RETINAL VENOUS OCCLUSIONS

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CHAPTER CONTENTS

INTRODUCTION

This chapter will discuss the following:

1. Central Retinal Vein Occlusion (CRVO)
    a. Ischaemic (non-perfused)
    b. Non-Ischaemic (perfused)
2. Hemi-Retinal Vein Occlusion
    a. Ischaemic (non-perfused)
    b. Non-Ischaemic (perfused)
3. Branch Retinal Vein Occlusion (BRVO)
    a. Ischaemic (non-perfused)
    b. Non-Ischaemic (perfused)

PATHOPHYSIOLOGY

Arteriosclerosis: Loss of elasticity, thickening and hardening of arteries and arterioles

- Thickening of the arteriole compresses the vein because both vessels share a common adventitial sheath at the AV crossing (BRVO) or posterior to the lamina cribrosa (CRVO)
- Results in turbulent blood flow, endothelial damage and thrombus formation

Retinal vein occlusions generally follow this sequence:
Vein occlusion → elevated venous and capillary pressure → stagnation of flow → hypoxia of retina → capillary endothelial damage → haemorrhage → further stagnation → hypoxia → tissue damage

CENTRAL RETINAL VEIN OCCLUSION

- Obstruction of the central retinal vein at lamina – central retinal artery and central retinal vein share the same connective sheath in lamina but not in optic nerve
- Arteriosclerosis creates less space for the vein
- Thrombus builds up from turbulent blood flow and the venous pressure elevates
- Blood flow becomes stagnant and this leads to hypoxia of the retina

Causes:

- Systemic
  - Vascular disease: Systemic hypertension, Diabetes Mellitus, Hyperlipidemia, Giant Cell Arteritis
  - Less commonly: hyperlipidemia, polycythemia, carotid artery insufficiency
- Ocular
  - Primary Open Angle Glaucoma
  - Ocular Hypertension
  - Optic nerve head drusen

Classification:

- Ischaemic: > 10 DD capillary non-perfusion determined by fluorescein angiogram
- Non-Ischaemic: < 10 DD capillary non-perfusion determined by fluorescein angiogram
- Size is based on fluorescein angiogram appearance

ISCHAEMIC CRVO

- **Symptoms:**
  - Unilateral, sudden and severe vision loss
  - Visual acuity < 20/200
- **Signs:**
  - Relative Afferent Pupillary Defect (RAPD) may be present
  - Fundus:
    - Marked tortuosity and engorgement of veins in all 4 retinal quadrants
    - Dot/blot and flame-shaped haemorrhages
    - Severe optic disc oedema
    - Macular oedema and haemorrhages
    - Hard exudates
    - Cotton-wool spots
- **Treatment and Management**
  - Fluorescein angiogram
  - Retinal neovascularization:
    - Panretinal photocoagulation
    - Intravitreal Injections of Anti-VEGF
  - Macular oedema:

        - Focal Laser
        - Intravitreal Injections:
            - Anti-VEGF
            - Triamcinolone (steroid)
    - Treat underlying systemic association (ie. systemic hypertension, cardiovascular disease, autoimmune disease)
- **Prognosis and Follow-up:**
    - **Monthly follow-up examinations to assess for development of neovascularization and macular oedema**
    - Acute signs resolve within 6-12 months
    - Chronic macular oedema is common
    - Optic disc collaterals (blood from optic disc being shunted from retinal veins to choroidal circulation; blood exits through vortex veins)
    - Chronic open angle glaucoma may develop from poor optic nerve vascular perfusion
    - Retinal neovascularization likely to develop secondary to retinal hypoxia
    - Rubeosis iridis in 50% of cases develops in first 2-4 months

