Pachychoroid Clinical Spectrum

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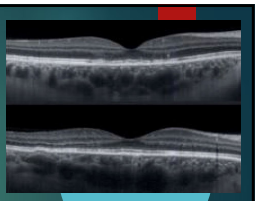
Pachychoroid disease



- Warrow et al 2013:
  - Described
    - pigmentary changes similar to fellow eyes of CSCR
    - Choroidal thickening
  - Called this: **pachychoroid pigment epitheliopathy**
    - **"Pachy" : thick**
  - Subsequent reports: other diseases manifest similar choroidal findings

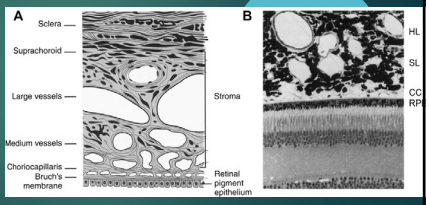
New group of macular diseases: **Pachychoroid Clinical Spectrum**

pachychoroid clinical spectrum

- Pachychoroid Pigment Epitheliopathy
- Central Serous Chorioretinopathy
- Pachychoroid Neovasculopathy
- Polypoidal Choroidal Vasculopathy

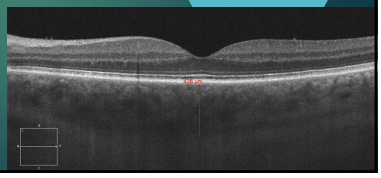
Anatomy of the Choroid

- Haller's layer
  - Large choroidal veins
- Sattler's layer
  - Medium sized choroidal veins
- Choriocapillaris
  - Fenestrated vessels
  - Supply outer retina
- Bruch's membrane
  - Adjacent to RPE



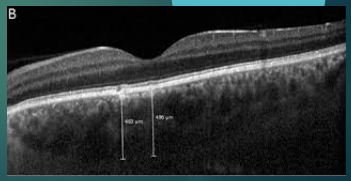
Choroidal thickness

- Varies with age, ethnicity, axial length
- Normal: 191-354 µm
- Thick choroid: >390 µm
- Enhanced depth imaging (EDI) OCT



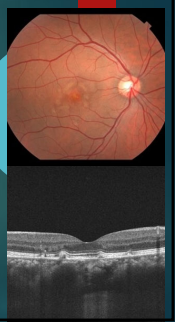
pachychoroid clinical spectrum

- Increased choroidal thickness
- "Pachyvessels"
- Dilated vessels in the outer choroid
- Compress inner choriocapillaris



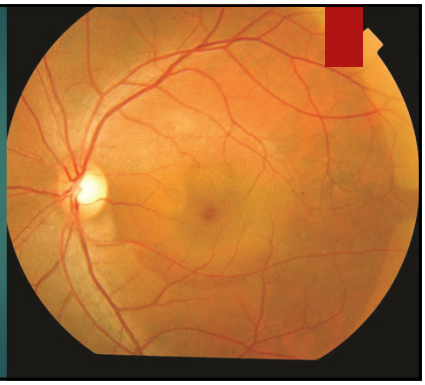
Pachychoroid Pigment Epitheliopathy

- ?forme fruste / precursor of CSCR
- RPE changes similar to CSCR
- No subretinal fluid
- Asymptomatic
- Thickened choroid
  - Appears reddish orange fundus
  - Reduced fundus tessellation
  - RPE abnormalities / small PED



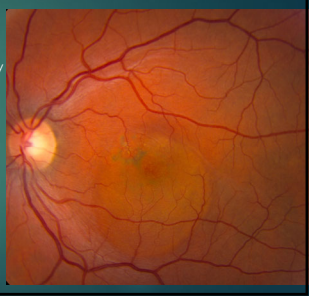
Central serous chorioretinopathy

- First describes by Albrecht von Gaefe 1866
- Serous retinal detachment with or without PED
- Most commonly seen in macular region
- Part of the spectrum of pachychoroid disease



