The spectrum of retinal vascular anomalies

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Choroidal Neovascularization Membrane (CNV or CNVM)

- Accounts for <20% of cases of age related macular degeneration (AMD). The common cause of severe vision loss among the elderly.
- Choroidal Neovascularization Types: Classic vs Occult
    - **Classic** CNVM (predominate and minimal)
        - Well defined membrane on FA with homogenous intense leakage
        - About 10% of cases
    - **Occult** CNVM
        - About 90% of cases
        - Ill defined membrane on angiogram
        - Fibrovascular PED: irregular elevation of RPE with speckled hyperflurescence (heterogenous leakage) more intensify in late stage
    - Leakage of undetermined source: FLAT RPE with speckled hyperfluorescence late stages
- Locations
    - CNVM may be subfoveal, juxtafoveal (1-199 microns from center of macula), or extrafoveal (> 200 microns from center of macula)
    - Type I or II
        - Type I: below the RPE. Commonly observed in AMD.
        - Type II: below neurosensory retina. Commonly seen in myopic degeneration and histoplasmosis
- Testing
    - OCT: variable presentations. If see any signs of retinal thicken or edema, suspect a CNV
    - FA and possibly indocyanine green (ICG) imaging: hot spots with late spread of hyperfluorescence.
    - ICG:
        - **Classic** CNV: delineating of the capillary pattern, branching arteriole pattern or mixed pattern
        - **Occult** CNV: demonstrate arteriolized vessels
    - ICG may be indicated to better visualize outline of membrane
    - ICG dye absorbs and emits fluorescence in the near IR spectrum
    - Better able to penetrate hemorrhage, melanin, fluid
    - Better for occult CNVM detection

Other Conditions Associated with Choroidal Neovascular Membrane Formation:

- Wet AMD in the elderly
- Choroidal rupture, ocular histoplasmosis, degenerative myopia, angioid streak, central serous chorioretinopathy (CSC), posterior inflammation or infection, and hereditary condition.

Management

Clinical pearl: FA within 72 hrs because membranes can grow 10 -20 microns/day

PCV: Polypoidal choroidal vasculopathy (PCV)

- First described in 1982 (Yannuzzi et al)
    - Posterior uveal bleeding syndrome
    - Multiple recurrent retinal pigment epithelial detachments in black women

PCV clinical presentation

- Dilated branching inner choroidal vessel + aneurysmal structures at the termination of a network of dilated choroidal veins
    - Recurrent serosanguineous neurosensory detachment of the retinal pigment epithelium (PED)
- Predilection for pigmented races.
    - Highest in Black women and Asian
    - Observed among Hispanic population
- Average age is 50's-70's
- Predilection for juxtapapillary or macula
- Commonly bilateral but can be UNILATERAL
- #1 mis-Dx is AMD

PCV Testing:

- FA appearance may look like an occult CNV
- OCT findings
    - Bolus sing: small elevation associated with the polyp
    - Double layer sign
    - Serosanguineous neurosensory and/or RPE detachmenet with high slopes
- ICG:
    - Abnormal branching choroidal vascular channels with associated dilated aneurysmal polyps.

PCV Management

- Unpredictable natural hx:
    - Spontaneous regression or recurrent with vision loss
- Combination PDT/ Avastin therapy (EVEREST study)

Retinal angiomatous proliferation (RAP)

- Pathological new vessel initiate from intra-retinal capillaries, invading superficial & subretinal/choroid space:
- Found in paramacular area but NOT subfoveal

Clinical presentation of RAP

- Bilateral abnormalities is the ELDERLY (80s)
- Presentation may include: pigment epithelium detachment, intraretinal hemorrhage, exudates, subretinal hemorrhages, possible VH.
- Common misDx is AMD

Stages of RAP

- Abnormal new vascular growth initiated in the intraretinal capillaries.
    - Abnormal vessels grow forward and then back, pulling upon the RPE and eventually breaking through to anastomose with the choriocapilaries (CNV)
        - ***Stage I-*** Capillary proliferation within the retina that originates from the deep capillary plexus in the paramacular region
        - ***Stage II-*** subretinal neovascularization (SRN)
        - ***Stage III.*** CVN seen clinically and angiographically, sometimes in association with a vascularized pigment epithelial detachment (vascularized-PED).)

Treatment options for RAP

- Laser is effective
- Because VEGF is a driven factors & RAP does react well to anti-vegf therapy

<u>PRIMARY RETINAL TELANGIECTASIA</u>

- Group of rare, idiopathic diseases having no known cause or systemic association
- Retinal vascular anomalies affecting the retinal capillaries.
- Pathogenesis: Ectatic (aneurysmal) retinal capillaries that lead to structural retinal changes, including increase leakage. Includes IMT, Coat's disease & Leber's military aneurysm.

- Clinical presentation:
    - Dilation of the retinal vessels
    - Tortuous retinal vessels
    - Multiple aneurysms
    - Varying degree of leakage
    - Deposition of lipid exudates and hemorrhages

Idiopathic Juxtafoveal Retinal Telangiectasia (JRT): Idiopathic macular telangiectasia (IMT)

1. **Group 1**	2. **1A**	3. Visible & **exudative**	4. Unilateral	5. w 90% male 6. w Lipid exuda 7. w No RPE cha 8. w Variant of C 9. w Laser ?effec
	10. **1B**	11. Visible, exudative & focal	12. Unilateral 13. (≤2 clock hours)	

