Vasculites rétiniennes: présentations cliniques et diagnostic

Dr Jean Vaudaux
Morges et RétinElysée, Lausanne

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Plan

• Definitions
• Epidemiology
• Clinical features
• Diagnosis – workup
• Conclusions – take home messages
Definitions

• Descriptive term for evidence of ocular inflammation and retinal vascular changes

• Perivascular sheathing and vascular leakage or occlusion on fluorescein angiography


• May be associated with infectious disease, systemic inflammatory disease, neoplasia or as an isolated, retina-restricted phenomenon (idiopathic retinal vasculitis)
Epidemiology – retinal vasculitis

- Prevalence is difficult to assess because of the lack of standardized definition

- 25-65% of patients with retinal vasculitis have an associated systemic disease; depends on geographical zone (Behçet’s)
Epidemiology – retinal vasculitis

- Incidence USA 1-2/100’000/yr (isolated retinal vasculitis and associated with systemic disease)
  

- Slightly female predominance (54%)
  Bilateral in 75% of patients
  

- 1 of 8 patients with uveitis has associated retinal vasculitis
  
Epidemiology – retinal vasculitis

• Retinal vasculitis associated with systemic vasculitis is rare (1.4%)
• Common systemic inflammatory associations include Behçet’s disease (30%), sarcoidosis (9%), multiple sclerosis (16%)
• Purely ocular inflammatory diseases commonly associated with retinal vasculitis: birdshot (27%) and pars planitis (19%)  
  
Clinical presentation - symptoms

• Asymptomatic: peripheral involvement, no associated vitritis, no retinal ischemia, no associated CME

• Symptomatic
  - floaters (associated vitritis)
  - decreased visual acuity (macular edema, dense vitritis)
  - scotomata (vascular occlusion, retinal ischemia)
Clinical presentation - signs

Active disease:

- Sheathing/cuffing of retinal blood vessels
- Signs of occlusive vasculitis/retinal ischemia:
  - cotton-wool spots
  - retinal edema
  - retinal whitening
  - intraretinal hemorrhage
Clinical presentation - signs

Active disease:

- Sheathing/cuffing of retinal blood vessels
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Clinical presentation - signs

Inactive disease – consequences of vascular occlusion and ischemia:

- Small vessel/capillary changes, anastomoses
- Ghost vessels (Behçet’s, SLE, syphilis)
- Neovascularization (Behçet’s, sarcoidosis)
- Retinal thinning/atrophy (CMV, ARN, toxo)
- Optic nerve atrophy (CMV, ARN, Behçet’s)
Clinical presentation - signs

- Associated intraocular inflammatory features:
  - Anterior uveitis
  - Vitritis
  - Retinitis (retinal infiltrates, retinal necrosis, retinal edema/whitening)
  - Choroidal involvement
  - Papillitis
Ocular workup – fluorescein angiography

• Mandatory in the assessment of retinal vasculitis
• More accurate assessment of retinal periphery
• May be more sensitive than clinical examination alone to reveal the extent of retinal vascular involvement
• Identification of retinal ischemia, capillary non-perfusion, and vascular occlusion
• Assessment of early/late complications (macular edema, neovascularization, macular ischemia)
Ocular workup – fluorescein angiography

- Vascular leakage secondary to inner blood-retinal barrier breakdown – diffuse or focal
- Staining of the blood vessel walls – diffuse or focal
- Optic disk leakage
- Cystoid macular edema
Ocular workup – fluorescein angiography

- Vascular occlusion
- Retinal ischemia
- Macular ischemia
- Neovascularization (retinal, optic disk)
Ocular workup – fluorescein angiography

Wide-field imaging:
Particularly to assess peripheral ischemia and neovascularization

HRA 2 w/150° Staurenghi lens SLO 230
Ocular workup – ICG angiography

- Assessment of choroidal involvement
- Assessment of choroidal vasculature
- Useful to detect choroidal changes that may be clinically subtle → diagnostic value
  - Birdshot chorioretinopathy
  - Sarcoidosis
  - Tuberculosis
  - Syphilis
Ocular workup – OCT-A

• Diagnosis and monitoring of:
  - macular ischemia/non-perfusion
  - neovascularization (retinal, pre-retinal, optic disk, subretinal, choroidal)
  - subtle posterior pole vascular changes (capillary hypoperfusion or non-perfusion, capillary loops, telangiectasias; changes in both superficial and deep capillary plexuses
Diagnosis

