Retinal Manifestations of Systemic Disease – Part 1
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The Retina and Systemic diseases
- Retinitis/Vasculitis
- Vitreous cells
- Serous detachments
- Choroidal lesions
- Pigmentary retinopathies
- Choroidal folds/Choroidal masses
- Retinal Vascular abnormalities
  - May be unilateral or bilateral
  - May or may not have systemic symptoms

Retinitis Differential Diagnosis
Infectious:
- Viral -
  - HSV/HZV - Acute Retinal Necrosis (ARN), Progressive Outer Retinal Necrosis (PORN)
  - CMV
- Fungal - especially Candida
- Bacterial - Syphilis, Tuberculosis, Septic embolus, Bartonella, Meningococcus, Cat-scratch (Bartonella)
- Parasitic - Toxoplasmosis, Cysticercosis, Onchocerciasis
Inflammatory:
- Sarcoidosis
- Behcet’s disease
Neoplastic:
- Masquerade syndromes - Lymphoma

Retinal Vasculitis
Retinal Phlebitis
- Sarcoidosis
- Tuberculosis
- Syphilis
- Multiple sclerosis
- Pars planitis
- Eales’ disease
- Antiphospholipid antibody syndrome
- HIV
- Frosted branch angiitis

Retinal Arteritis
- Behcet’s disease
- Collagen vascular disease – Systemic Lupus, Polyarteritis nodosa, Wegener’s granulomatosis
- Toxoplasmosis
- Viral – HZV/HSV/CMV/HIV
- Antiphospholipid ab synd.
- Eales’ disease
- Syphilis
Lab Results:

- RPR Positive
- FTA-ABS Positive
- Treponemal antibody current best test
- Diagnosis: Syphilitic Retinitis
- Treated with Neurosyphilis regimen
- IV PCN G 2 million units q 4 hours for 14 days

Inner Retinitis

Bacterial Septic Embolus

Septic Bacterial Retinitis with overlying vitreous involvement

Roth spot

White centered hemorrhage, fibrin/immune complex. Also in leukemia, vasculitis, diabetes, but non-specific.
Endogenous Candidal Infection

Systemic Fungal infection

Classic "fluff balls"

Acute Retinal Necrosis

Patchy peripheral retina that coalesce. Vitreous haze/inflammation. HZV/HSV. Immunocompetent patients.

Acute Retinal Necrosis (ARN)

Note: Vitreous haze

ARN

Hemorrhage often seen with retinitis

Acute Retinal Necrosis (ARN) OCT (Inferior)
Progressive Retinal Necrosis – usually HZV/HSV. PCR test helpful.

Clear vitreous, immunocompromised patients.

Progressive Outer Retinal Necrosis

Note: Clear Vitreous

Rocky Mountain Spotted Fever

Rickettsial infection

Retinitis with severe vaso-occlusion

Cytomegalovirus Retinitis (CMV Retinitis)

Immunosuppressed, though may occur in diabetics after steroid injections. Treat with oral Valganciclovir and intravitreal Foscarnet or Ganciclovir.

CMV Retinitis

Complex RD requiring SO in extensive cases.
Cat-scratch Test for Bartonella hensalae and quintana.

Toxoplasmosis: Any positive blood titer (IgM or IgG) helpful. PCR helpful. Acquired or Congenital (adjacent to a hyperpigmented scar).

Bactrim DS works well. Can densensitize patients. Intravitreal Clindamycin for vision threatening disease.
Retinal Vasculitis

Think collagen vascular disease

Vasculitis-Lupus

Vasculitis-Lupus

Wegener's (ANCA +) Vasculitis
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Behcet’s disease


Behcet’s disease

Classic Masqueraders

Always keep these conditions in the differential of retinitis/vasculitis as they can mimic many different conditions

Syphillis - “The great masquerader”

Sarcoidosis
Sarcoidosis

Tuberculosis

Lymphoma - another “great masquerader”