NON-ISCHAEMIC CRVO

- **Most common CRVO**
- **Symptoms**
    - Unilateral vision loss, usually better than 20/200
- **Signs**
    - RAPD usually **absent**
    - Tortuosity and dilation in all four quadrants
    - Dot/blot and flame haemorrhages
    - Mild-moderate disc oedema
    - < 10 DD capillary non-perfusion
- **Treatment and Management**
    - Fluorescein angiogram
    - IOP monitoring if IOP rises
    - Treat underlying systemic condition
    - Retinal management is needed if conversion to ischaemic form occurs or if macular oedema occurs
    - Macular oedema management:
        - Focal laser
        - Triamcinolone
- **Prognosis and Follow-up:**
    - **Monitor every 1 month for first 6-12 months (depending on severity)**
        - **Be aware of development of '90 DAY GLAUCOMA'**
            - **Neovascularization of the retina, iris and angle can develop as a response to retinal ischaemia**
            - **If neovascularization develops in the angle and obstructs the trabecular meshwork causing decreased outflow – IOP will rise leading to a risk of glaucoma if IOP is not controlled**
    - Acute signs resolve in 6-12 months
        - Acuity will return back to normal in 50% of the cases
    - Conversion to ischaemia is most rapid in the first 4 months
    - Can develop chronic macular oedema
    - Residual findings (after resolution)
        - Disc collaterals (blood from optic disc being shunted from retinal veins to choroidal circulation; blood exits through vortex veins)
        - Epiretinal membrane
        - Macular pigmentary changes

COMPARISON OF ISCHAEMIC VS NON ISCHAEMIC VASCULAR OCCLUSION

Test	Non-Ischaemic	Ischaemic
Visual Acuity	Usually better than 20/200	Usually worse than 20/200
Pupils	(-) RAPD	(+) RAPD
Visual field test		
Fluorescein Angiography	Area of non-perfusion less than 10 DD	Area of non-perfusion greater than 10 DD
Fundus Appearance	Absence of: cotton-wool spots, flame-shaped haemorrhages, macular oedema	Presence of: cotton-wool spots, flame-shaped haemorrhages, macular oedema

