Sorting through the Misconceptions of Anomalous Retinal Vessels

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GOAL
Using case presentations, this course will sort thorough the misconceptions of anomalous retinal vascular changes such as retinal collaterals, variation of collaterals (optociliary shunt vessels, IRMA, and AVM), retinal arterial aneurysm, retinal telangiectasia, and familial retinal arterial tortuosity.

LEARNING OBJECTIVES
At the conclusion of this presentation the participant should be able to:

1. Discuss the variable clinical presentation of the entities presented.
2. Familiarize the OD with the use of auxiliary testing (FA, ICG, OCT) in the evaluation of the entities present.
3. Discuss the latest in management and treatment options for the entities presented

ABSTRACT
Anomalous retinal vascular changes can lead be a diagnostic dilemma for many practitioners. Nonetheless, proper identification is important. The clinical features of each entity and the current treatment options available will be reviewed in this presentation.
I. Retinal collaterals

- Vessels that develop within the framework of existing retinal capillary 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 caliber 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**
  - Fluorescein angiography
    - Collaterals DO NOT leak on FA
    - Neovascularization leaks in the early phase
  - OCT
  - ICG

II. Variations of Collateral Vessels

- **IRMA (intraretinal microvascular abnormalities)**
  - Variation of collateral formation in Diabetic Retinopathy
  - Dilated capillaries in areas of nonperfusion
  - Consider pre-neovascularization

- **OPTOCILIARY SHUNT VESSELS**
  - Shunt venous blood from the ONH to the choroid
  - Retinochoroidal 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 hemorrhages
- Exudation
- Aneurysm formation
- Vascular occlusion
- Neovascular glaucoma
- Vitreous hemorrhage

Systemic Association
- Racemose Hemangioma
- Wyburn-Mason Syndrome
  - Coexistence of facial, retinal, orbital, and central nervous system (CNS)

Management:
- 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

III. Retinal Arterial Aneurysm
I. Macroaneurysm (RAM)
- Typically associated with Hypertension or atherosclerosis
- RAM clinical presentation
  - Acquired, focal dilation of a retinal artery
    - Typically occurring within the first three bifurcations of the central retinal artery
  - Abnormal vessel structure, hemorrhage, or exudates, and the retinal location
  - Most commonly observed in elderly females
  - Two types of RAM are described in the literature
    - Fusiform (cuffed)- uniform widening of retinal artery
    - Saccular (blowout)- localized outpouching of the arterial wall
- FA/ICG appearance
  - FA shows weaken of the vasculature at site of macroaneurysms = associated with hyperfluorescence
- OCT
  - Monitor associated retinal edema from RAM and to determine the presence and extent of subretinal fluid, macular edema and hemorrhage.

Management
- Unpredictable natural history:
  - Spontaneous regression or recurrent with vision loss
- Anti-VEGF

II. Microaneurysm
- Small saccular out pouching that involve capillaries
- Typically caused by systemic conditions- Diabetes
III. PRIMARY RETINAL TELANGIECTASIA

- 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 hemorrhage

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<th>Idiopathic Juxtafoveal Retinal Telangiectasia (JRT): Idiopathic macular telangiectasia (IMT)</th>
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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 retina) 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 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 4th-5th 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 Coat’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

**IV. Familial retinal arterial turtousity**

- A/D (FHx)
- Not associated with any other systemic disease
- Only affects the arteries
  - Usually 2nd-3rd order (after 1st bifurcation)
  - Usually progressive and may lead to intermittent hemorrhages