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CHOROIDAL IMAGING IN UVEITIS: AN UPDATE

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Aprile 2022

ARTICLE IN PRESS

JID: SOP [mNS;January 28, 2022;22:48]

SURVEY OF OPHTHALMOLOGY XXX (XXXX) XXX

Available online at www.sciencedirect.com

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Survey of Ophthalmology

Review article

Choroidal imaging in uveitis: An update

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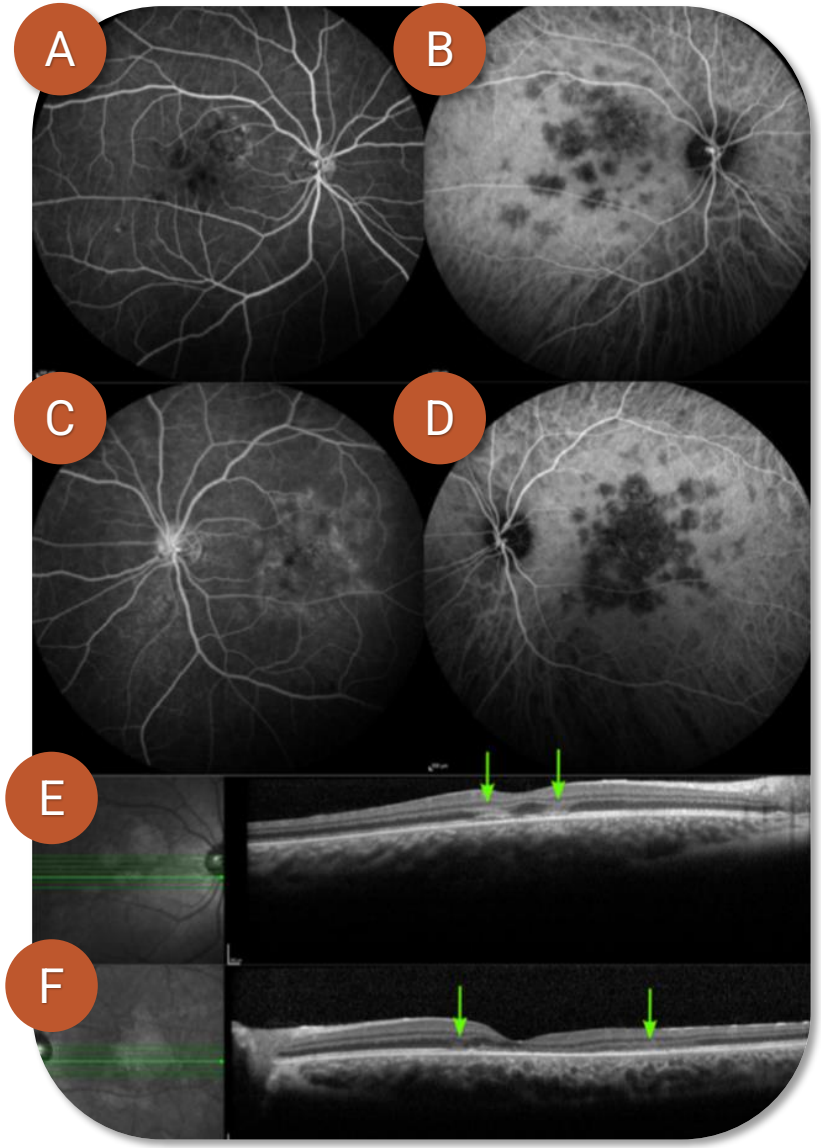
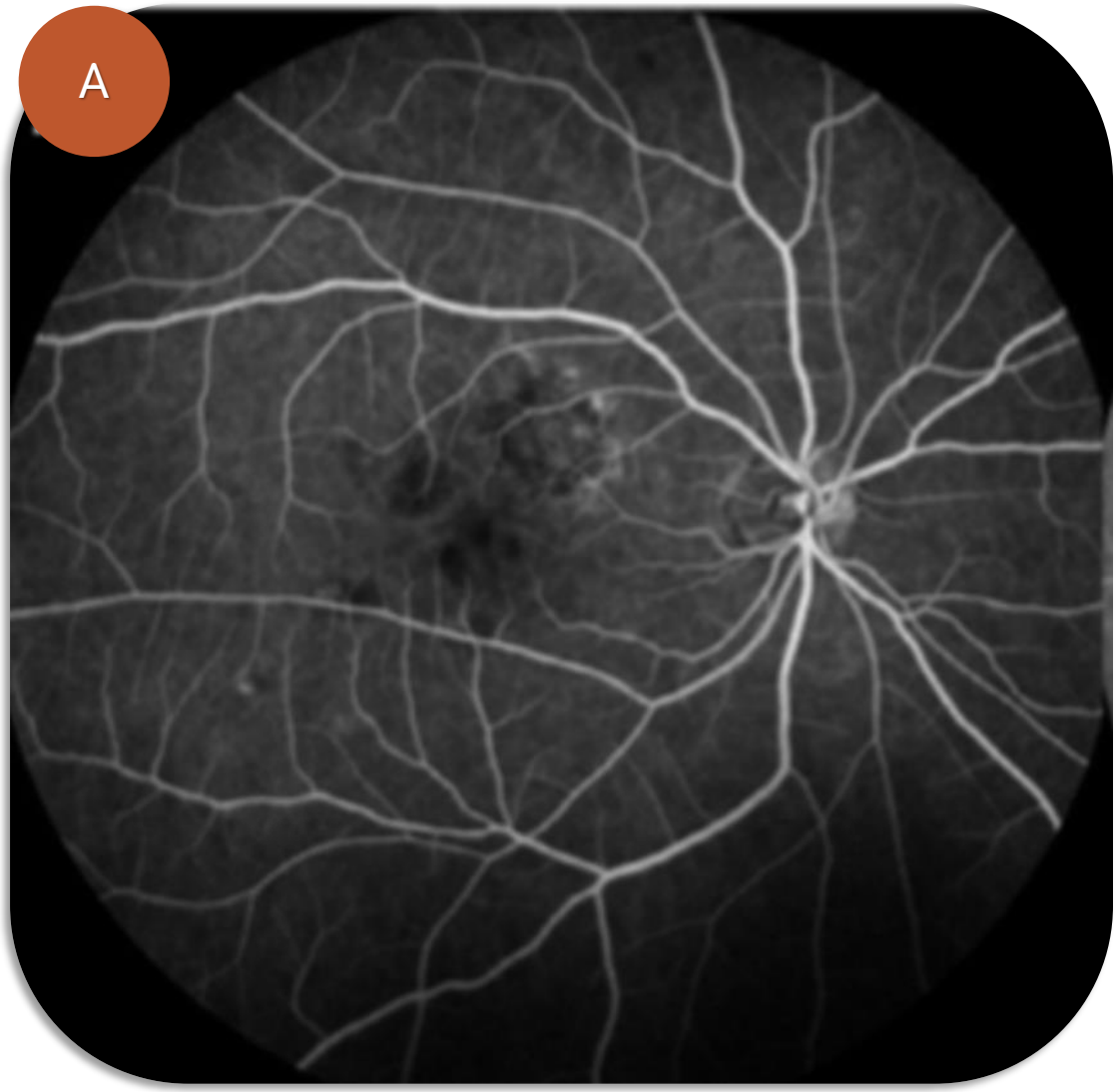
Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

- Inflammatory disease
- Sudden deterioraton in vision
- Young individuals
- Multiple, discrete, yellowish white, flat lesions, mostly in the posterior pole
- Gass originally described and named the disease (pigment epithelium tissue most affected)
- Acute inflammation of the choriocapillaris with secondary involvement of the RPE