Figure 1: *Comparison of ischaemic vs. non ischaemic vascular occlusion*

IMAGES OF NON-PERFUSION VS. PERFUSION

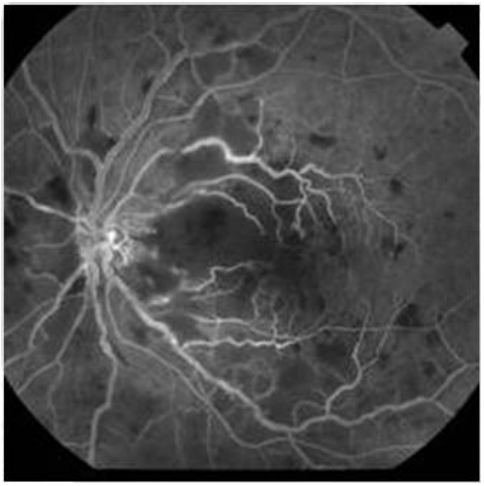

Figure 2: *> 10DD of non-perfusion on fluorescein angiography*

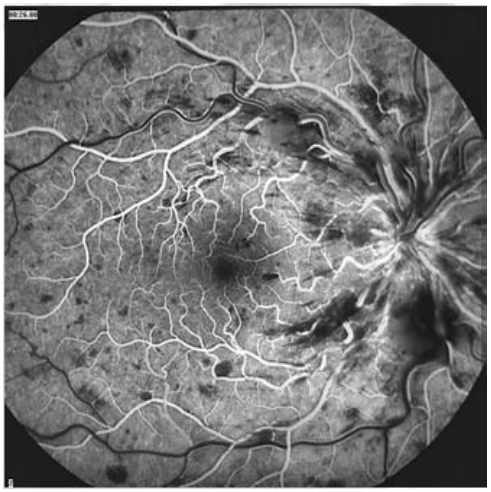

Figure 3: *< 10 DD of non-perfusion on fluorescein angiogram*

HEMIRETINAL VEIN OCCLUSION

- Accounts for approximately 5-13% of vein occlusions
- Also referred to as an altitudinal BRVO
- Similar pathology and clinical picture to CRVO but only affects one hemi-sphere of the retina
    - Occlusion of the central retinal vein at its bifurcation location at the lamina cribrosa
    - Superior or inferior retinal vein becomes blocked at the lamina cribrosa
    - Causes turbulence and deposition of material on the vessel wall resulting in stagnant blood flow
- **Non-Ischaemic (67% of cases)**
    - <10 DD capillary non-perfusion
    - Arterial attenuation throughout the retina
    - Dot/blot haemorrhages out to periphery in one hemisphere of retina
    - Optic disc oedema, intraretinal oedema and macular oedema
    - Dilated and tortuous veins in one hemisphere of the retina
- **Ischaemic (33% of cases)**
    - >10 DD of capillary non-perfusion
    - Arterial attenuation throughout the retina
    - Dot/blot and flame-shaped haemorrhages to periphery in one hemisphere
    - Dilated and tortuous veins in one hemisphere of the retina, arterial attenuation
    - Macular oedema is likely
    - Cotton-wool spots
    - Optic disc oedema
    - Threat:
        - Neovascularization of the disc, retina, iris, angle
        - Neovascular Glaucoma
- **Course and Prognosis for non-ischaemic and ischaemic HRVO:**
    - Optic disc collaterals
    - Macular oedema and retinal hypoxia
    - Neovascularization may develop in 6 months
- **Management:**
    - Fluorescein angiography
    - Monitor monthly for development of neovascularization and Neovascular Glaucoma (for at least first 6 months)
    - Treat underlying systemic disease: (ie. systemic hypertension, cardiovascular disease, autoimmune disease)
    - Management of neovascularization:
        - Panretinal photocoagulation
        - Intravitreal injections
            - Anti-VEGF
    - Management of macular oedema
        - Panretinal photocoagulation
        - Focal laser
        - Intravitreal injections
            - Triamcinolone (steroid)
            - Anti-VEGF

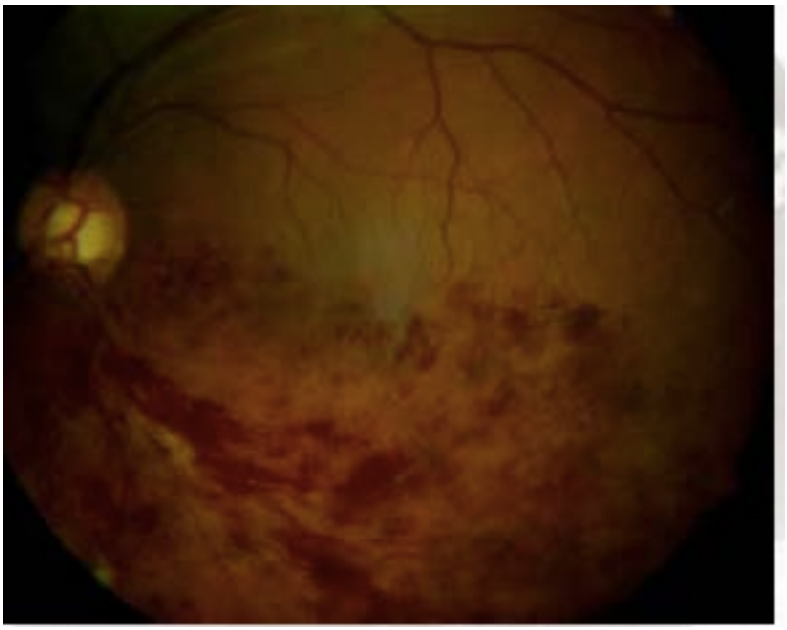

Figure 4: *Hemiretinal vein occlusion*

BRANCH RETINAL VEIN OCCLUSION

- Second most common retinal vascular disease after diabetes
- 60-70 year old, most commonly
- Strong association with: systemic hypertension (50-70% of the cases), cardiovascular disease, diabetes, Open angle glaucoma
- **Different Classifications dependent on occlusion location:**
    - **Major Branch Occlusion:**
        - Occlusion of first order temporal branch at optic disc OR
        - Occlusion of first order temporal branch away from disc but involving branches to macula
    - **Minor Macular Branch Occlusion:**
        - Occlusion involves vessel involved in the macular branch
    - **Peripheral Branch Occlusion:**
        - Occlusion of a vein that does not involve the macula
        - Will be seen in mid-periphery
- **Symptoms**
    - Symptoms depend on the site and severity of the occlusion
        - May be asymptomatic if no macular involvement, however visual blurring involving the sector of visual field corresponding to the area of the retina involved can occur
        - Sudden-onset blurred vision if macula is involved
    - Relative visual field defect
- **Signs**
    - Visual acuity: variable and dependent on macular involvement
        - (6/6 - worse than 6/120)
    - Initial retinal findings:
        - Dilated, tortuous veins distal to A/V crossing (occlusion)
        - Sector of intraretinal and flame-shaped haemorrhages (wedge-shape with apex pointing to occlusion site)
        - Haemorrhages do not cross horizontal raphe
        - Exudates, cotton-wool spots, macular oedema – dependent on amount of non-perfusion
    - Later stages
        - If non-perfused: Development of venous collaterals, optic disc collaterals or neovascularization may occur
        -

