structural role in outer segment discs in rods and cones
  - BEST1 - proper function in RPE
  - IMPG1 and IMPG2 genes - associated with cones and rods and an interphotoreceptor matrix protein, respectively
- Single-nucleotide polymorphism (SNPs) mutations, commonly associated with age-related macular degeneration (AMD) also puts an individual at risk for AFVD

Natural Course of AFVD

- AFVD shares observable characteristics with Best vitelliform macular dystrophy (BVMD)
- Stages of the disease as proposed for typical BVMD
  - Vitelliform
  - Pseudohypopyon
  - Vitelliruptive
  - Atrophic

Stage: vitelliform

- Most commonly diagnosed at this stage
- Classic clinical findings
  - Bilateral (sometimes unilateral), usually asymmetric
  - Foveal
  - Yellow, solitary, round to oval elevated subretinal lesion
  - Often with central pigmentation

Vitelliform

- A/B color fundus: bilateral, vitelliform, circular, foveal lesions and associated infrared images C/D
- E/F FAF: central hypoAF surrounded by ring of hyperAF
- G/H Red free frames: central white spots
- FA: mottled foveal hyperfluorescence (I/J), late staining without leakage (K and L)
- M OCT (H-cut): disrupted ELM, mottled and hyperreflective PR layer that overlies at subretinal hyperreflective dome-shaped lesion
- N OCT (V-cut): mottled ELM and ellipsoid zone, overlies heterogeneously hyperreflective subretinal material

Vitelliform

- A/F: central hypoAF surrounded by hyperAF
- B OCT: hyperreflective lesion between RPE and ellipsoid zone
- FA: early hypofluorescent (C) and hyperfluorescent from edges towards the center in later phases (D)
- ICGA: hypocyanescent in both early (E) and late phases (F)
- OCT Angiography: subretinal material leads to displacement of blood vessels in superficial (G) and deep capillary plexuses (H)
- Associated decreased blood flow or perfusion at choriocapillaris (I)
Stage: pseudohypopyon

- Clinical findings
  - Yellow-orange lesion with heterogeneous appearance as lesion material moves toward bottom of the fovea
  - Creates fluid layers: usually bottom is the yellow yolk-like fluid, top is clear fluid
  - Increase in size from vitelliform stage

Clinical findings: pseudohypopyon
- Yellow-orange lesion with heterogeneous appearance as lesion material moves toward bottom of the fovea.
- Creates fluid layers: bottom is yellow yolk-like fluid, top is clear fluid.
- Increase in size from vitelliform stage.

Stage: vitelliruptive

- Clinical findings
  - Decrease in lesion size from pseudohypopyon stage
  - "Scrambled" appearance
  - FAF (inset): central hypoautofluorescence surrounded by hyperautofluorescence.
  - SD-OCT: mixed hyperreflective/hyporeflective lesion.
  - Central interface of IS/OS junction disrupted.

Clinical findings: vitelliruptive
- Decrease in lesion size from pseudohypopyon stage.
- "Scrambled" appearance.
- Central interface of IS/OS junction disrupted.
- Overall decrease in lesion size.
Vitelliruptive
- A FAF: central hypoAF
- B FA and (C) ICGA: central hypofluorescence/cyanescence with areas of window defect and staining
- D OCT: flattened lesion by resorption of the majority of fluid between RPE and EZ
- OCTA:
  - Displacement of blood vessels at superficial (E) and deep capillary plexus (F)
  - Decreased density of vessels at choriocapillaris (G)

Stage: atrophic
- Clinical findings: decrease in lesion size
- FAF (left): central hypoAF surrounded by hyperAF (baseline) and mixed hypo/hyperAF (12 mons)
- SD-OCT
  - Baseline: hyperreflective lesion, IS/OS junction (EZ) relatively normal
  - 12 mons: atrophic lesion (complete resorption of vitelliform lesion and IS/OS junction absent at site of lesion)
  - Overall decrease of lesion size

Choroidal Neovascularization
- A FAF: central hyper/hypoAF
- B/C OCT: mixed hyper- and hyporeflective lesion
- D/E FA and F/G ICGA: early and late phase, respectively
  - Central hypofluorescence with areas of window defect and staining but no leakage
  - OCTA:
    - Displacement of blood vessels at superficial (H) and deep capillary plexus (I)
    - Arrow points to CNV with tangled vascular network and decreased vascular density at choriocapillaris (L)

