

Retinal Vasculitis

Retinal vasculitis is characterized by inflammatory involvement of retinal arterioles, venules and/or capillaries. It is commonly associated with (posterior, or pan-) uveitis.

Diagnoses of retinal vasculitis (retinal biopsy - not commonly done -high risk of damage):

(1) **Dilated eye examination:** Sheathing, perivascular cuffing & exudation, vascular occlusion... +

(2) **Retinal imaging** e.g. fluorescein angiogram: stains the vessel wall or leaks beyond the vessel or optical coherence tomography angiography [OCTA]

Classification based on etiology

(1) Infectious cases

Bacterial disorders

- Tuberculosis: Esp in young Asian Indian males
- Syphilis
- Lyme (Borrelia)
- Cat-scratch disease (Bartonella): BRAO can also be seen
- Brucellosis
- Rickettsial disease
- Leptospirosis
- Whipple's disease
- Exogenous/endogenous infection (e.g. endophthalmitis)



(2) Non-infectious causes

Autoimmune systemic disorders

• Primary Vasculitis

Behcet's disease**

- ANCA-associated vasculitis (EGPA>GPA>MPA)
- Takayasu's arteritis
- Polyarteritis nodosa
- CNS vasculitis

• Sarcoidosis**

- Collagen Vascular disorders: **SLE***, RA, Sjogren's syndrome, Inflammatory myositis, systemic sclerosis
- HLA B27-associated uveitis
- Antiphospholipid syndrome
- Crohn's disease
- Susac syndrome (Encephalopathy, BRAO, hearing loss)
- Multiple Sclerosis
- RVCL (retinal vasculopathy with cerebral leukodystrophy - mutation in TREX1 gene)
- A20 haploinsufficiency (Behçet's-like disease - mutations in TNFAIP3)
- Vogt-Koyanagi-Harada disease



Viral disorders

- Herpes: HSV, VZV, EBV, CMV
- RNA viruses: Dengue, West Nile, Chikungunya
- HIV
- HTLV1
- Hepatitis B and C
- Rift valley fever (Bunyaviridae) - in Africa



Isolated retinal disorders

• Idiopathic (~5-20%)

- Birdshot chorioretinopathy (strong association with HLA-A29 allele)
- Eales' disease (hypersensitivity response to mycobacterial antigen)
- IRVAN - Idiopathic recurrent retinal branch occlusion
- ADNIV - Autosomal Dominant Neovascular Inflammatory Vitreoretinopathy; Causes anterior chamber + vitreous inflammation and retinal + iris neovascularization mutations in Calpain 5 [CAPN5] gene)



Parasitic disorders

- Toxoplasmosis (Host: Cats): Most common infectious cause
- Toxocara



Other:

- Drug-induced vasculitis (e.g. Hep C Rx, checkpoint inh)
- Tattoo-related vasculitis
- Vaccination-associated vasculitis



Fungal disorders

- Candidiasis
- Coccidioidomycosis
- Amoebiasis
- Histoplasmosis



Hematologic disorders (Mimickers)

- Leukemia
- Ocular lymphoma
- Paraneoplastic syndromes



Investigations: R/o infectious & non-infectious causes

Baseline: Complete blood count, ALT, albumin, calcium, lytes, CRP (+/-ESR), creatinine, urinalysis

Basic Immunologic: ANA, C3 and C4, RF, ANCA, ACE; *To consider:* ENA, APL Ab, HLA-B51/A29 ...

Basic infectious: TB quantiferon (TB skin test), Hep B/C, HIV, VDRL; *To consider:* Toxo, Lyme ...

Imaging: Chest X-ray +/-CT chest, Brain imaging (Brain + orbit MRI)

+/- **Aqueous fluid analysis** (by eye specialist): HSV, VZV, CMV, Toxo PCR + cytology

Treatment of isolated primary retinal vasculitis:

Local + systemic steroids; **Steroid-sparing agents:** Methotrexate, Azathioprine, Mycophenolate Mofetil, Cyclosporine, Tacrolimus; **Refractory cases:** anti-TNF (e.g. Infliximab), Cyclophosphamide, Tocilizumab, Secukinumab