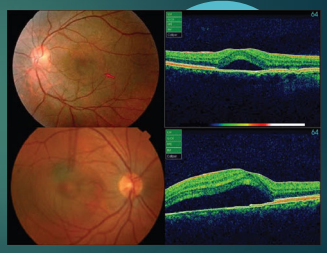
Epidemiology

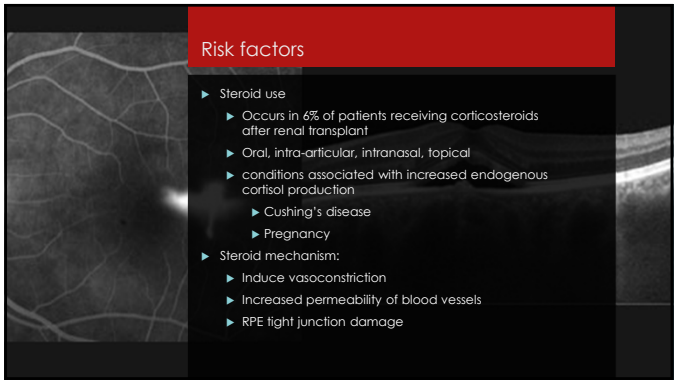
- Age of onset: 30-50
  - Can continue as chronic disease in elderly
- Older patients:
  - Worse vision
  - Diffuse retinal pigment epitheliopathy
  - Increased rates of CNVM
- Annual incidene:
  - 9.9 / 100,00 men
  - 1.7 / 100,00 women
- Bilateral : 14-40%



Risk factors

- Type A behavior
  - Elevation of catecholamine levels
- Hypertension
- Helicobacter pylori infection
- Obstructive sleep apnea
- Autoimmune disease
- Familial predisposition



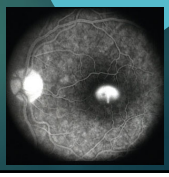


Risk factors

- Steroid use
  - Occurs in 6% of patients receiving corticosteroids after renal transplant
  - Oral, intra-articular, intranasal, topical
  - conditions associated with increased endogenous cortisol production
    - Cushing's disease
    - Pregnancy
- Steroid mechanism:
  - Induce vasoconstriction
  - Increased permeability of blood vessels
  - RPE tight junction damage

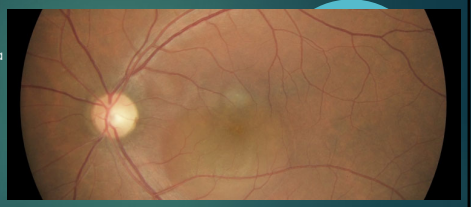
pathophysiology

- Multiple etiologies and mechanisms
- Widespread choroidal circulation abnormality
  - Choroidal vascular hyperpermeability
  - Increased hydrostatic pressure in choroid leads to breakdown of RPE barrier
  - Leakage of fluid into subretinal space



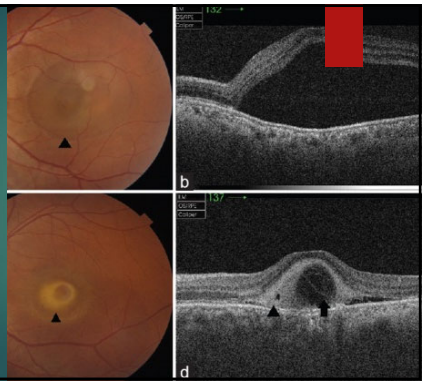
Clinical features: symptoms

- Central scotoma
- Metamorphopsia
- Micropsia
- Blurred vision
- Hyperopic shift



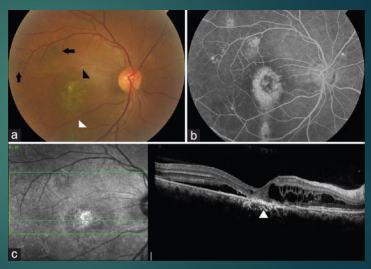
Clinical feature: acute

- Serous macular detachment
- Yellow subretinal deposits in atypical cases
- RPE defects
- Usually resolves in 3-4 months