- Non-Ischaemic vs. Ischaemic
    - Non-Ischaemic: If < 5 DD of non-perfusion on fluorescein angiogram
    - Ischaemic: If > 5 DD of non-perfusion on fluorescein angiogram = ischaemic
        - Conversion occurs in up to 30% of BRVO patients
        - Usually occurs within first 6-12 months, but can occur years later
        - Can lead to: vitreous haemorrhage, tractional retinal detachment, neovascular glaucoma (same time frame as CRVO)
- **Prognosis:**
    - Resolution takes 6-12 months
    - Venous sheathing and vascular sclerosis can occur peripheral to the occlusion
    - Some haemorrhages may persist for years
    - Collateral retinal venous channels develop in areas that are non-perfused (ischaemic)
    - Macular RPE changes
    - Epiretinal membrane may form
    - Severe complications:
        - Neovascular glaucoma
        - Chronic macular oedema
            - Most common cause of poor acuity following BRVO
            - Associated with hard exudates and microaneursyms
- **Management**
    - Ocular:
        - Fluorescein angiogram
        - Monitor for chronic macular oedema
            - Treatment:
                - Laser photocoagulation (grid laser)
                - Intravitreal kenalog injections
                - Intravitreal Lucentis (ranibizumab)
        - Monitor for development of neovascular glaucoma
            - Panretinal photocoagulation
    - Systemic:
        - Strong association with systemic hypertension, hyperlipidemia, Diabetes Mellitus and cardiovascular disease
        - If > 50 years old:
            - Hypertension (BP and pulse **in office**)
            - Diabetes Mellitus
            - Hyperlipidemia
            - Carotid artery disease (if elderly >65)
            - Consider:
                - Hyperviscosity syndromes, hypergammaglobulinemia, Polycythemia, Leukemia
        - If < 50 years old consider *adding:*
        - Homocysteine
        - Anti-Phospholipid Antibody (APA), Antinuclear antibody (ANA), Erythrocyte Sedimentation Rate (ESR)
        - Other autoimmune disease
        - Protein S and protein C deficiency

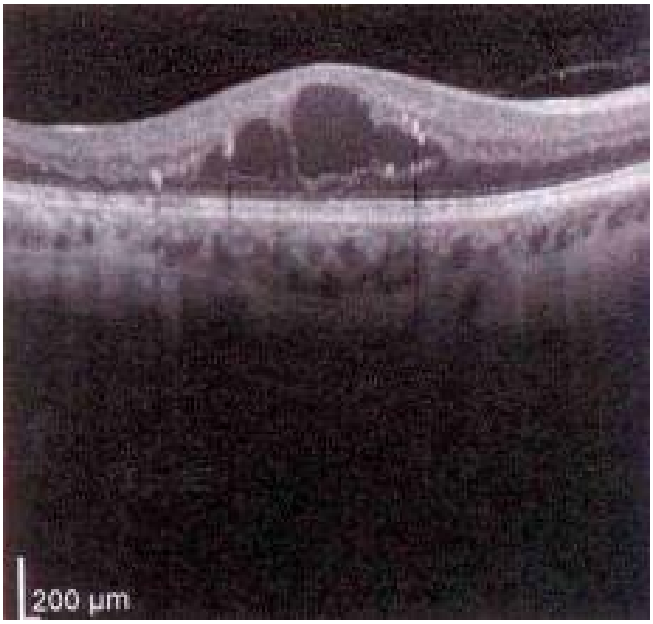

Figure 5: *Chronic macular oedema*

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