Macular disease when it’s not AMD

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Disclosures – Leo Semes 2019
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86 YOWM
• Presents with reduced VA OS
• POH: repaired peripheral retinal hole SN OS X
  11 yrs
• Pseudophakic in each eye
• Medicated for HTn X 20 yrs

20/80
What’s the diagnosis and management?

Caliper to measure
Retinal thickness = 462 µ
Retinal angiomaticous proliferation

- Aka Type II neovascularization (intraretinal NV)
- Management
  - Avastin injection ⇒ 20/40 @ 3 weeks
  - 20/60 with 4 additional treatments @ 2 years

Neovascularization
Type I – CNVM
Type II – retinal telangiectasia

Another example (Stage II, w/PED)

A. RAP with hyperfluorescent PED
B. ICGA with RAP hot spot, and hypo-fluorescent PED
C. OCT: intraretinal RAP (arrow), cystic spaces and PED
D. Another RAP with PED
E. Arrow indicates intraretinal lesions, cystic spaces overlay PED

A. ICGA: showing communication among intraretinal, subretinal and choroidal NV.
B. OCT shows choroidal invasion into subretinal space; RAP not well defined

A. RAP (note drusen)
B. OCT suggesting choroidal invasion of subretinal space

Recent update (2017): RAP

Key points for contemporary management

- Anti-VEGF therapy is first line treatment
- Variable treatment response portends a guarded prognosis
- Future research will clarify pathophysiology, definition & classification as well as optimal treatment

Retinal Angiomatous Proliferation

- Questions
- Comments

57 AA F

- Presents to UAB Eye Care with a history of reduced VA (OD>OS) because,
  She had been told elsewhere that she had glaucoma.
- Ocular and family histories non-contributory
- Medical history: Tx for HTN X 12 yrs.
- VA 20/200, 20/80
Rare presentation of *Malattia Leventinese* (aka Dyme’s honeycomb macular dystrophy)


Malattia Leventinese

- Questions
- Comments

55 W/M C/O reduced VA (OD ≈ OS)
BSCVA: 20/25 OD, OS

Genetics and BVMD
(Best vitelliform macular dystrophy)
- AD
- BEST 1 gene that codes for “bestrophin 1”
  - Protein that is involved in membrane integrity
    at the PR/ RPE layers.
  - This loss of function allows accumulation of
    lipofuscin — seen clinically as well as on OCT

Note lipofuscin accumulation beneath the macula (OD)
Management

- Genetic counseling
- Monitor for CNVM – definitive tx (anti-VEGF)
- Genetic testing* / engineering in the future
- OCT, FA, FAF, electro-diagnostic testing to confirm
- Low vision!!!

*EyeGENE

One differential to consider

The so-called flying saucer sign

Ocular Toxicity

- Acute
  - Retina (Photic [solar] Retinopathy, Maculopathy)

  - Sungazing, Unprotected eclipse viewing
  - Operating microscope over-exposure
  - Symptoms and signs
    - History of exposure
    - Decreased vision
    - Central scotoma

20 WM

- Laser injury to face with both eyes potentially involved
  (Micra laser with a legend amplifier, 800 nm, generally set at 2.5 watts, mode lock pulsed laser)
- Persistent after image; no further VA reduction subjectively
- VA 20/20 in each eye

6-week F/U with ultra high resolution imaging
Vertical orientation shows intact and fully formed PIL

Summary

- 7 cases over 2 years; mean age 18.7 yrs (12-26)
- Exposure: several seconds, 5mW green laser pointer
- Presenting VA range: 20/20 to CF; all reported central/paracentral scotomas.

- Distance from source, spot size were not recorded.
  (ME calculation was, therefore, impossible)
Summary

- 5 eyes showed ophthalmoscopic signs of round retinal discoloration (yellowish/orange) at the macula
- 2 eyes showed macular subhyaloid hemorrhage
- 1 eye had FTMH with CME
- Baseline SD-OCT revealed disruption of the PII/ellipsoid zone band, and extended toward the inner aspect of the RPE band.

- Treatment = oral steroids 0.5 mg/kg/day, i.e., 40 mg/40 kg
- All but two eyes returned to 20/20 (20/40, CF 2 m.)
- Follow-up 2-12 mo.