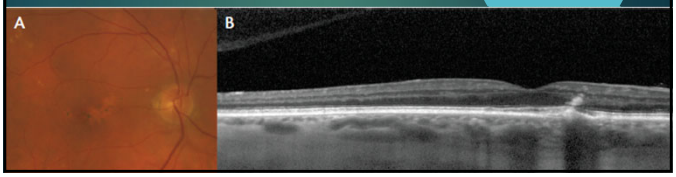
Clinical features: Chronic

- RPE degeneration
- Shallow SRF
- Cystoid edema / schisis
- RPE degeneration in a teardrop configuration due to gravity
- CNVM
- Foveal atrophy



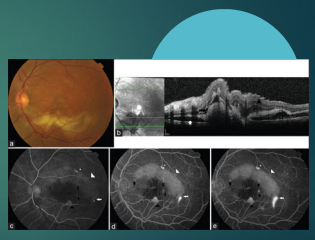
Clinical features

- Tesselation: visibility of large choroidal vessels
- Reduced fundus tessellation
- Due to changes in choroidal pigmentation or to choroidal thickening



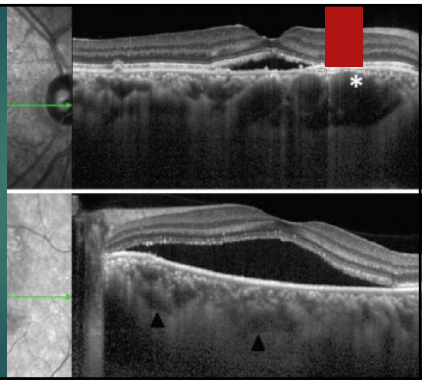
Atypical CSCR

- Inferior exudative RD
- Multifocal PED
- Large RPE tears
  - Usually in patients undergoing systemic corticosteroid therapy



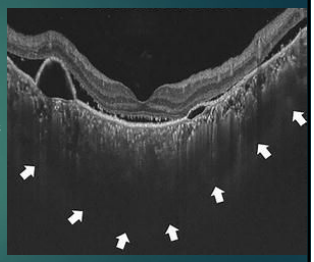
Optical coherence tomography

- Choroid
  - Increased choroidal thickness
    - Increased thickness in fellow eye
    - Not a mandatory criterion
  - Enhanced depth imaging



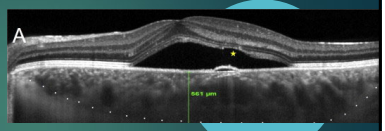
OCT: choroid

- Thinning of inner choroidal layer
- Outer choroidal vessel dilation
- Hyperreflective dots inner choroid
- Hyperreflectivity of wall of dilated vessels



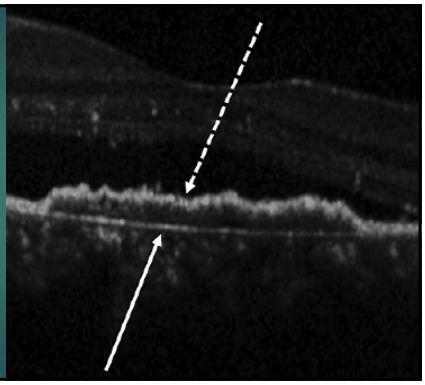
OCT: retinal pigment epithelium

- PED
  - 50%
  - Can be within or outside SRF
  - RPE atrophy
  - "double layer sign"
    - Undulated RPE layer with hyporeflective content over intact Bruch's membrane
    - Sign of chronicity



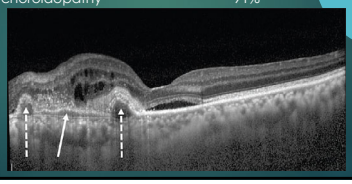
Double layer sign

- Separation of the irregularly elevated RPE from the inner layer of Bruch's membrane
- Fluid between basement membrane of RPE and Bruchs's membrane



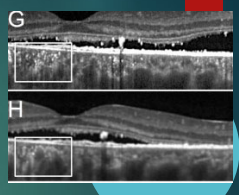
Double layer sign

- Pachychoroid pigment epitheliopathy 0%
- Pachychoroid neovasculopathy 93%
- Chronic central serous chorioretinopathy 31%
- Polypoidal choroidopathy 91%