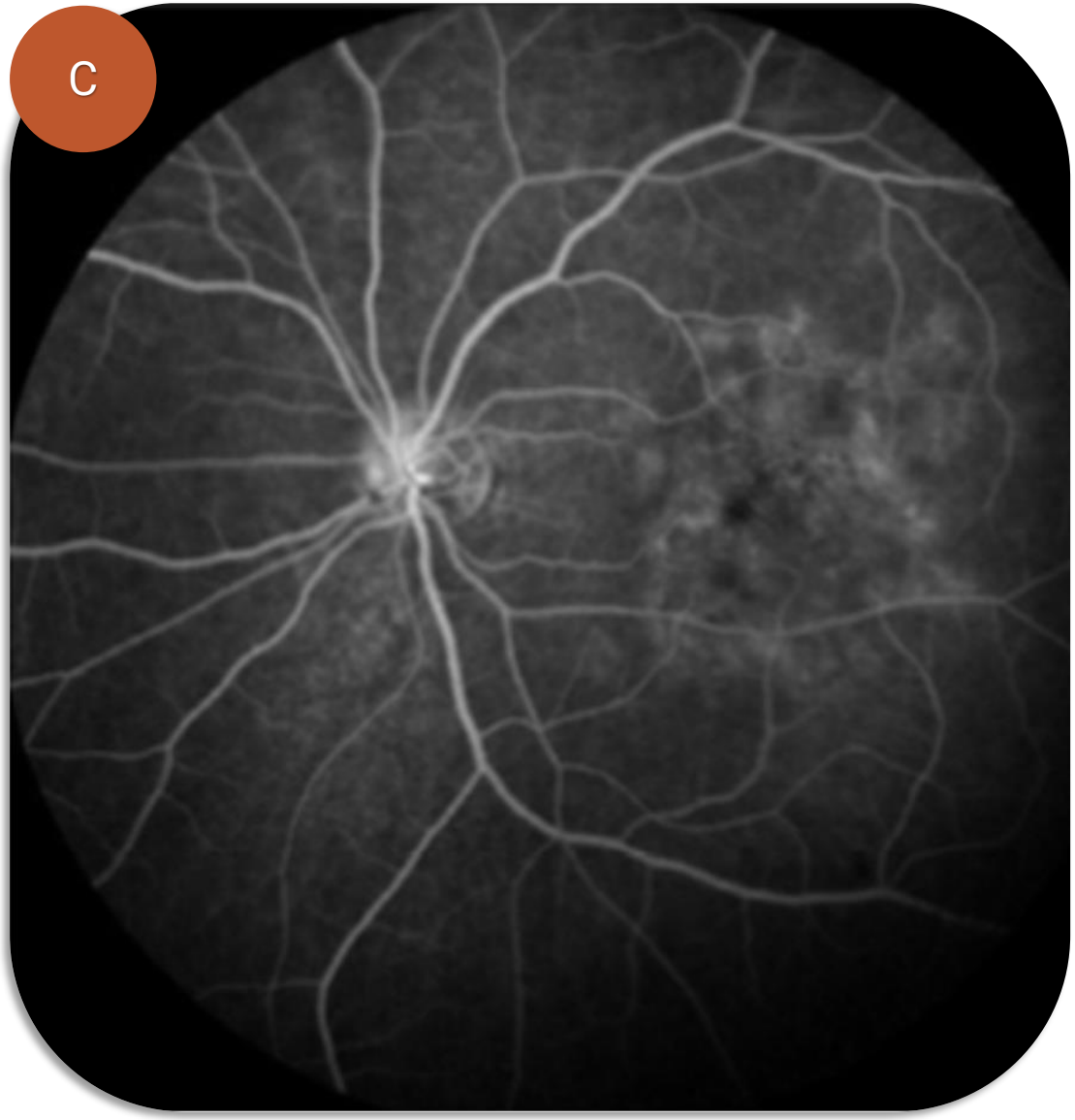
APMPPE: FA

- ACTIVE LESIONS: early hypofluorescence and late staining
- Early hypofluorescence**: masking effect** from acute alteration of the RPE (swelling of RPE or infiltrates)
- Others interpreted this finding **as delayed choriocapillaris filling** (occlusion of precapillary arterioles that feed the lobules of choriocapillaris or inflammatory infiltrates leading to compression of choriocapillaris)

- **Late staining**: not all the hypofluorescent lesions seen in the early phase FA

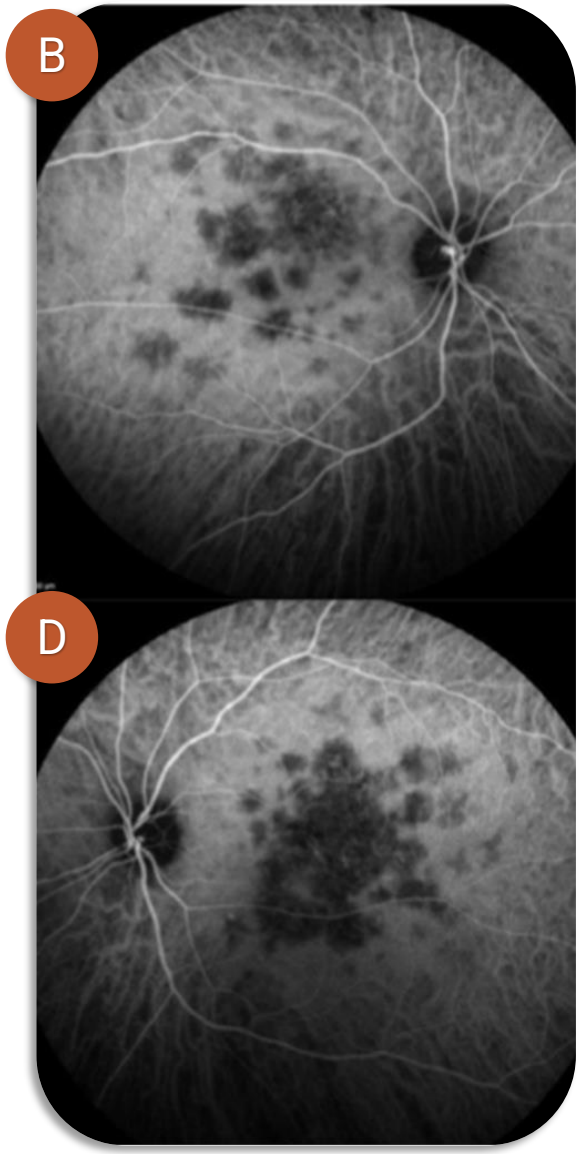
- **Diffusion of dye from the choroid to the demaged RPE cells or staining of cellular infiltrates located in the choriocapillaris and RPE**

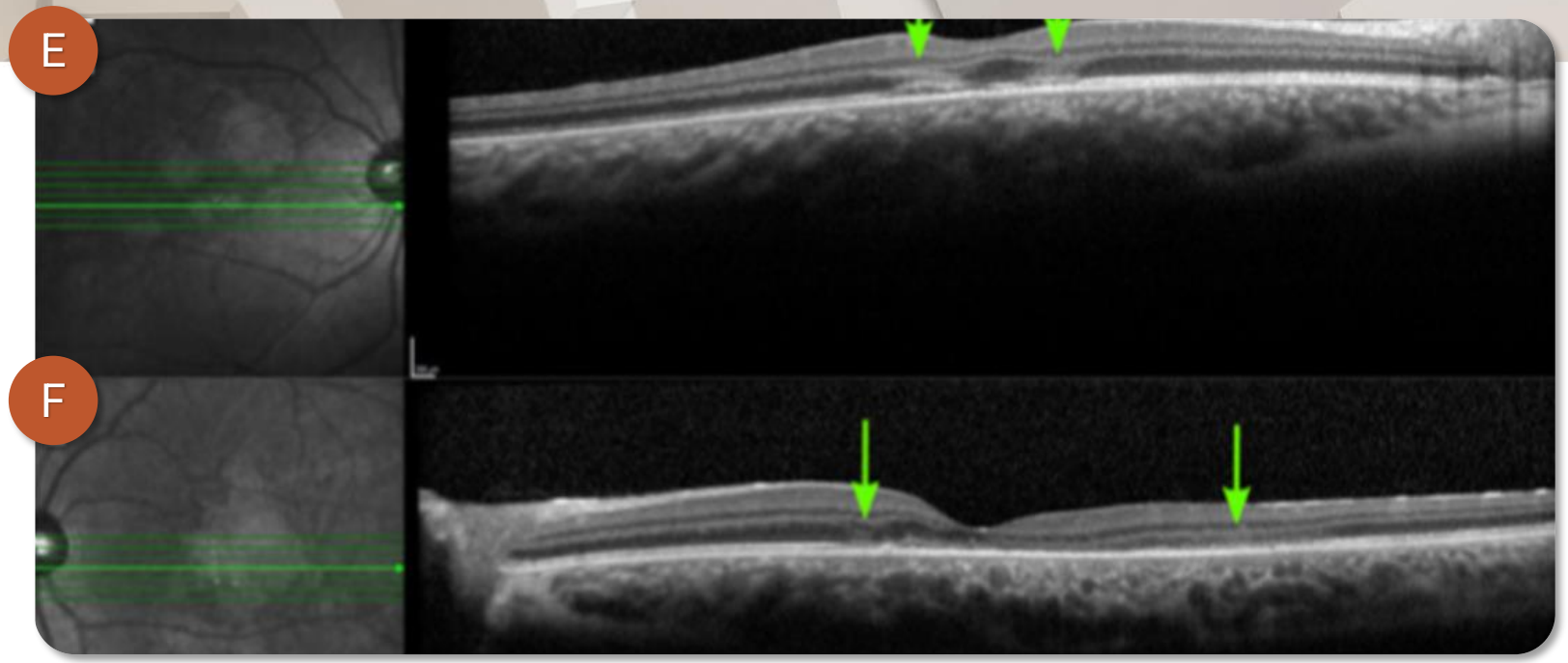
- Since **FAF** lesions are less numerous than the early phase hypofluorescent lesions seen an FA → RPE involvement appears to be secondary to choroidal involvement



APMPPE: ICGA

- **Delayed choroidal filling** and multiple areas of **early hypocyanescence** which remain hypocyanescent in the late phase
- Areas of hypocyanescence **correspond to hypofluorescence on FA** and are **more numerous than the placoid lesions** seen clinically
- Visualization of underlying large choroidal vessels in areas of hypocyanescence in the early phase, persistence of hypocyanescent lesion in the late phase → **PRIMARY CHORIOCAPILLARIS PERFUSION DEFECT WITH DELAYED AND SECONDARY INVOLVEMENT OF RPE**