Stages of AFVD
A. Vitelliform
B. Pseudohypopyon
C. Vitelliruptive
D. Atrophic
Prognosis

- Visual prognosis generally favorable
- Patients with progressive central scotoma use self-adaptive strategies to use peripheral retina in place of damaged fovea
- Low vision rehab effective in such cases - maintain and optimize reading ability

Future investigations

- Current challenge includes identifying the underlying cause in the majority of cases
- Although OCTA presented with limitations, a study showed OCTA was able to visualize two cases of CNV that FA and ICGA were not able to pick up
- May provide useful information in pre-therapeutic assessment of AFVD
- New technologies may aid in understanding pathogenesis and lead to future therapeutics like gene therapy

Summary

- Accumulation of subretinal fluid and debris are related to an abnormal function of the PR-RPE complex
- Natural course AFVD progresses from the classic dome-like lesion as it enlarges then collapses leading to outer retinal atrophy
- Single central yellow lesion in early stages differentiate AFVD from ARM and CNV
- Genetic mutations are found in only a minority of AFVD cases
- OCT is regularly utilized to confirm the subfoveal lesion; FA and OCTA utilized together to help confirm CNV
- OCT has limitations
- Unknown potential artifacts, signal strength and final image quality influence by signal masking and mechanical forces exerted by vitreomacular material

References

THANK YOU!

Mentors:
Dr. Bryan Deck
Dr. Dirk Dittemore

Director of Residency Programs:
Dr. Michela Kenning

Residency Programs Coordinator
Nora Garfias
Something's Lurking Underneath the Retina: Pachychoroid Spectrum

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Learning Objectives

- By the end of the presentation, attendees will be able to:
  - Understand the pathophysiology of pachychoroid spectrum diseases
  - Identify pachychoroid
  - Distinguish between the 4 main groups of pachychoroid spectrum
  - Differentiate it between other similar diseases
  - Treat, if necessary/in our scope of practice

Choroid

- The layer between the Retina and Sclera
- Consists of 5 different layers:
  - Bruch Membrane
  - Choriocapillaris
  - Sattler Layer (Layer of small diameter blood vessels)
  - Haller’s Layer (Layer of large diameter blood vessels)
  - Suprachoroid
  - Supplies posterior 1/3 of the retina

What is a Pachychoroid?

- Pachychoroid (pachy-[prefix]: thick) - abnormal and permanent increase in choroidal thickness often showing dilated choroidal vessels and other structural alterations of the normal choroidal architecture
- Eyes with increased thickness of Haller’s layer (pachy-vessel) causing compression of overlying Sattler’s layer and choriocapillaris develop changes in the RPE
- Thickest Subfoveally ~300 microns
- Normal range 191 to 350 microns
- Depends on age, axial length, refractive error, blood pressure, diurnal cycle and some racial variations
- Having a thick choroid does not mean disease but puts people more at risk
- Detection with Swept Source OCT or Spectral Domain OCT with enhanced depth imaging if indocyanine green angiography is not available.

Disclosures

The presenter and Organizers for “Something's Lurking underneath the Retina”

By Dr. Eugene Cheung has no financial relationship with any company or products mentioned in this presentation
4 Main Disease Groups of Pachychoroid Spectrum

1. Pachychoroid pigment epitheliopathy
2. Central serous chorioretinopathy
3. Pachychoroid neovasculopathy
4. Polypoidal choroidal vasculopathy

Case 1

- 61yo Hispanic male
- CC: Vision stable since last exam OU. Mild distortion inferior to visual axis but no changes in amsler grid. No other ocular complaints.
- OCULAR HISTORY:
  - (+) Pigment epithelial detachments (x3) OS
  - Previous FA ruled out neovascular retinitis pigmentosa
  - First noted 2013 at Phoenix VA MC
  - (+)Anatomically narrow angles OU
- No ocular meds
- No Hx/FS
- MHx: DM type 2 and sleep apnea

Exam findings

- Entrance testing: Unremarkable
- Amsler Grid:
  - OD: unremarkable
  - OS: 2 areas of distortion inferior and 1 superior
- Manifest Refraction: No changes
- Anterior segment: Unremarkable other than anterior chamber
  - IOPl 14, 15 in both eyes
- Gonioscopy: safe to dilate
- Posterior Segment: See pictures below
Assessment/Plan

- Pigment epithelial detachments (PED) OS secondary to pachychoroid; stable, longstanding
- Educated patient on pachychoroid and being at higher risk of developing further complications. Discussed due to superior position of PEDs and proximity to fovea that it is extremely important to monitor with home amslor grid. Return in 6 months for dilated exam and macular scan, or immediately if any changes in amslor.