OCT: subretinal space

- Acute CSCR:
  - SRF is serous
  - Hyporeflective area between retina and RPE
- Chronic CSCR:
  - Hyperreflective dots
    - May reflect photoreceptor outer segments shedding, fibrin or lipids
  - Atypical CSCR: clump or band like deposits

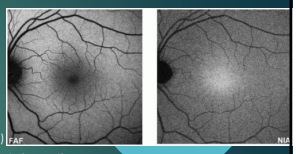


OCT: neurosensory retina

- Acute: retina intact
- Chronic:
  - changes in photoreceptor outer segments
  - Intraretinal hyperreflective dots
  - Intraretinal cysts or schisis
  - Gradual loss of outer segments resulting in poor visual prognosis

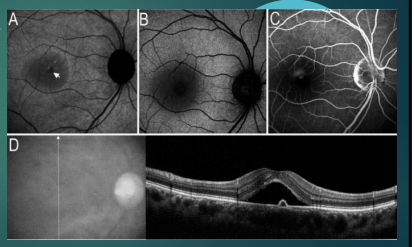
Fundus autofluorescence (FAF)

- Noninvasive imaging modality
- Two types :
  - Short wavelength FAF
    - Originates from lipofuscin pigment of RPE
    - Optic nerve and vessels hypoAF (no lipofuscin)
    - Fovea hypoAF (absorption of blue light by luteal pigment)
  - Infrared FAF
    - Originates from melanin pigment of the choroid and RPE
      - Optic nerve and vessels hypoAF (lack of melanin)
      - Fovea: hyerAF (more dense melanin)



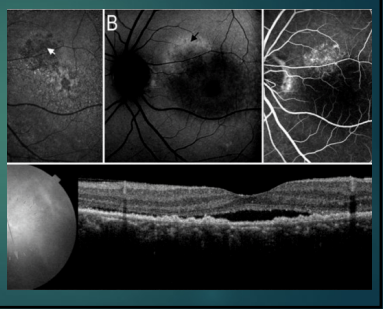
Fundus autofluorescence Acute CSCR

- Hypoautofluorescence at leaking point
- Hypoautofluorescence in areas of SRF
  - Due to masking of RPE autofluorescence by SRF



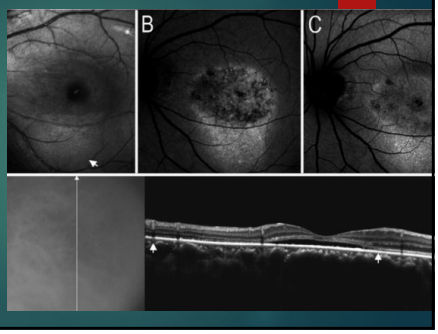
Fundus autofluorescence Chronic CSCR

- Hypoautofluorescence at leaking point
- Hyperautofluorescence
  - Outer retinal atrophy / loss of photoreceptors
  - Unmasking of normal RPE autofluorescence



Fundus autofluorescence: long term sequelae

- Mixed FAF patters
- Later Hypoautofluorescence
  - In areas of RPE damage in pathway of SRF
- Granular Hyperautofluorescence

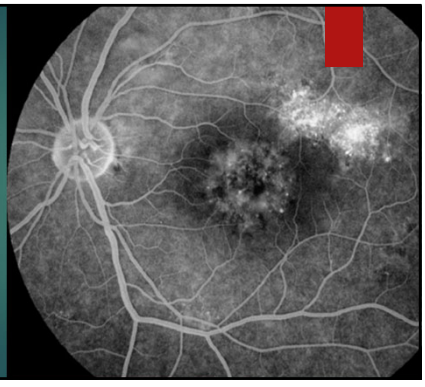


Fundus Fluorescein Angiography: Acute CSCR

- Single or mulifocal point leaks
- Early punctate hyperfluorescence
- Late:
  - "ink blot"
  - "smoke stack" leakage