Unusual Vitreal Cells

51 y.o with progressive floaters OS

Fundus Photos
Fluorescein Angiogram

OCT- OS

Differential Diagnosis of Chronic Large Vitreous Cells
- Chronic fungal endophthalmitis
- Lymphoma
- Sarcoidosis
- Amyloidosis
- Whipple’s disease
- Metastatic cancer cells
- Inflammatory Bowel disease
- Tuberculosis
- Lyme disease
- Idiopathic intermediate uveitis
  - Often require vitreous biopsy to diagnose

Diagnosis: Whipple’s disease

Vitreous biopsy
- Blood/Vitreous PCR positive for *T. whippelli* positive
- Light microscopy:

Whipple’s disease
- Chronic multisystem bacterial disease: weight loss, chronic diarrhea, vague abdominal pain, migratory arthralgias

Pars Planitis
- Lyme
- Tuberculosis
- Syphilis
- Sarcoidosis
- Multiple sclerosis
- Toxocariasis
- Idiopathic

Endogenous Fungal Infection
Vitreous Amyloidosis

Glass wool like or cobweb sheets. Clear with vitrectomy

Metastatic Melanoma


Lymphoma

Sheets of clumped cells in vitreous

Ocular Lymphoma

Ocular Lymphoma
Serous Detachments

Differential Diagnosis:
- Central serous retinopathy – steroids, catecholamines, pregnancy, renal transplant
- CNV
- Optic pit
- Posterior Scleritis
- VKH (Vogt-Koyanagi-Harada Syndrome)
- Rhegmatogenous RD
- Circumscribed Choroidal Hemangioma
- Tumor (amelanotic melanoma, metastasis)
VKH – bilateral uveitis, serous RD's, associated with cutaneous, auditory, and neurologic abnormalities.

Vogt-Koyanagi Syndrome (VKH)

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Need steroids or immunosuppression. Rapid response, but careful to taper very slowly. Can follow EDI Choroidal thickness. Dex implant may help.

Optic Nerve Pit with Schisis

Management – Laser, Vitrectomy/Laser, Retinal fenestration. Takes a long time to see results.

Choroidal Lesions

47 year old healthy Asian-Indian male. Blurry spot OD, 20/25 OU. No cells.
Differential Diagnosis of Multifocal Placoid Chorioretinal Lesions

- AMPPE (Acute Multifocal Posterior Placoid Epitheliopathy)
- Serpiginous Choroidopathy
- Ampiginous or “Relentless” Choroidopathy
- Masquerade Syndromes - Lymphoma, TB, Syphilis
- Multifocal Choroiditis – West Nile Virus, Lyme, Pneumocystis, PIC, Idiopathic
- Histoplasmosis

AMPPE

- Multiple placoid lesions
- Often bilateral
- HLA B7 / DR2
- Recurrences rare
- Vision loss with foveal involvement
- Steroids may speed recovery

Usually resolves within 4-8 weeks, but reported to last up to 6 months

Serpiginous

- Peripapillary: bilateral, progressive. Males more common. Recurrences can be months to years apart. CNVM can develop. Requires immunosuppressive therapy. Test for TB.
Diagnosis: Ampiginous/Relentless placoid choroiditis.
Associated with TB.
Quantiferon gold testing.
Patient quiescent after 9 months of TB rx.

Multifocal Choroiditis

Can be associated with subretinal fibrosis. CNVM can cause vision loss.
Immunosuppressive Rx.

West Nile Virus

Lyme Choroiditis
Pneumocystis Carinii choroiditis

Ophthalmomyiasis

**OPHTHALMOMYIASIS**
- Larval form of flies from the order Diptera
- Botflies: cattle, sheep, horses, deer, rodents, human
- Most common: *Cuterabra* (rodent botfly)
- Usual host: rabbits, squirrels, field mice, rats, chipmunks
- Eggs transported to eye by fly or hands
- Bore into eye may occur in AC, vitreous, SR space

Stay tuned for Part 2