A little different story

(Left) 5/20  Right macula with pigmentary disruption.
(Right) 20/20  Left macula is uninvolved

Close up of right macula. Note proliferative pigmentary response

High-resolution scans showing dimension of the laser injury lesion. Note the disorganization of the outer retina corresponding to other clinical findings.

Laser retinal exposure

- Comments
- Questions
**Back to RAP...**

The following 2 cases appeared in:


- Images courtesy M. Gonzalez, MD

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**47-year-old Caucasian woman**

- Referred to our clinic for evaluation of blurred vision in the right that began 2 years earlier.
- Medical history was remarkable for anxiety (systemic medications: Citalopram Hydrobromide and Clonazepam).
- The remainder of her medical and family histories were non-contributory.
- BCVA: 20/70-2 OD and 20/20 OS.
- Pupillary testing was normal without RAPD.
- Confrontation visual fields were FTFT in each eye and ocular motilities were full and smooth.
- SLE was unremarkable for each eye
- IOP was 10mmHg OD and 12mmHg OS.

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**Fundus images**

Note subtle gray thickening in the paravascular area of each eye (OS > OD)

Note lack of leakage on FA (3:18) but capillary disorganization

Note lack of leakage on FA (3:22) but capillary disorganization
56-year-old Caucasian woman
- Referred for evaluation prior to cataract surgery.
- She reported a decline in vision of each eye for over 2 years.
- Medical history was remarkable for Type 2 diabetes, hypertension, and hyperlipidemia, all for > 10 years.
- Her medications included Janumet, Glybera, Lipitor, Spirulactone and Lisinopril.
- BCVA: 20/25 OD and 20/30 OS.
- Pupillary testing was normal without RAPD.
- Confrontation visual fields were FTFC in each eye and ocular motilities were full and intact in each eye.
- SLE revealed age-appropriate lens changes in each eye.
- IOP was 14mmHg in each eye.

Fundus images
- Note parfoveal discoloration in each eye

Angiogram @ 3:45, Minimal leakage
Following a 5-year interval during which the patient was lost to follow-up...

- S/P Cataract extraction with PCIOL in each eye
- 2-month history of decreasing VA (OD)

Management

- Treatment (OD) with serial intravitreal bevacizumab (Avastin) was offered and following 5 injections, IVFA confirms regression of the extrafoveal SRN in the right eye. Her visual acuity measured 20/40-1 OD.
Recent report on MacTel

- Capillary perfusion density is reduced in all quadrants of all vascular plexi (superficial to deep)
- Capillary perfusion density is significantly reduced in later stages
- Capillary perfusion density is reduced to the greatest extent in the deeper plexus
- MacTel may be a disease of the Mueller cells

One more case...

- 40s AA Male
- Longstanding diagnosis of MS with systemic treatment
- BCVA 20/40, 20/40
- Normal IOP and anterior segment

Suprathreshold VF

“Macular” OCT
• **RESULTS (Cont)**
  
  • OCT showed greater differences in the inferior and temporal RNFL thicknesses in both groups.
  
  • In MS patients only, OCT revealed a moderate correlation between the increase in EDSS [Expanded Disability Status Scale] and temporal and superior RNFL thinning.
  
  • Temporal RNFL thinning based on OCT results was correlated moderately with decreased QOL.

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Some of the latest (February 7, 2017)

- 100 normal, 50 MS patients followed for 5 years
- VA, CV, VF, OCT, SLP [GDx-VCC], VEP & QoL
- **RESULTS**
  
  • Optical coherence tomography (OCT) revealed reduced RNFL thicknesses in both groups.
  
  • In the MS group, changes were detected in average thickness and in the mean deviation using the GDx-VCC and in the P100 latency of visual evoked potentials;
  
  • No changes were detected in visual acuity, color vision or visual fields

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**Comments and clinical application**

- Descending OA may show up early as ganglion-cell damage
  
  - Other neurodegenerative disorders may show sentinel damage in the retina
  - Parkinson
  - Alzheimer
  - Concussion
  - Glaucoma ?!
Thank you