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)

Viral disorders



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

Parasitic disorders



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

Fungal disorders



- Candidiasis
- Coccidiodomycosis
- Amoebiasis
- Histoplasmosis

(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

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)

Other:



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

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

Adapted from: Agarwal A et al. J Clin Med. 2022 Apr 30;11(9):2525; CAVALI textbook by Dr C. Pagnoux

Created by M. Soowamber