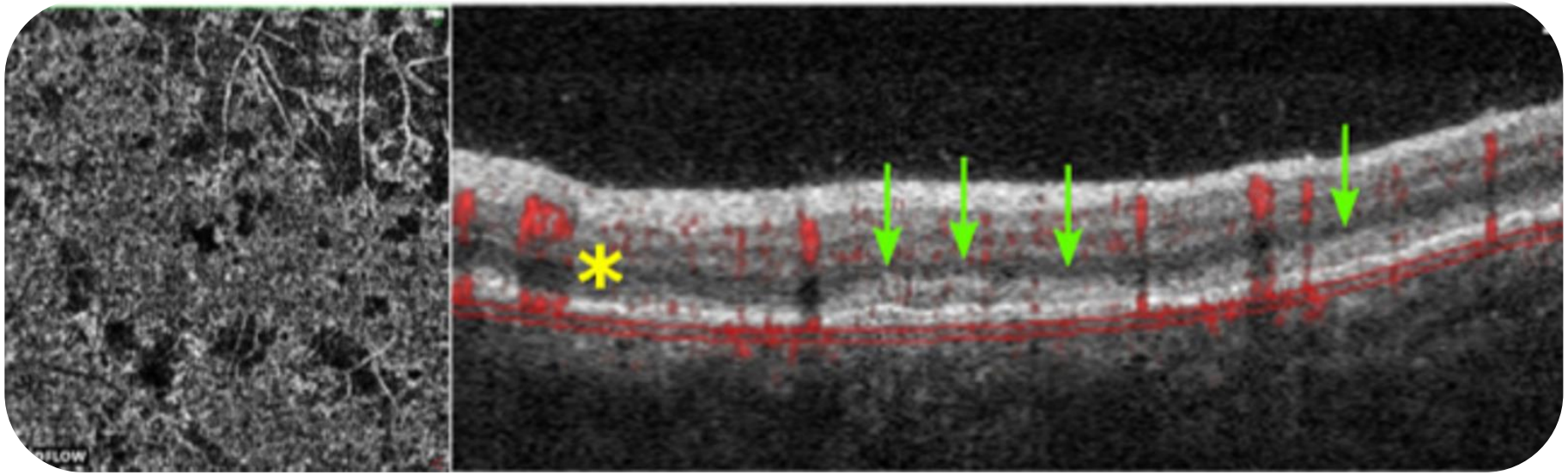


APMPPE: OCT

- **EDI-OCT**: **distinct involvement of the RPE and choroid**
- **Outer retinal opacification and ellipsoid zone disruption**
- **Increased thickness of the choroid and inner choroidal lucency**
- **Lucency**: **compression or obliteration of the small choriocapillaris** consistent with the hypofluorescence in the early phase FA
- **HEALED APMPPE**: **normalization of choriocapillaris reflectivity and choroidal thickness.**
- **Choroidal thinning** in case of healing with atrophy

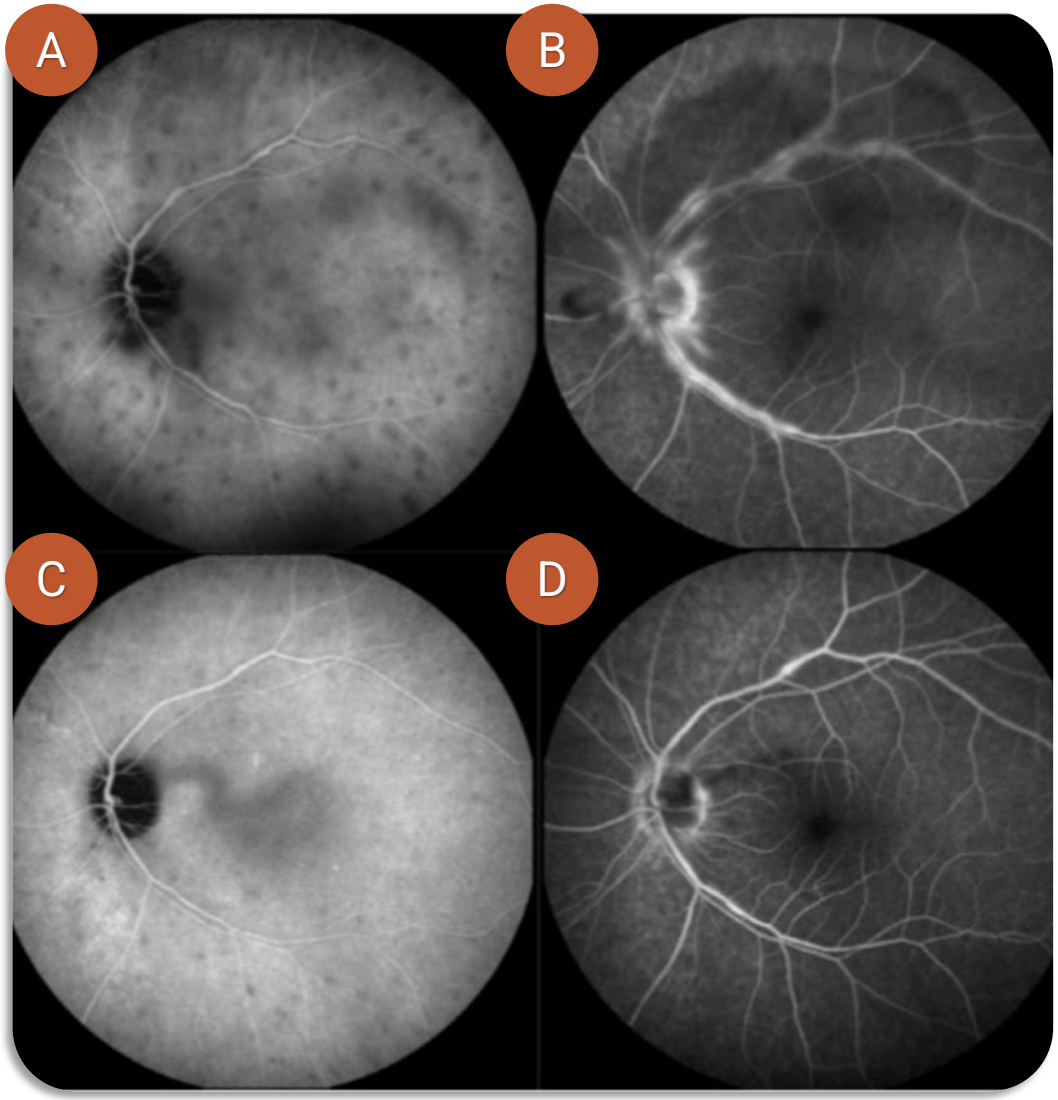
APMPPE: OCGA

- **ACUTE STAGE:** multifocal areas of choriocapillaris flow void
- **Primary inner choroidal insult**
- **OCTA: more sensitive** than FA and ICGA in detecting active lesions
- **HEALED STAGE**: Prompt steroid therapy➔ reperfusion of areas of apparent ischemia
- Transient choriocapillaris ischemia➔ **RPE irregularities are not severe and photoreceptor damage is reversible** ➔ persistent outer retinal thinning and persistent hypoautofluorescence on FAF



Birdshot chorioretinopathy (BCR)

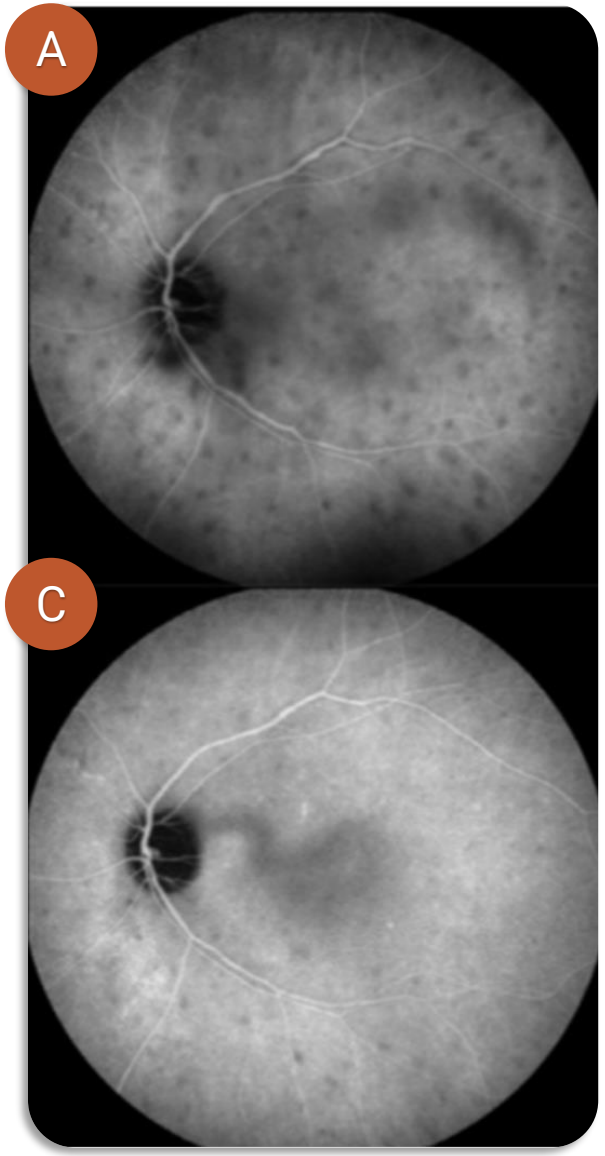
- Autoimmune
- Bilateral posterior uveitis
- No apparent systemic involvement
- Mostly in Caucasians
- Low-grade anterior segment inflammation
- Vitritis
- Retinal vasculitis (large retinal veins)
- **Rice-shaped hypopigmented choroidal lesions**
- **HLA-A29** in almost 100% of patients
- **DUAL INDIPENDENT INFLAMMATION IN THE CHOROID AND IN THE RETINA → CHOROID PRIMARY SITE**