• Clinical presentation
• Angiography: mandatory!
• OCT: monitoring of CME or vitreo-macular interface
• OCT-A
• Laboratory workup: based on clinical and angiographic findings; always rule out TBC and syphilis!
<table>
<thead>
<tr>
<th></th>
<th>Inflammatory</th>
<th>Infectious</th>
<th>Other/Not classified</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Arteritis</strong></td>
<td>Systemic vasculitis (SLE, PAN, granulomatosis with polyangiitis = Wegener, Churg-Strauss)</td>
<td>ARN (HSV, VZV) CMV Toxoplasmosis</td>
<td>IRVAN</td>
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<tr>
<td><strong>Phlebitis</strong></td>
<td>Behçet’s Sarcoïdosis Multiple sclerosis Pars planitis JIA Birdshot</td>
<td>Tuberculosis Eale’s disease</td>
<td>Idiopathic retinal vasculitis</td>
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<tr>
<td><strong>Mixed - arteritis and phlebitis</strong></td>
<td>Granulomatosis with polyangiitis Inflammatory bowel disease (Crohn’s, ulcerative colitis) Relapsing polychondritis</td>
<td>Tuberculosis</td>
<td>Hemorrhagic occlusive retinal vasculitis (HORV, vancomycin)</td>
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<td><strong>Retinal infiltrates</strong></td>
<td>Behçet’s</td>
<td>Tuberculosis Syphilis Cat-scratch disease</td>
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<tr>
<td><strong>Retinal necrosis</strong></td>
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<tr>
<td><strong>Cotton-wool spots</strong></td>
<td>Systemic vasculitis (SLE, PAN, granulomatosis with polyangiitis, Churg-Strauss) Behçet’s</td>
<td>HIV retinopathy</td>
<td></td>
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<td><strong>Vascular occlusion (FA)</strong></td>
<td>Behçet’s (primarily BRVO) Sarcoïdosis SLE, antiphospholipid antibody syndrome (Multiple sclerosis)</td>
<td>Toxoplasmosis ARN (HSV, VZV) Tuberculosis, Eale’s disease Cat-scratch disease</td>
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<td><strong>Ghost vessels</strong></td>
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<td>Syphilis Tuberculosis</td>
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</table>
Conclusion – messages to take home

• Descriptive diagnosis
• Wide clinical spectrum
• May be isolated, or associated with:
  - systemic inflammatory disease (Behçet’s, sarcoidosis), infectious disease (syphilis, ARN, CMV, toxoplasmosis, tuberculosis), neoplasia (lymphoma)
• Only rarely associated with systemic vasculitides
• Epidemiology primarily depends on geographical location
Conclusion – messages to take home

• Non-occlusive versus occlusive
• Ocular and visual morbidity secondary to complications (neovascularization, vitreous hemorrhage, CME), usually when occlusion is present → worse prognosis
• Angiography is mandatory, use wide-field imaging whenever available
• Multimodal imaging
• Systemic workup is necessary & multidisciplinary
• Clue to DDx: type of vessel involved
What sign IS NOT highly suggestive of occlusive retinal vasculitis:

- A) Cotton-wool spots
- B) Intraretinal hemorrhages
- C) Vascular wall staining on fluorescein angiography
- D) Capillary non-perfusion on fluorescein angiography
- E) Retinal neovascularization
Quizz #1

What sign IS NOT highly suggestive of occlusive retinal vasculitis:

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- C) *Vascular wall staining on fluorescein angiography*
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Quizz #2

Which of the following diseases is associated with predominantly arterial retinal involvement:

- A) Behçet’s disease
- B) Sarcoidosis
- C) Multiple sclerosis
- D) Birdshot chorioretinopathy
- E) Systemic lupus erythematosus
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Quizz #3

Which of the following regarding Behçet’s disease is FALSE (only one answer):

- A) May present with non-occlusive or occlusive retinal vasculitis
- B) Complications may include retinal vascular anastomoses
- C) Vasculitis predominantly involves retinal arterioles
- D) Complications may include optic disk neovascularization
- E) May present with isolated branch retinal vein occlusion
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Quizz #4

Which of the following is TRUE regarding retinal vasculitis (only one answer):

- A) Is never occlusive in ocular sarcoidosis
- B) Presents as severe occlusive retinal phlebitis in ARN syndrome
- C) When associated with systemic disease, retinal vasculitis is almost always associated with systemic vasculitis
- D) Vascular leakage = disruption of the external blood-retinal barrier
- E) None of the above
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