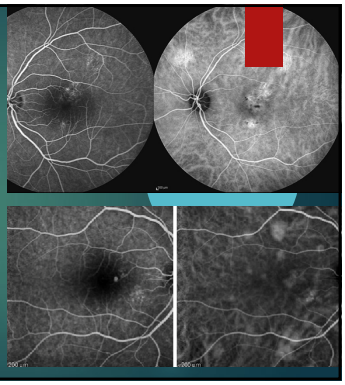
Fundus Fluorescein Angiography Chronic CSCR

- Multifocal leakage
- Diffuse oozing of dye
  - Diffuse RPE defect



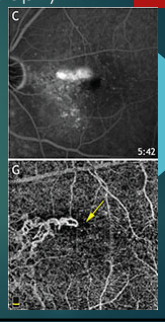
Indocyanine Green Angiography

- Choroidal filling delay
- Dilated choroidal vessels
- choroidal hyperfluorescence
- focal patchy areas of choroidal hyperfluorescence
- Choroidal vasculature abnormality more widespread than FA
  - Commonly present in fellow eye



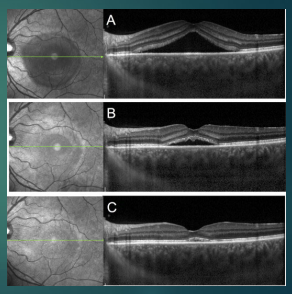
Optical Coherence Tomography Angiography

- New noninvasive technique
- Detects changes in blood flow in vessels
- No need for dye injection
- Segmentation of different vascular layers
- CSCR:
  - Dilated choriocapillaris
  - Abnormal flow patterns suggestive of choroidal ischemia
  - Can detect CNVM



CSCR: natural history

- 90% improve spontaneously
  - Residual metamorphopsia, scotoma
  - Reduced contrast sensitivity
- Chronic CSCR:
  - Poor vision on presentation
  - Prolonged duration of SRF
- 50% recurrence rate
- Hx of psychiatric disease
  - Higher recurrence rate

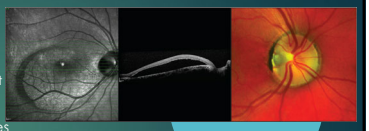


CSCR: natural history

- Irreversible vision loss:
  - RPE atrophy
  - CNVM (6%)
  - Transformation into polypoidal choroidopathy

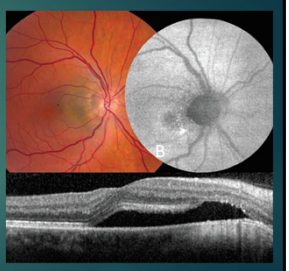
CSCR differential diagnosis

- Optic disc pit
  - 45% serous macular detachment
- ARMD
- Inflammatory / infectious diseases
  - Vogt-Koyanagi-Harada (VKH) disease
  - Posterior scleritis
  - Sympathetic ophthalmia
  - Uveal effusion syndrome
  - Benign reactive lymphoid hyperplasia



CSCR differential diagnosis

- Autoimmune / vascular disorders
  - SLE
  - Polyarteritis nodosa
  - Malignant HTN /
  - Toxemia of pregnancy
  - Disseminated intravascular coagulopathy
- Intraocular tumors
  - Choroidal hemangioma
  - Choroidal melanoma
  - Choroidal metastasis



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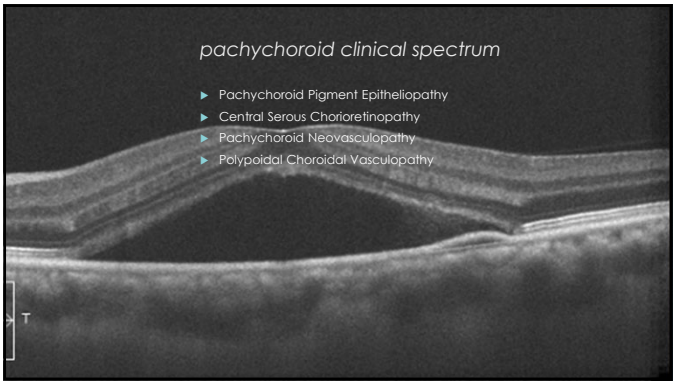
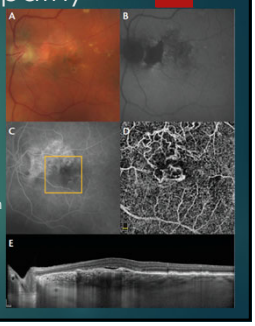
- Polypoidal Choroidal Vasculopathy