BCR: ICGA

- **ICGA findings precede fundus lesions**
  1. **Bilaterally symmetrical, round oval, hypocyanescent spots in the early-intermediate phase** (GRANULOMATOUS INFILTRATION)➜isocyanescent or hypocyanescent in the late phase ➜more numerous than seen on FA
  2. **Late diffuse hypercyanescence**
  3. **Fuzzy and indistict appearance of the choroidal vessels in the intermediate phase** ➜ choroidal vasculitis with leakage

- **HEALED**: **isocyanescent spots** or persistent **hypocyanescence in case of scarring**

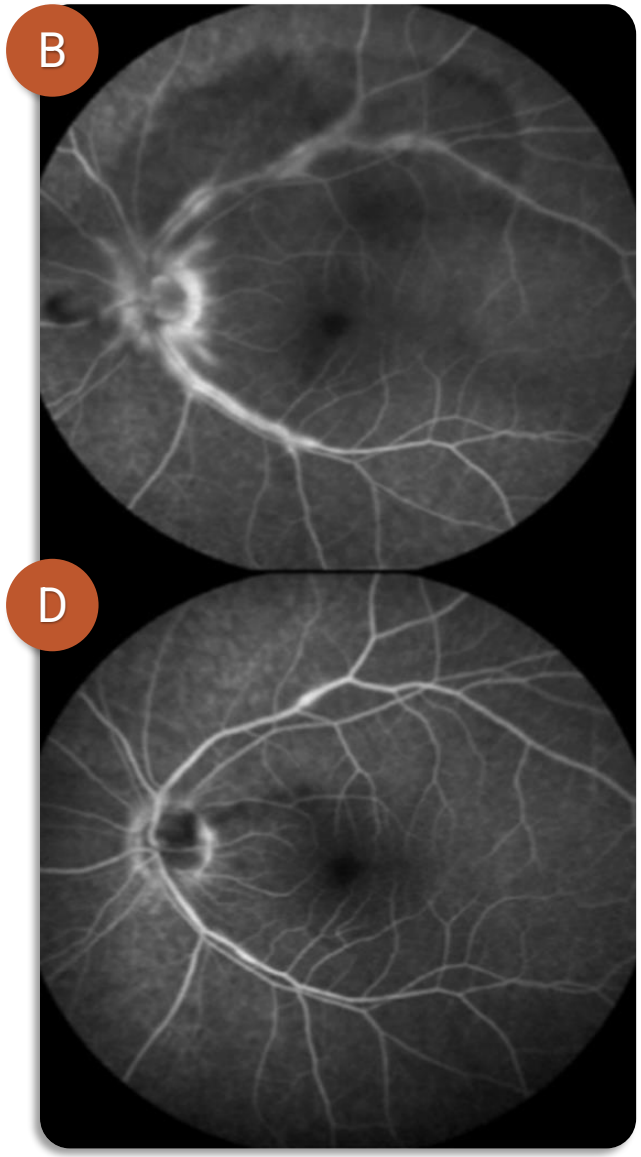


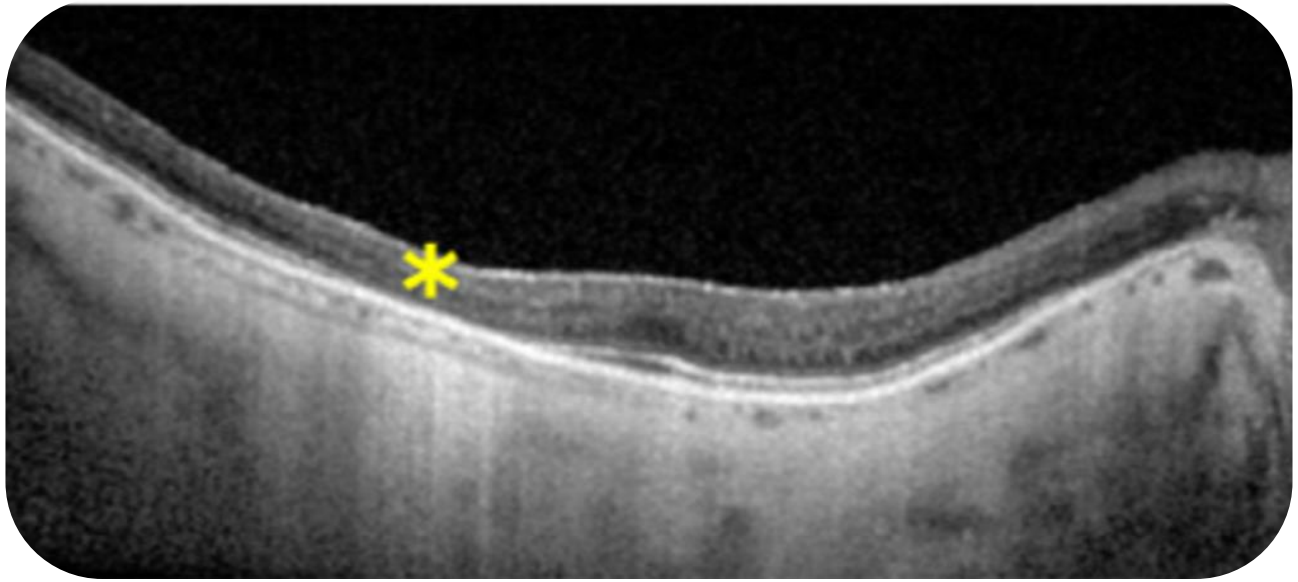
BCR: FA

- ICGA findings do not correspond with those seen on FA
- TWO DUAL INDEPENDENT INFLAMMATORY PROCESSES IN THE RETINA AND IN THE CHOROID

- ACTIVE:
  1. Disc hyperfluorescence
  2. Cream coloured fundus lesions silent/ hypofluorescent
  3. Posterior pole leak with cystoid edema
  4. Hyperfluorescent foci (mottled appearance)

- HEALED:
  1. early hypofluorescence
  2. late hyperfluorescence of atrophic areas



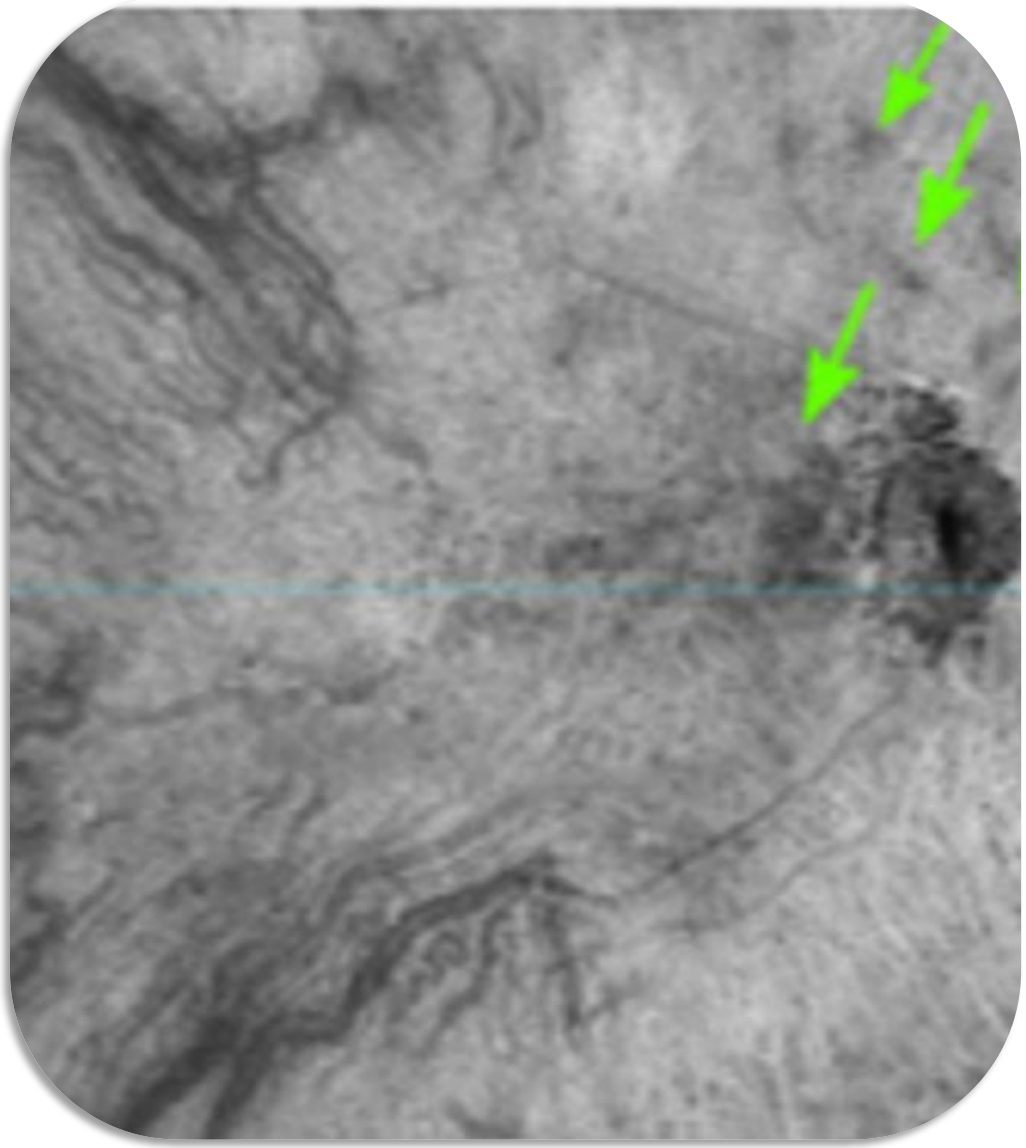


BCR: OCT

- Progressive choroidal thinning
- Thinning and atrophy mainly involve the Sattler layer
- Ellipsoid layer disruption (not strictly related to disease activity)
- SCF: suprachoroidal fluid
- Hyperreflective choroidal foci in the vicinity of BCR lesions ➜ inflammatory cells or pigment clumps