Pachychoroid Pigment Epitheliopathy (PPE)

- Disruption in RPE causing small pigment epithelial detachment
- Thought to be a form fruste or precursor to Central serous chorioretinopathy
- Signs: Localized disruption in RPE on OCT
- Symptoms: blurry or distorted vision, asymptomatic
- Treatment: monitor for self-resolution
- DDx: Dry Age-related macular degeneration, or pattern dystrophy of the RPE
  - But… (Manjunath et. al, 2011)
  - Wet AMD (194.6 μm (n = 40)
  - Dry AMD (213.4 μm (n = 17).

Drusen

- Drusen
  - Metabolite material between RPE and Bruch’s membrane
  - Certain lipids and inflammatory protein
  - “soft drusen”
- Pachydrusen
  - Seen on red-free or blue-light fundus photography
  - More defined borders
  - Seen over thickened choroid with increased Haller thickness with attenuation of choriocapillaris

http://www.ijo.in/article.asp?issn=0301-4738;year=2019;volume=67;issue=3;spage=371;epage=375;aulast=Singh
Central Serous Chorioretinopathy (CSCR)
- Serous detachment of the neurosensory retina
- Signs: Localized serous RD, serous PEDs, RPE disruption, usually unilateral
- Symptoms: Blurred vision, metamorphopsia, central scotomas, asymptomatic
- Etiology: Patients have a thick choroid, increased choroidal vascular area and increased choroidal vascular permeability. In these areas PEDs occur leading to compromise of the blood retinal barrier and localized RPE disruption with subsequent serous RD.
- Risk factors: type A personality, steroid exposure, sleep apnea, male and pregnancy.
- Treatment: monitor for self-resolution, oral antimineralocorticoid (25mg BID), oral carbonic anhydrase inhibitors (500mg TID), anti-VEGF or photodynamic therapy

Pachychoroid Neovasculopathy (PCN)
- Late stage complication of PPE or chronic CSCR
- Hypothesis is long term compression of pachychoroid on to retinal layers causes the Type 1 choroidal neovascular membrane (CNVM)
- Occurs over the areas of increased choroidal thickness
- Types of Neovascularization
  - Type 1: located below the RPE
  - Type 2: passes through the RPE and is located above the RPE in the subretinal space
  - Type 3: within the neurosensory retina
- PCN continued
  - Usually lacks drusen, younger age vs AMD
  - May have surrounding changes consistent with PPE
- Bosquet et. al:
  - 88 patients with chronic CSCR for 1 year
  - 35.6% of their patients with chronic CSCR went on to develop Type 1 neovascularization
- Treatment: Anti-VEGF

Polypoidal choroidal vasculopathy (PCV)
- Serosanguineous detachments of the RPE and exudative changes that can commonly lead to subretinal fibrosis
- Polyp-like dilations in the peripapillary or macular region on indocyanine green angiography
- Demographics:
  - 50 to 65 years old
  - More common in Asian and African populations
  - Asian men are affected more than women
  - Caucasian women are affected more than men
  - Smoking is a risk factor
  - Associated with a history CSCR
- Is likely underdiagnosed due to mimicry of AMD and CSCR
PCV Continued

- Signs: Polyps that can lead to sub retinal bleeding, exudates, CNVM, serosanguinous PEDs
- Symptomatic: reduced acuity, metamorphopsia
- OCT
  - dome-like elevations of RPE with moderate internal reflectivity
  - highly reflective line just below these lesions consistent with location of vascular branching network
  - double-layer sign
- Treatment: Can spontaneously self resolve as monitor in 1-2 months with OCT and FA/ICG or refer for PDT + Anti-VEGF

Summary

- Abnormally dilated blood vessels in Haller's layer
- Compression of Sattler and choriocapillaris
- Compromised RPE
- Complications

Clinical Pearls

- If you have a patient with:
  - Atypical drusenoid PEDs
  - Chronic CSCR
  - Atypical CNVM in younger pt
  - Polypoidal choroidal vasculopathy
  Look for an underlying cause with enhanced depth imaging if ICG angiography is not available.

References