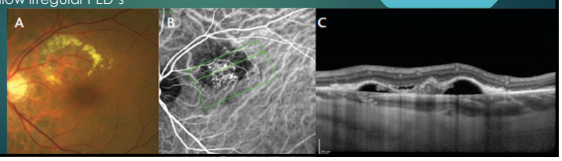
Pachychoroid Neovasculopathy

- Form of type 1 (sub-RPE) CNVM
- Differences from AMD:
  - Younger age
  - Relative absence of drusen
  - Thick choroid
  - Pachyvessels
- Elderly patients can exhibit characteristics of both

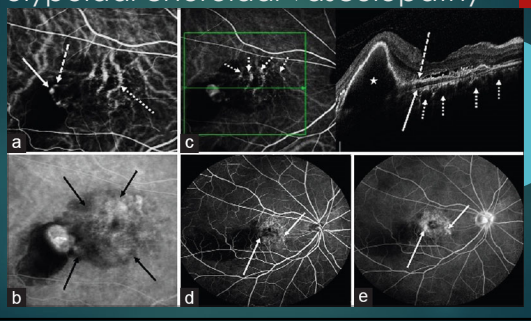


POLYPOIDAL CHOROIDAL VASCULOPATHY

- Traditionally considered a variant of wet AMD
- Minimal / absent drusen
- Thicker choroid
- Pachyvessels with inner choroid attenuation
- Shallow irregular PED's



Polypoidal choroidal vasculopathy

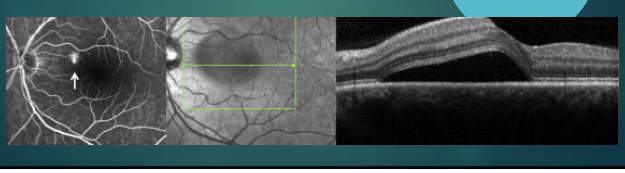


Treatment: CSCR

- Observation
  - Stop corticosteroids
    - PO, topical, nasal sprays, injections
  - Lifestyle modifications
- Photodynamic therapy
  - Choriocapillaris narrowing
  - Reduction of choroidal exudation
  - Choroidal hypoperfusion
  - Choroidal vascular remodeling