BCR: OCTA

- Deep choroidal en face slab → **hyporeflective lesions** of BCR
- **Flow void in the choriocapillaris layer under areas of RPE disruption** → true vessel atrophy or greatly reduced flow

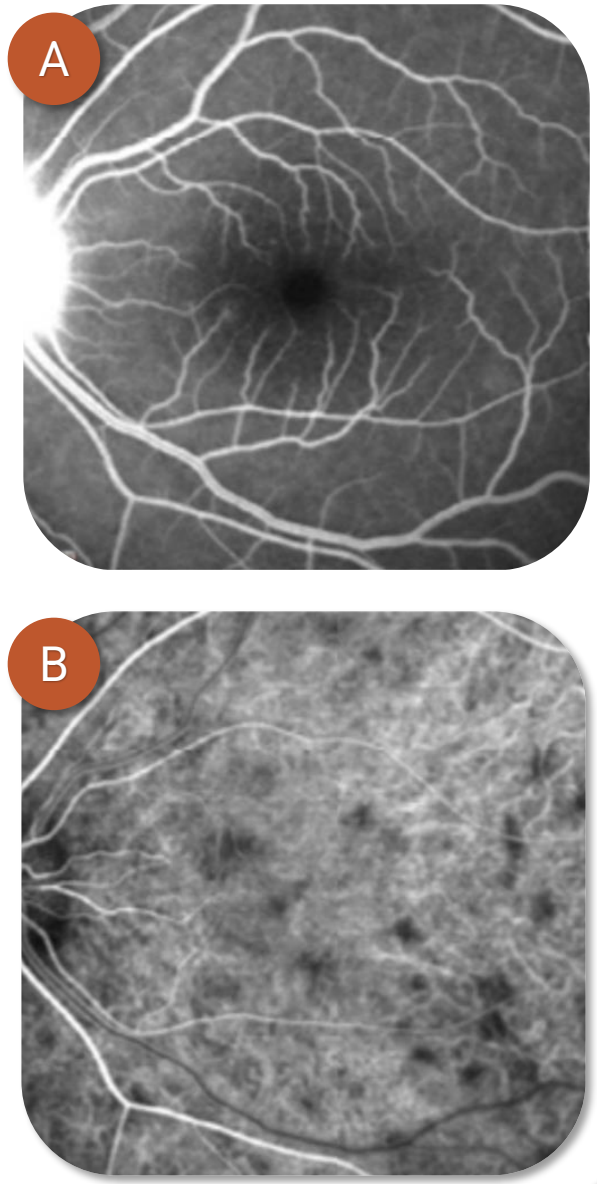


Tuberculosis and sarcoidosis: FA/ICGA

- **GRANULOMA**: the **primary lesion of stromal choroiditis** → immune cells, macrophages, epithelioid cells encircled by lymphocytes

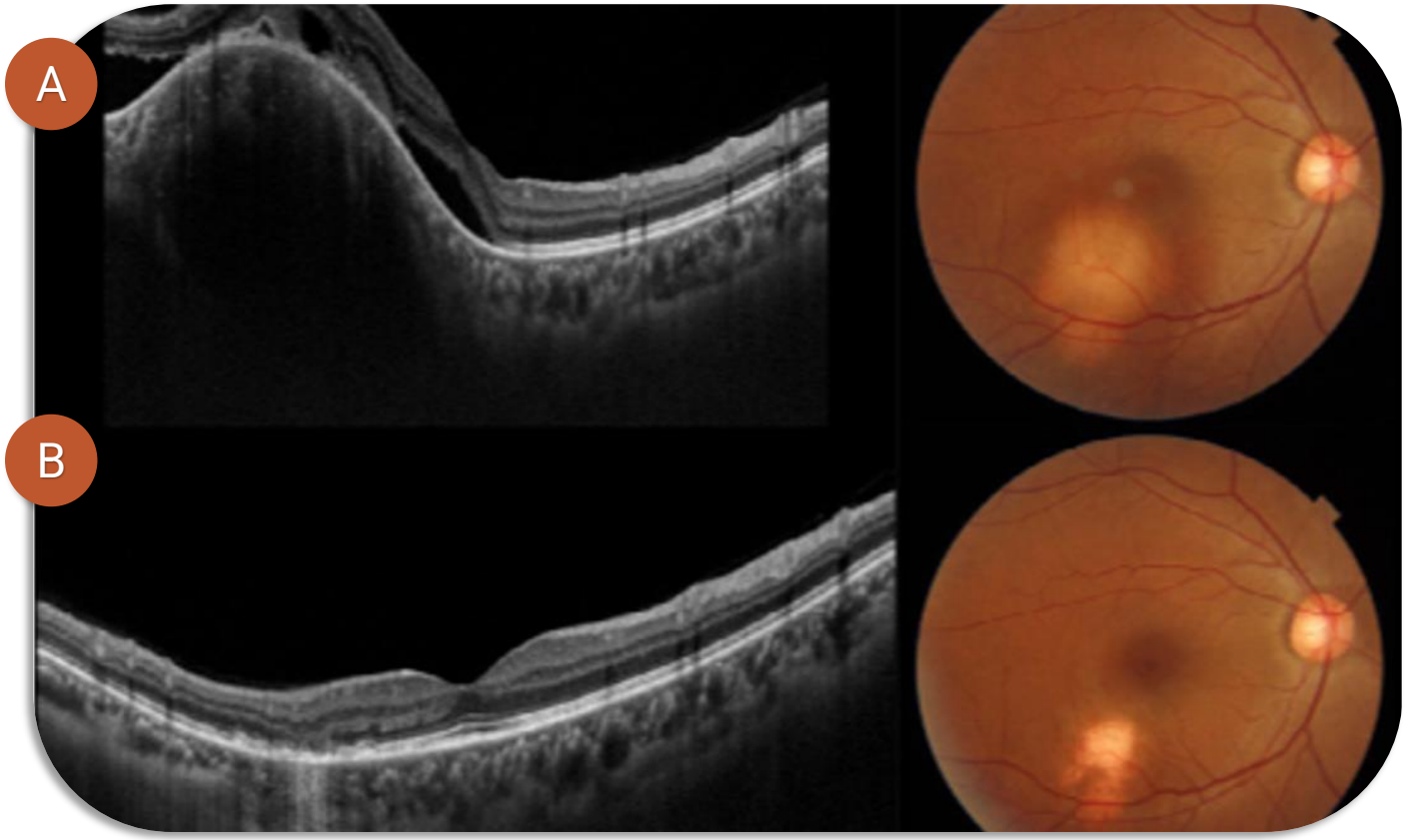
- **ICGA**
  1. Multiple irregularly distributed **hypocyanescent spots** in the **early and intermediate phase** → **isocyanescent (type 1)** or remain **hypocyanescent (type 2**, choroidal atrophy) in the **late phase**
  2. **Fuzziness (reversible) of the choroidal vessels** in the **intermediate** phase
  3. **TB**: fuzziness of choroidal vessels and diffuse zonal hypercyanescence (**choroidal staining**) → less common

- **Sarcoidosis: fuzziness of choroidal vessels and choroidal staining seen in 100% eyes**