14. **Group 2**	**15. 2A**	16. **Occult** & non- exudative	17. Bilateral (98%)	18. w 50yo both g 19. w Retinal thick 20. w "Right-angle 21. w RPE plaques
	22. **2B**	23. Juvenile occult familial	24. Bilateral	
25. **Group 3**	26. **3A**	27. Occlusive	28. Usually bilateral	29. w Rare 30. w No gender d 31. w Capillary oc 32. § > 1 DD 33. w Associated v 34. systemic disea
	35. **3B**	36. Occlusive with CNS vasculopathy	37. Usually bilateral	

Differential Dx include:

- Type I:BRVO, SCR, DME & HTN
- Type II: AMD
- Type I: leaking form with edema, hemorrhages and exudates. Found <2DD and usually temporal to the fovea
- Type II: pigmentary changes & crystal deposits
- JRT is a rare cause of macular edema and reduced acuity. A developmental anomaly with subsequent leakage. Similar to macroaneurysm, but too close to fovea
- Type II is divided into non proliferative and proliferative
- Vision typically 20/30 or better
- This condition is greatly under-diagnosed
- Always consider this condition in patients presenting with idiopathic parafoveal edema or dot/blot hemorrhages *especially if there is no history of ischemic vascular disease*

JRT/IMT Dx tests

- OCT: shows variable presentations- retinal thickening and migration of the retinal pigment.
    - Type II is described as a draped ILM (superficial retinba) over a cyst (that may seem collapse)
- FA: is required to make the Dx, delineating the abnormal capillary plexus

JRT/IMT Management

- Observation: typically for type II non-proliferative
- Photocoagulation with grid argon green or krypton red if there is progressive loss of vision. Mostly for type I and even type Ii proliferative stage
- Intravitreal injection of Avastin has been used for type I and usually reserved for proliferative type II
- PDT been used in past
- Consider testing for HTN and DM in patients with parafoveal hemorrhaging. If these diseases are not present, then telangiectasia is the likely cause. Keep in mind that there is no strong relationship between this condition and any systemic disease.

Leber's Miliary Aneurysm

- Localized cluster of dilated capillaries and aneurysms and telangiectasia.
    - Similar to Coat's disease- there is no exudation and minimal leakage
    - A variant of Coat's disease
    - Affects one quadrants (superior temporal) in the mid-periphery
    - Male predilection with unilateral presentation presenting in 4^{th}-5^{th} decade of life
    - Typically asymptomatic, unless macula is involved

Coat's Disease

- Formation of telangiectasis and breakdown of the inner blood-retinal barrier are the fundamental causes of all changes found in coat's disease.
- Characteristic Findings:
    - mild to massive aneurysmal exudation
    - Retinal hemorrhage, edema & exudates
    - Capillary changes including increased permeability and non-perfusion.
    - Serous (exudative) retinal detachment can develop as a common complication
- Because of capillary closure at the telangiectasis, retinal neovascularization with subsequent vitreous hemorrhage and traction detachment can also occur.
- Predominately a unilateral presentation affecting males (85%) between the ages of 18 months and 18 years. Affects the retinal periphery
- When massive exudation results from any disease, it is termed C*oat's response.*

- **Coat's Disease: Treatment**
- Treatment is aimed at eliminating abnormal blood vessels & decreasing edema.
- Prognosis is guarded. There is a gradual progression with increasing exudation over time. Final result can be macular edema and/ TOTAL retinal detachment
- Laser photocoagulation or cryoretinopexy to area of telangiectasia/exudation for thermal necrosis of vessels.
- Anti-vegf therapy is being evaluated
- Scleral buckle for RD

Retinal collaterals

- Vessels that develop within the framework of existing retinal vessels to route blood from a hypoperfused vascular territory to a patent vascular bed
- Pathogenesis:
    - Increased intravascular pressure within the capillary network adjacent to the obstructed vessel
    - Enlargement of the vessels
        - Seen ophthalmoscopy
    - Assume same calibre and cellular characteristic as obstructed vessel
    - Occur several weeks after the obstruction
        - Single or multiple
    - Frequently forms across the horizontal raphe

Three types of collateral formation

- Arteriole-Arteriole (A-A)
- Venule-venule (V-V)
    - Venous occlusion
- Arteriovenous (A-V)

Collateral vs Neovascularization Dx tests

- Fluorescein angiography
    - Collaterals DO NOT leak on FA
    - Neovascularization leaks in the early phase
- OCT
- ICG

Variations of Collateral Vessels

- IRMA (intraretinal microvascular abnormalities)
    - Variation of collateral formation in Diabetic Retinopathy
    - Dilated capillaries in areas of nonperfusion
    - Consider pre-neovasculaization
- OPTOCILIARY SHUNT VESSELS
    - Shunt venous blood from the ONH to the choroid
    - Retinochoriodal Collaterals
    - Associated with:
        - CRVO
        - Chronic glaucoma
        - Chronic papilledema
        - Compressive lesion- Glioma & Meningioma
        - ONH Drusen
- ARTERIO-VENOUS MALFORMATION (AVM)
    - rare retinal vascular anomalies
    - Artery and vein communicate without an intervening capillary bed
    - Purest form of retinal vascular shunts
    - unilateral involvement- isolated or widespread

- Complications of AV malformation

    - Intraretinal haemorrhages
    - Exudation
    - Aneurysm formation
    - Vascular occlusion
    - Neovascular glaucoma
    - Vitreous haemorrhage
- **Systemic Association**
    - Racemose Hemangioma
    - Wyburn-Mason Syndrome
        - Coexistence of facial, retinal, orbital, and central nervous system (CNS) AVM

Mgmt:

- Benefits of Collateral Formation
    - Maintain structure and function of the sensory retina
    - Indicate a past or present retinal vascular disease
    - Ascertain underlying systemic cause

❑NEVER photocogulated