Treatment: CSCR

- Argon laser photocoagulation
- Micropulsed diode laser
- Anti-VEGF therapy

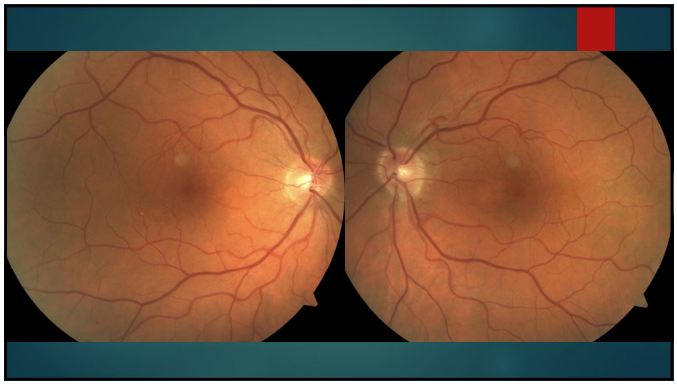


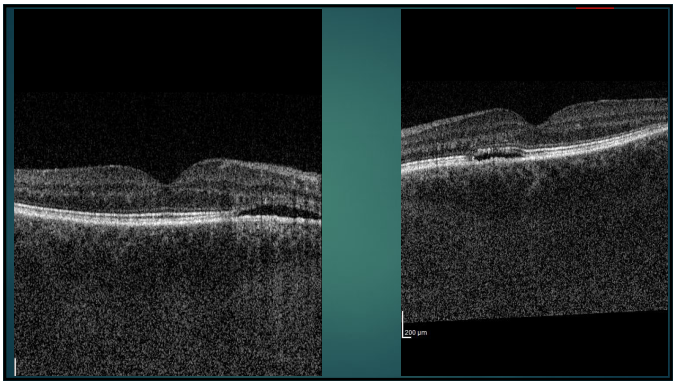
CSCR: corticosteroids

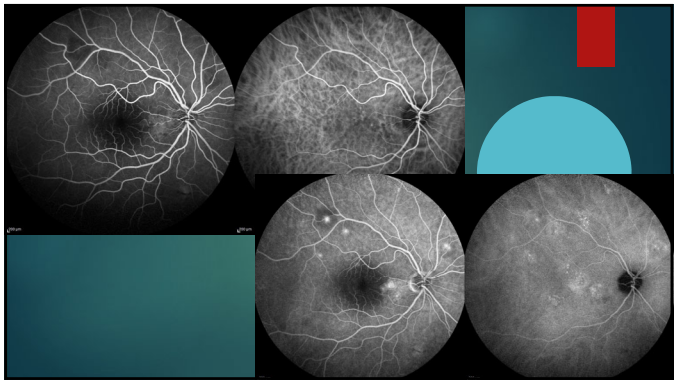
- Elevated glucocorticoid hormone levels
  - Caused by systemic / endogenous steroids
  - Bind to glucocorticoid receptor & mineralcorticoid receptor
  - MR present in RPE and endothelium of retinal / choroidal vessels
  - Activation of MRs in choroidal vessels
    - Vessel dilation
    - Leakage
    - Choroidal hyperpermeability
  - Choroidal hyperpermeability can be blocked by MR antagonists

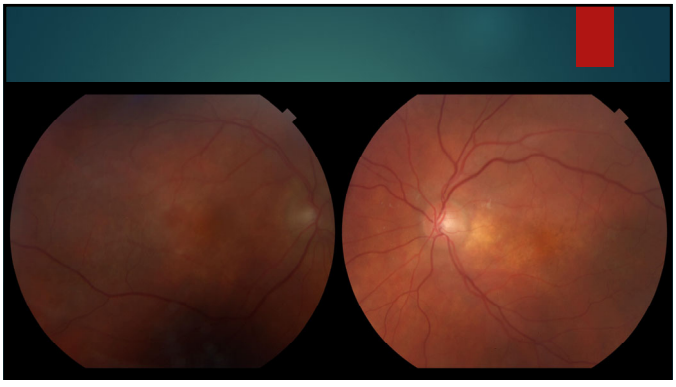
Treatment: CSCR

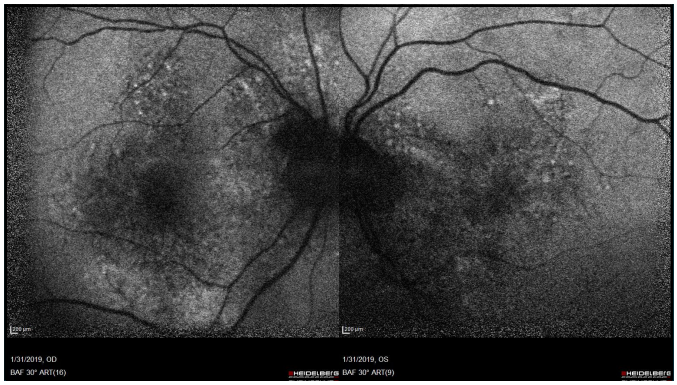
- Anticorticosteroid treatment
  - Mineralocorticoid receptor antagonists
    - Spironolactone (50 mg / day)
    - Eplerone (50 mg / day)
- Other
  - Rifampin










1/31/2019, OD
BAF 30° ART(16)
1/31/2019, OS
BAF 30° ART(9)