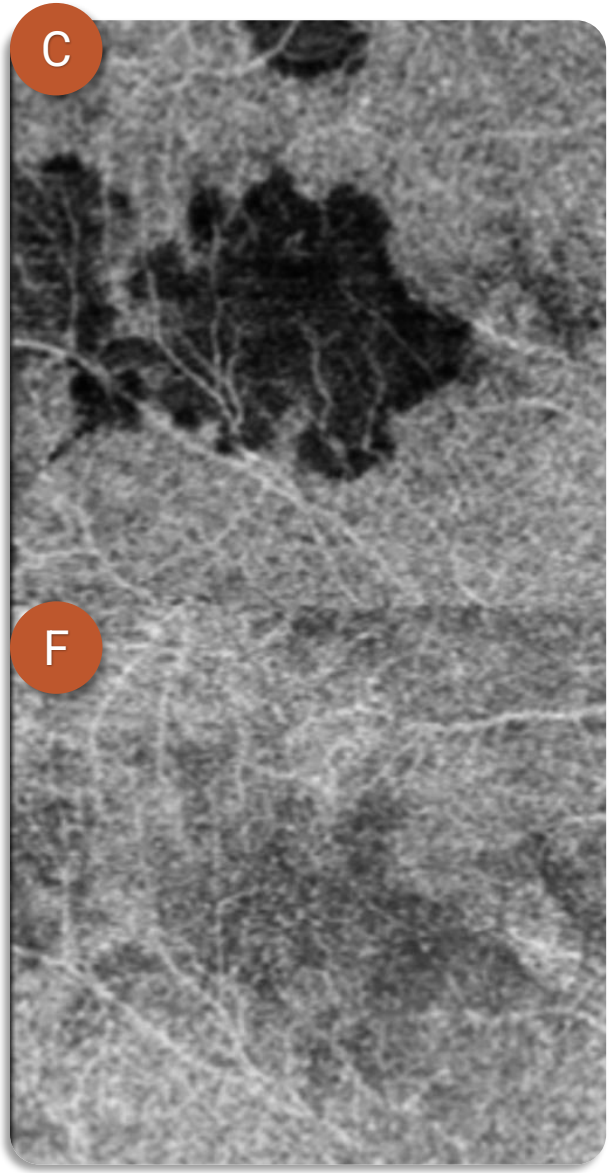
TB and sarcoidosis: OCT

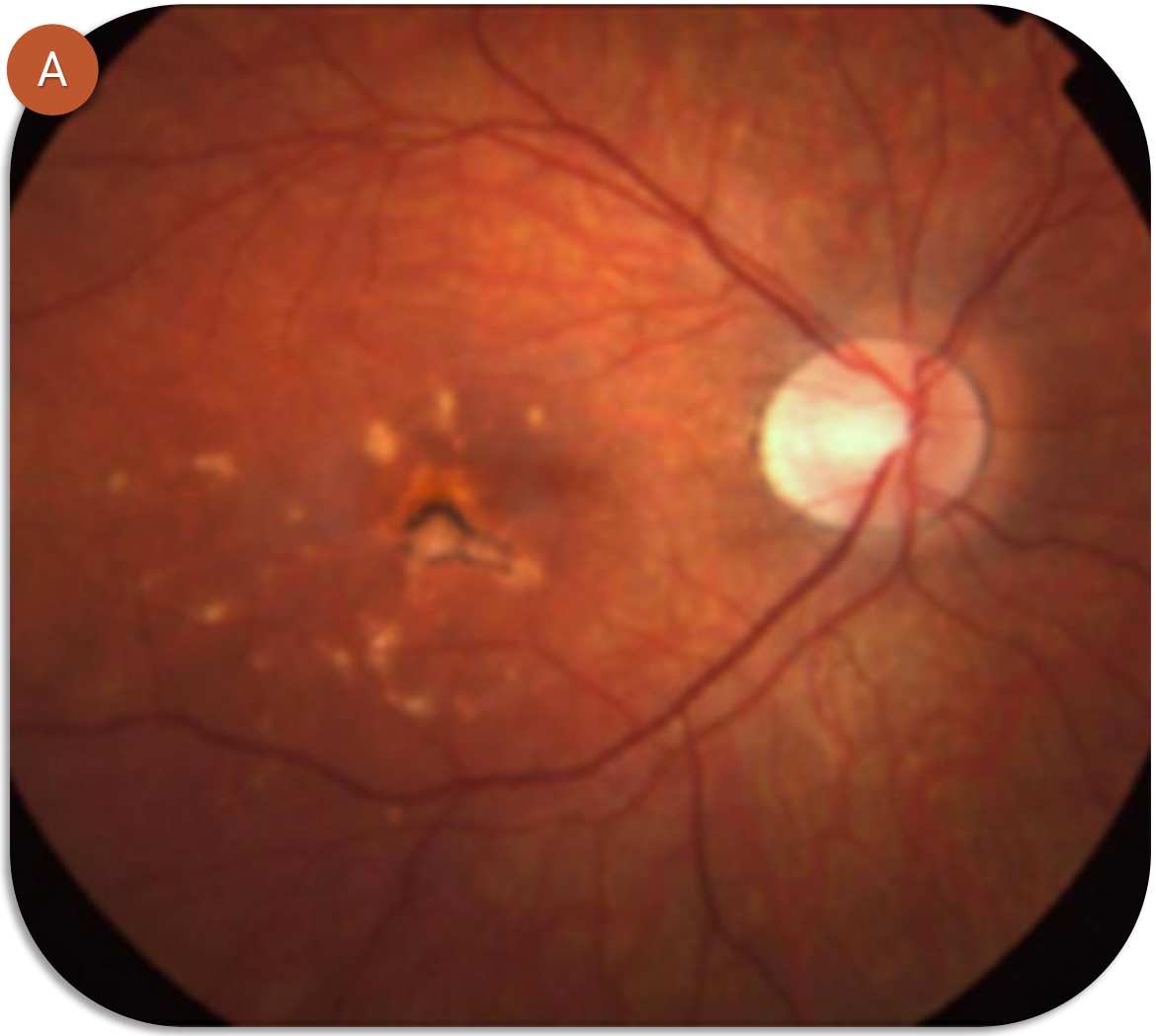
- **TB choroidal granuloma → elevation of the choroid** with a **localized attachment of the choriocapillaris-RPE complex to the overlying neurosensory retina** with **surrounding subretinal fluid**
- **TB: generalized choroidal thickening**
- **TB GRANULOMAS**: lobulated shape and a non-homogenous internal pattern
- **Sarcoidosis**: generalized choroidal thickening with **Sattler medium vessel layer disproportionately thickened**
- **Sarcoidosis granulomas →** more commonly localize in the **inner choroidal stroma**



TB and sarcoidosis: OCTA

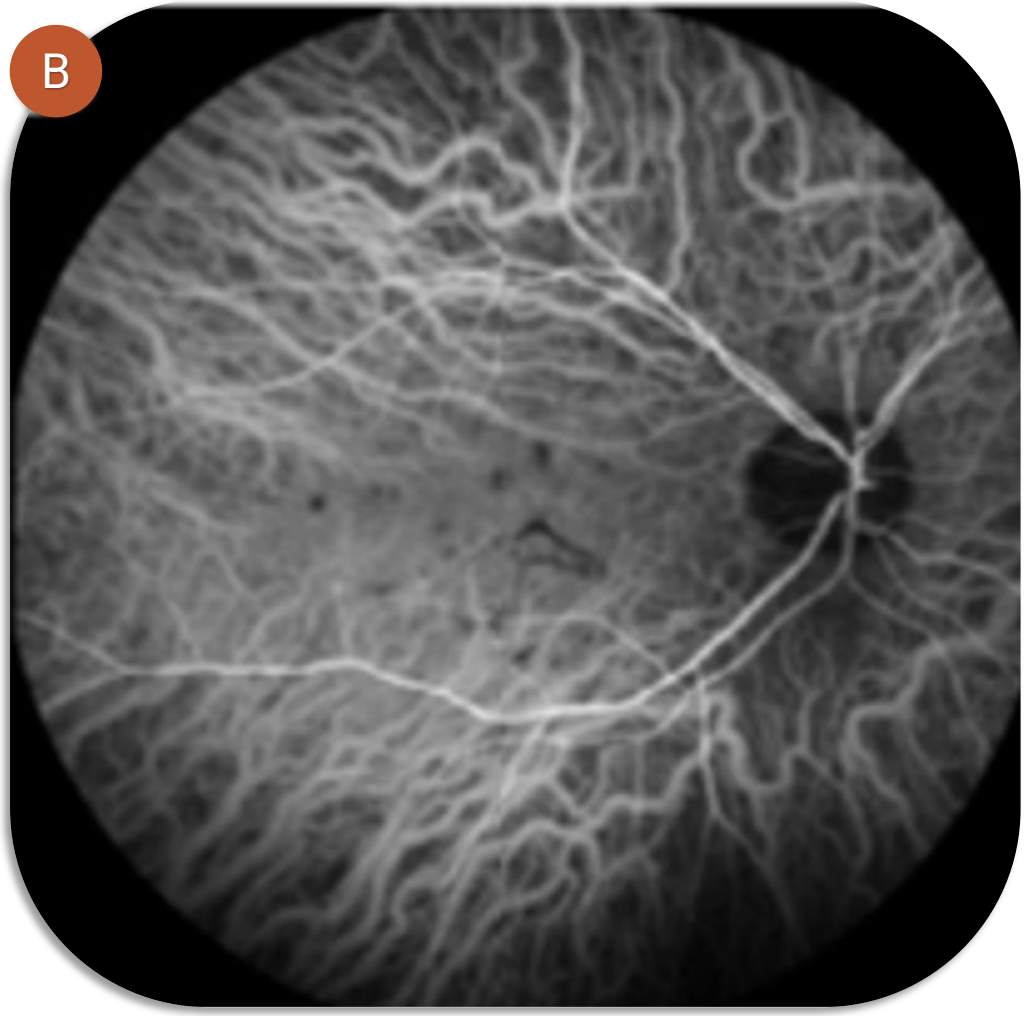
- The small granulomas are not identified on OCTA
- Larger, full-thickness choroidal granulomas → areas of choriocapillaris non-flow on OCTA
- OCTA shows **reperfusion of choriocapillaris** with a **reduction in density** **after treatment**
- Compared to ICGA → **OCTA**: better resolution images of choriocapillaris and **choriocapillaris hypoperfusion and atrophy** can be distinguished





Multifocal choroiditis and panuveitis (MCP) punctate inner choroidopathy (PIC)

- Both entities: in **young myopic woman** → **multifocal yellowish chorioretinal lesions** in the posterior pole that evolve into **punched out scars**
- **Photopsias**
- **Visual field defects**
- **CNV**
- **Difference**: anterior chamber and vitritis and larger size of lesions in MCP



MCP/PIC: FA/ICGA

- **ICGA: ACTIVE DISEASE → early hypocyanescent spots**, more **prominent in the late phase**
- They do not correspond to lesions detected clinically or on **FA → they correspond to areas of visual field defects**
- **ICGA**: **HEALED: They disappear**
- **FA**: **ACTIVE DISEASE → early isofluorescence and late leakage**
- **FA**: **HEALED → early isofluorescence and late staining**
- **CNV**: **early hyperfluorescence** with **late leakage**

