White Spot Syndromes

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Inflammation and dysfunction of:
- Outer retina
- Retinal pigment epithelium
- Choroid
- Vitritis / iritis not a necessary finding

Etiology:
- Unknown
- Autoimmune cause hypothesized for some WSS
- Several of these may be related / spectrum of same process

Common characteristics:
- Blurred vision
- Photopsias
- Visual field changes
- Floaters
- Change in contrast sensitivity
- Symptoms can be present prior to clinical findings
- White spots
  - Can be subtle or prominent
  - One or both eyes
  - Can be asymmetric
White spot syndromes

Differential diagnosis:
- Granulomatous diseases (sarcoidosis, sympathetic ophthalmia, Tuberculosis)
- Masquerade syndromes (syphilis, lymphoma)
- Infectious diseases (toxoplasmosis, POPS, Behcet’s disease)
- Degenerative conditions (drusen)

40 y.o. white female

CC: metamorphopsia os

Vision:
- OD: 20/20  -2.50
- OS: 20/40  -3.75
- IOP 15/18
- Ant Seg: no cells / flare OU
- Vitreous: no cells OU
Work up

- Syphilis serologies: negative
- Bartonella Henselae titers: negative
- Viral titers (HSV, Varicella, CMV): negative
- Toxoplasma titers: negative
- ACE: 24
- ANA: positive 1:160 homogenous pattern
Punctate inner choroidopathy (PIC)

- Healthy myopic women (90%)
- Mean age: 30
- Blurred vision
- Paracentral scotomas
- Small yellow/white lesions of inner choroid / RPE
- Overlying SRF
- Change to atrophic scars
- Usually unilateral (85%)
- No anterior or posterior uveitis

Asymptomatic after one month
Scars become atrophic / pigmented over time
40% develop CNV

Multifocal choroiditis and panuveitis

- Decreased central vision
- Photopsias
- Floaters
- Metamorphopsia
- Paracentral / temporal scotomas
- Photophobia
- Vision can be 20/20 to LP
Multifocal choroiditis and panuveitis

- Mostly white myopic women
- Age 20-60
- Most in their 30s

- Yellow round / oval lesions in outer retina and RPE
- 50-500 um in size
- Posterior pole / midperiphery / PP region
- Often cluster in nasal retina
- Peripheral linear scars parallel to ora
- Can see SRF
- Change to "punched out" scars with healing

- Optic disc edema / atrophy
- Peripapillary subretinal fibrosis ("napkin ring")
- CNV: 25-30%
- Mild / moderate anterior uveitis
- +/- mild or moderate vitritis
Multifocal choroiditis and panuveitis

- Waxes and wanes
- 25% chronic course
- CNV
  - Most common cause of vision loss
- POHS
- Sarcoidosis
- VVK
- Sympathetic ophthalmia
- Infectious / neoplastic
- Other white dot syndromes

MFC / PIC differential diagnosis

- POHS
- Sarcoidosis
- VVK
- Sympathetic ophthalmia
- Infectious / neoplastic
- Other white dot syndromes

MFC / POHS

**MFC**
- Punched out lesions
- PP scarring
- CNV
- Inflammation
- Photopsias, VF defects
- Mixture of acute and chronic lesions
- Growth of lesions

**POHS**
- Punched out lesions
- PP scarring
- CNV
- No inflammation
MFC / PIC

- **MFC**
  - Median age: 45
  - CNV: 28%
  - More inflammation

- **PIC**
  - Median age: 29
  - CNV: 77%

MFC and PIC treatment

- Corticosteroids
- Immunosuppressives
- Anti-VEGF
- PDT

Multiple evanescent white dot syndrome (MEWDS)

- Acute multifocal postviral retinopathy
- Unknown cause
- Young myopic females (75%)
- Multiple white dots
- Level of outer retina or RPE
Multiple evanescent white dot syndrome (MEWDS)

- Acute onset of blurred vision
- Temporal field loss (enlarged blind spot)
- Photopsias
- Flu-like symptoms
- Vision: 20/20 - 20/300
- Usually returns to normal
- Duration: 6 weeks

Numerous small para-macular white spots
- Level of RPE or deep retina
- Spare the fovea
- "Granularity" to macula
- Spots disappear over time
- Left with mild RPE mottling
- Mild iritis
- +/- vitritis

FA:
- Early and late hyperfluorescence of white dots
- "Wreath" like pattern

ICG:
- Numerous hypofluorescent spots
- Usually no treatment required
Birdshot Chorioretinopathy

- Chronic bilateral inflammatory dx
- Healthy men or women: 30-60 yrs old
- Symptoms:
  - Vitreous floaters
  - Decreased vision (especially at night)

- Blurred vision, floaters, photopsias
- Vision usually > =20/40
- No pain or redness
- Severe nyctalopia
- Changes in color or visual fields
- Purely ocular disease
- Mean age: 53
- Predominately white patients
- Bilateral disease

- Multiple small cream-colored lesions
- Scattered around the optic nerve towards the equator
- "shotgun pattern"
Birdshot Chorioretinopathy clinical findings

- Birdshot lesions:
  - Round/oval
  - ½ to ¼ DD in size
  - Appear deep retina
  - RPE and overlying retina intact
  - Tend to cluster near the nerve
  - Minimal A/C and vitreous reaction
  - No posterior synechiae
  - Posterior inflammation signs: disc edema, vasculitis, CME

