Sorting through the Misconceptions of Anomalous Retinal Vessels

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Course Outline

Anomalous retinal vessels can lead be a diagnostic dilemma for many practitioners. Nonetheless, proper identification is important, as these vessels can be a sign of potentially life-threatening conditions. This course will review and provide current information on common anomalous vessels.

I. Collaterals -Microvessels that develops within the framework of existing retinal capillaries joining obstructed to non-obstructed adjacent vessels.

- Pathogenesis: Increase flow within working capillaries or increased intravascular pressure within the capillary network adjacent to the obstructed vessel
- Clinical Signs: Enlargement of the vessels that assume same caliber and cellular characteristic as obstructed vessel
    - Occur several weeks after the RVO
    - Single or multiple and frequently forms across the horizontal raphe
    - Fluorescein angiography- Collaterals DO NOT leak on FA
    - SDOCT/ OCTA
- Misconceptions
    - Collateral vessels are benign
    - Truth- indicates underlying systemic diseases or prior ocular complication. Warrant medical work-up ~60% of patients developed collateral vascularization in BRVO
    - ONH collaterals are OPTOCILIARY SHUNT VESSELS
        - Shunt vessels- typically artery-vein connection without intervening capillaries
    - Correct term- Retinochoroidal Collaterals
        - CRVO/ Chronic glaucoma/ Chronic papilledema/ Compressive lesion- Glioma & Meningioma/ ONH Drusen
    - IRMA (intraretinal microvascular abnormalities) are NOT collateral vessels
        - Truth- Variation of collateral formation in Diabetic Retinopathy

II. Arterio-venous Malformation (AVM)- true shunt vessels (artery and vein connection). Also known as racemose hemangioma

- Pathogenesis: Rare congenital high-flow vascular anomalies (Magnus 1874).
- Clinical Sign
    - Unilateral involvement- isolated or widespread
    - Marked arterial and venous dilation

        - Type I- involves a capillary plexus connecting artery and vein.
        - Type 2- Direct AVMs with no capillary plexus
        - Type 3- is characterized by widespread retinal AVMs
    - The presence of stage 3 AVM with ipsilateral facial and intracranial vascular malformations is **Wyburn–Mason Syndrome**
    - Complications:
        - Intraretinal hemorrhages, exudation, RVO, aneurysm formation, vitreous hemorrhage, NVG
    - FA and OCT evaluation
- Misconceptions- AVMs are benign
    - AVMs require neurologist consultation/ MRI

III. Retinal Arterial Macroaneursym (RAM)

- Idiopathic or acquired focal dilation of retinal artery
- Within first three bifurcations of the central retinal artery)/ often found on the temporal retina
- Elderly females
- Associated with hypertension or atherosclerosis
    - Up to 80% of patients presenting with RAM are shown to have hypertension
- Clinical finding: Multilayer hemorrhage, exudation, and macular edema)
- FA/ICG
- SD/OCT or OCTA
- Unpredictable natural history
    - Spontaneous regression or recurrent with vision loss
    - Anti-VEGF therapy
    - RAMs are misdiagnosed at a rate of 75%
    - Clinical pearl: look for leakage and/or hemorrhages surrounding an arterial area

IV. Retinal Telangiectasia

- Idiopathic Macular telangiectasia (IMT) / formerly known as idiopathic juxtafoveolar retinal telangiectasia (IJRT)
    - A retinal vascular malformation-irregular ectasia and dilation of retinal capillaries
    - Type I- Aneurysmal telangiectasia
    - Type II- Perifoveal telangiectasia (non-proliferative)
        - OCT- Internal limiting (ILM) drape)
            - Müller cell loss
        - Observation
    - Type II- proliferative (CNV)

- Misconceptions- No medication work-up necessary for IMT
- Rule-out HTN and DM in patients with parafoveal hemorrhaging/microaneurysms. If these diseases are not present, then telangiectasia is the likely cause.

- **Leber's Miliary Aneurysm**
  - Localized cluster of dilated capillaries and aneurysms and telangiectasia.
  - A variant of Coat's disease- there is no exudation and minimal leakage
  - 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**
  - Mild to massive aneurysmal exudation, retinal hemorrhage, edema and exudates
  - Serous (exudative) retinal detachment can develop (common complication)
  - Predominately a unilateral presentation affecting males (85%) between the ages of 18 months and 18 years.
  - Affects the retinal periphery

V. Familial retinal arterial tortuosity (Beyer 1958)

- Autosomal dominant (AD) disorder
- Clinical finding-Tortuous small retinal arterioles with normal veins
  - Intra/preretinal hemorrhages
- Usually 2^{nd}-3^{rd} order (after 1^{st} bifurcation)
- May be associated with coarctation of the aorta

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