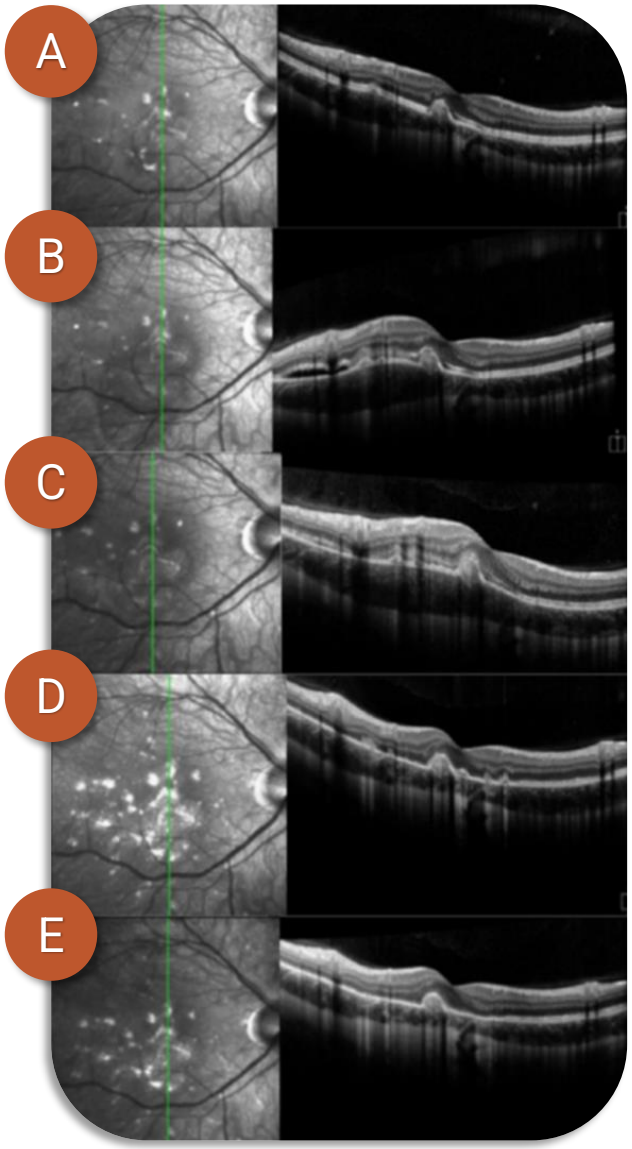
MCP/PIC: FAF/OCT

- **FAF: ACTIVE → hypo- isoautofluorescence in the center** with a **hyperautofluorescence surround**
- **FAF: HEALED → hypoautofluorescent** lesions

- **OCT: ACTIVE →**
  1. **Conical elevation of the RPE**
  2. **Underlying homogenous hyperreflective material**
  3. **Widespread disruption of the ellispoid zone**
  4. **Choroidal thickening**
  5. **Increased choroidal reflectivity**

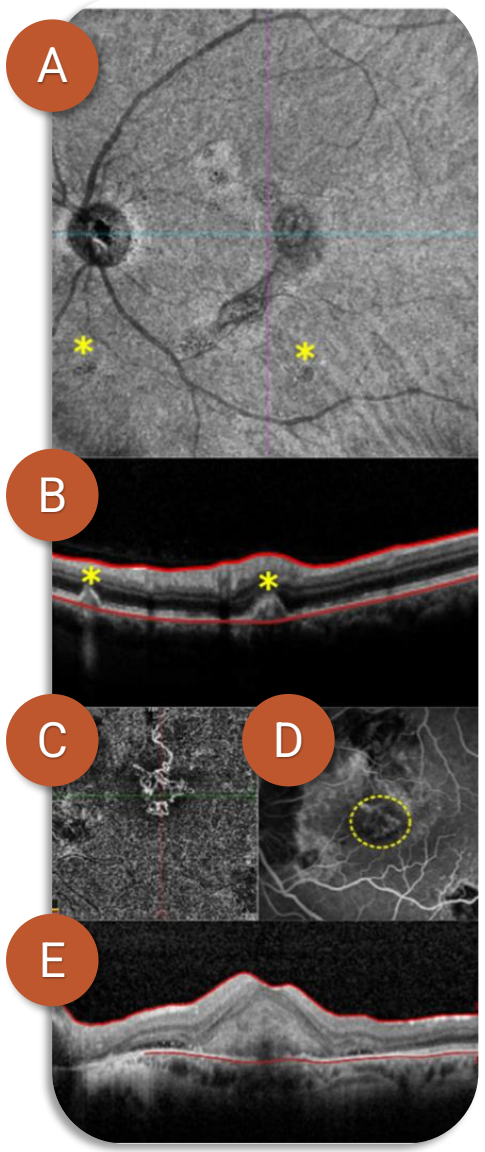
- **OCT: HEALED →** reformation of the ellipsoid zone, choroidal thinning, decrease in choroidal reflectivity

- **NO CONSISTENT INVOLVEMENT OF THE CHOROIDAL STRUCTURE → OUTER RETINA AND RPE ARE MAINLY INVOLVED**



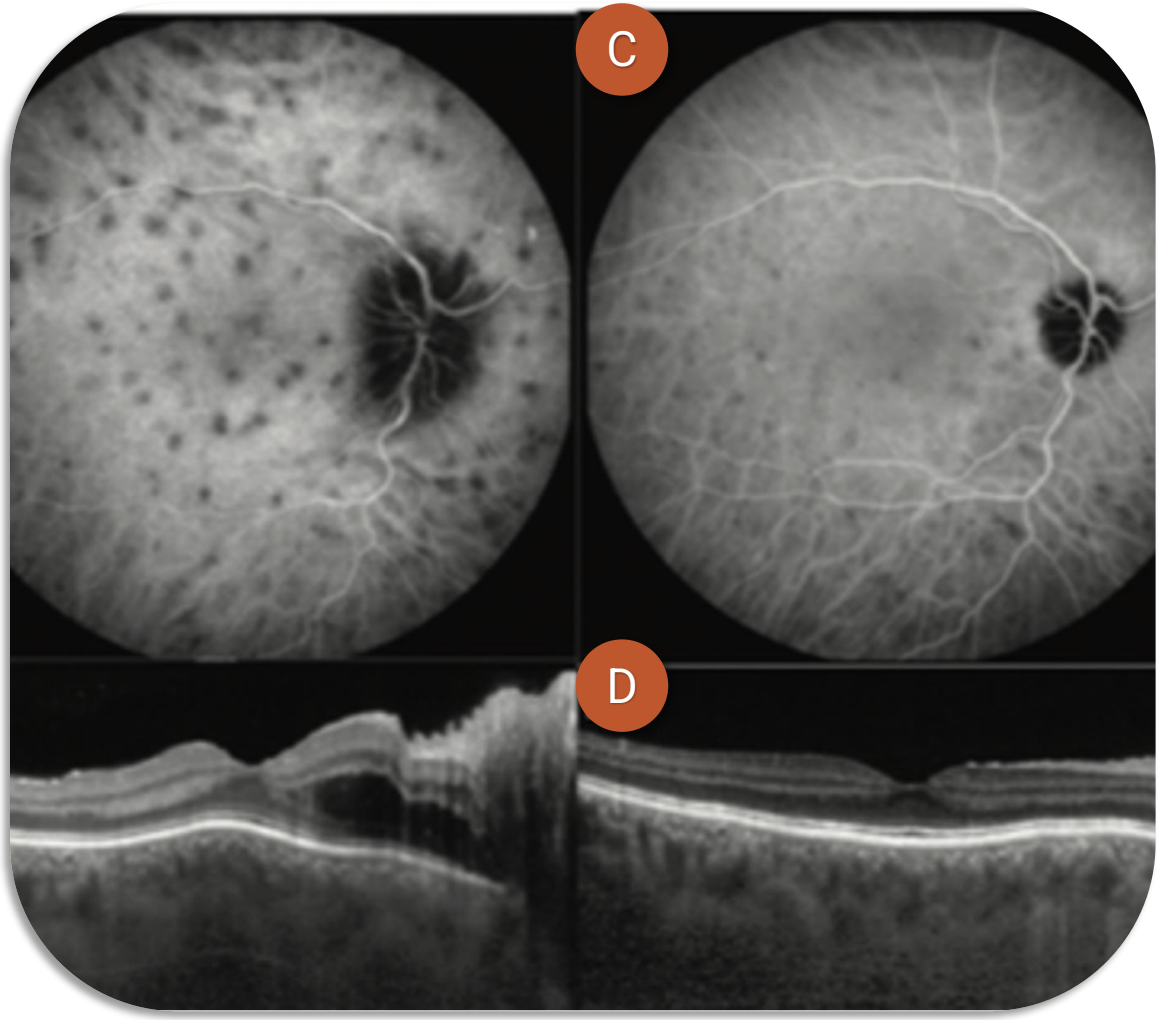
MCP/PIC: OCTA

- **CNV** is the primary cause of vision loss in MCP/PIC
- **Type 2 CNV**: subretinal heterogenous hyperreflective material
- **CNV**: vascular net in the outer retina segmentation → **multifocal, smaller and more well-circumscribed** compared to CNV associated with AMD and CSC
- **ACTIVE CNV** in MCP/PIC: **neovascular network with surrounding hyporeflectivity, leakage on FA**
- **INACTIVE CNV**: vascular net, no flow signal, no leak on FA
- **QUIESCENT CNV**: vascular net, flow signal present, no leak on FA