Birdshot Chorioretinopathy

- Strongly associated with HLA-A29 allele
- 90% of patients with BCR are HLA-A29 positive
- Abnormal ERG: 88%
- Visual field defects

Birdshot Chorioretinopathy DDx

- Pars planitis
- Intracocular B-cell lymphoma
- Syphilitic chorioretinitis
- Sarcoidosis
- Sympathetic ophthalmia
- Other white dot syndromes (MFC with panuveitis)
Birdshot Chorioretinopathy Treatment

- Corticosteroids
- Immunosuppressive therapy
- Anti-VEGF if there is CNVM
- Yearly ERG, visual field

Placoid Diseases

- Acute posterior multifocal placoid pigment epitheliopathy (APMPE)
- Serpiginous choroiditis
- Relentless placoid chorioretinitis
- Persistent placoid maculopathy
  - "placoid lesions"

Acute posterior multifocal placoid pigment epitheliopathy (APMPE)

- Rapid onset of central vision loss
  - Can see central/paracentral scotomas
  - Photopsia
  - Metamorphopsia
- Unilateral or bilateral (more common)
  - If unilateral, fellow eye can become involved in a few weeks
  - Headache, stiff neck, malaise
  - Hx of antecedent viral syndrome
  - Young adults
APMPPE fundus findings

- multiple round cream colored flat lesions
- Indistinct margins
- Lesions not found anterior to equator
- Usually bilateral
- Fresh lesions can develop
- Lesions of differing ages can be seen
- Lesions cleared gradually leaving
  - Hypopigmentation
  - Pigment clumping

APMPPE

- Localized serous RD over the lesions
- Vitritis not common
- Sometimes can see AC reaction

APMPPE: clinical course

- Improves over 2-4 weeks
- Good prognosis
- Foveal involvement good predictor of final vision
APMPPE: imaging

- FA: early hypofluorescence, late hyperfluorescence
- ICG: shows more lesions than clinical exam

APMPPE: systemic associations

- CNS:
  - Cerebral vasculitis
  - Meningo-encephalitis
  - Stroke
  - CN VI palsy
  - Transient hearing loss
  - Headache
  - Systemic vasculitis

APMPPE: etiology unclear

- Etiology: unclear
  - HLA-B7 and HLA DR2 association
  - Possible viral cause
  - Found post various vaccinations
- Differential Dx:
  - Other white dot syndromes
  - Herpes zoster
  - TB, sarcoid, syphilis
  - Choroidal metastasis, lymphoma --
APMPPE: treatment

- Observation
- Corticosteroids / immunosuppressives
  - IFCH involvement

Serpiginous choroiditis

- Bilateral progressive chronic inflammatory chorioretinitis
- Usually one eye active at a time
- Ages 30-70
- Unknown cause

Serpiginous choroiditis: clinical findings

- Classic appearance (80%)
  - Geographic patches of gray / creamy yellow placoid lesions
  - Peripapillary
  - Progresses in centrifugal manner
  - Fingerlike / serpentine projections
Serpiginous choroiditis: clinical findings

- Edematous outer retina
- Serous RD can be seen
- Active lesions heal
- Extensive RPE/choriocapillaris atrophy
- Recurrences can occur at edge of old scars
- Symptomatic when fovea becomes involved
- Macular serpiginous choroiditis

- White eye
- 1/3 have fine vitreous cells
- A/C reaction not common
- Multiple recurrences over months/years
- 75% of patients develop central vision loss
- Final vision <20/200 in 25% regardless of treatment

Serpiginous choroiditis: differential dx.

- APMPPE
  - Younger pts
  - Lesion scattered throughout posterior pole
  - Recurrences rare
- Other white dot syndromes
- Tuberculous serpiginous choroiditis
- Sarcoidosis, syphilis, toxoplasmoids, posterior scleritis etc.
Serpiginous choroiditis: treatment
- Corticosteroids
- Immunosuppressives

Acute zonal occult outer retinopathy (AZOOR)
- Idiopathic inflammatory disorder
- Young healthy women
- Photopsia and acute progressive VF loss
  - VF starts as enlargement of blind spot
- Due to damage to broad zones of outer retina
- Unilateral initially
- Can progress to bilaterality

Fundus initially appears normal
- Retinal atrophy and mottling
- Attenuated arterioles
- Often peripapillary involvement
- May resemble:
  - Sectoral retinitis pigmentosa
  - Diffuse unilateral subacute neuroretinitis (DUSN)
- No a/c rxn; minimal vitreous cells
Acute zonal occult outer retinopathy (AZOOR):

- Usually active for 6 months then stabilize
- Can be chronic progressive disease
- Fundus autofluorescence:
  - Involved areas show hypoautofluorescence

Differential dx:
- Hereditary retinal diseases (RP) -
- Retinal dystrophies
- Cancer associated retinopathy

Treatment:
- No proven therapy

Acute macular neuroretinopathy

- Young middle aged women
- Acute onset of mild decreased vision in one or both eyes
- One/multiple paracentral scotomas
- Wedge shaped intraretinal lesions:
  - Point to fovea
  - Flower petal arrangement
  - Outer retina
  - Spare RPE and retinal vessels
Acute macular neuroretinopathy

- Self-limited
- Scotomas can persist
- No treatment proven effective