VOGT-KOYANAGI-HARADA disease (VHK)

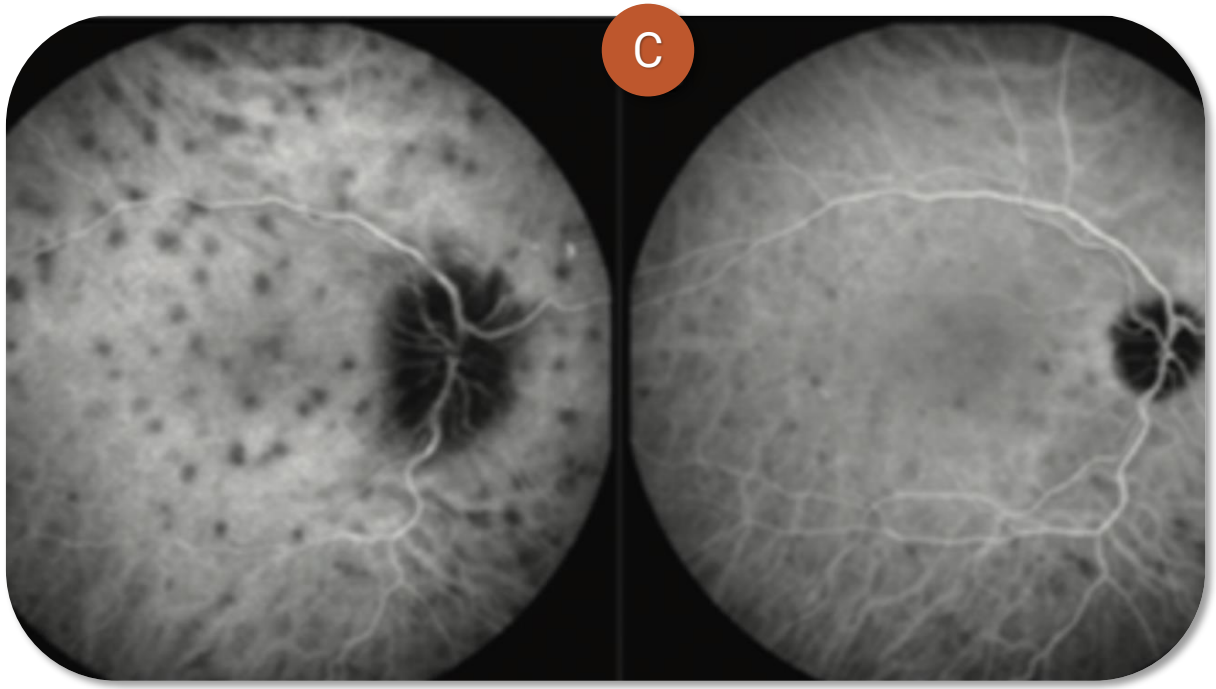
- Idiopathic
- Multisystem
- Granulomatous
- Autoimmune
- **Cell-mediated immunity against melanocytes**
- Ocular, neurologic, integumentary and auditory manifestations
- In the eye ➜ it primarily involves the CHOROIDAL STROMA and manifests as a BILATERAL GRANULOMATOUS PANUVEITIS

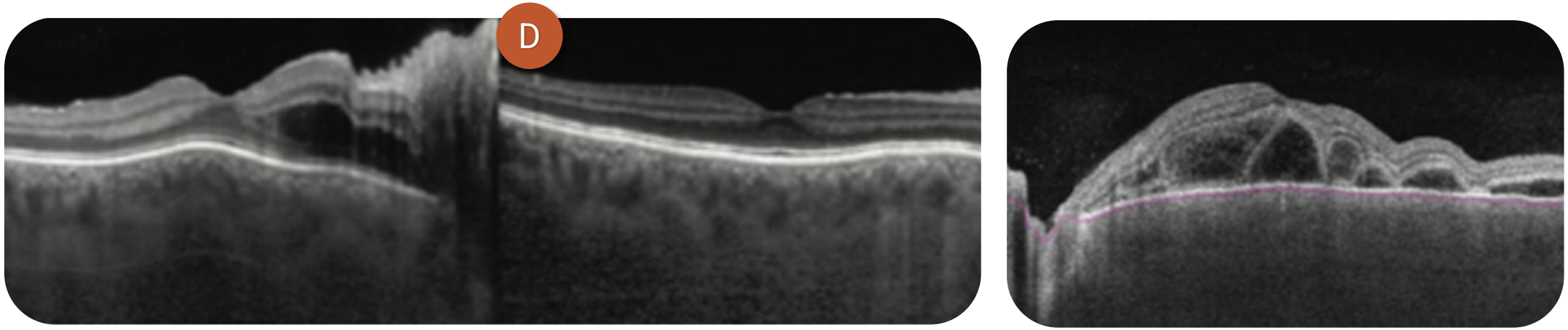


VKH:FA/ICGA

- **FA: ACTIVE** ➜ numerous puctate hyperfluorescent pinpoints of dye at the level of RPE ➜they gradually enlarge and pool in the subretinal space (serous retinal detachment)

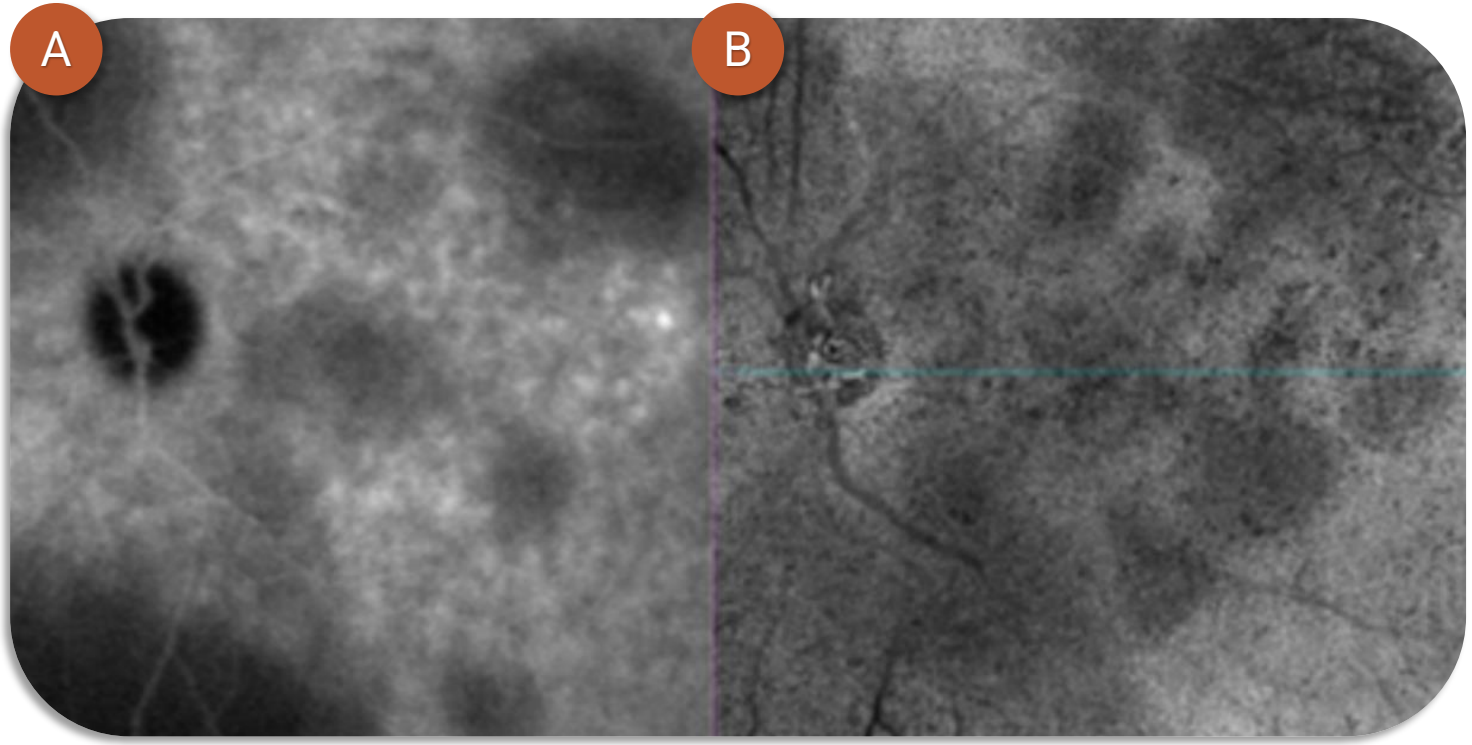
- **ICGA: ACTIVE** :
  1. Early choroidal vessel hypercyanescence (CHOROIDAL VASCULITIS)
  2. Disc hypercyanescence
  3. Fuzziness of choroidal vessels in the mid and late phase
  4. Multiple hypocyanescent dark dots (HDD) which become isocyanescent in the late phase or remain hypocyanescent ➜ HDD are the LAST TO RESOLVE ➜INFLAMMATORY INFILTRATION OF THE CHOROIDAL STROMA





VKH: OCT

1. Increased choroidal thickness ➜ inflammatory infiltration and exudation in the choroid
2. Choroidal thickening: marker of development of serous retinal detachment
3. Hyperreflective dots in the inner choroid (HDD on ICGA)
4. Reduced choroidal vascularity index (CVI) ➜ LUMINAL AREA (LA)/TOTAL CHOROIDAL AREA (TCA)

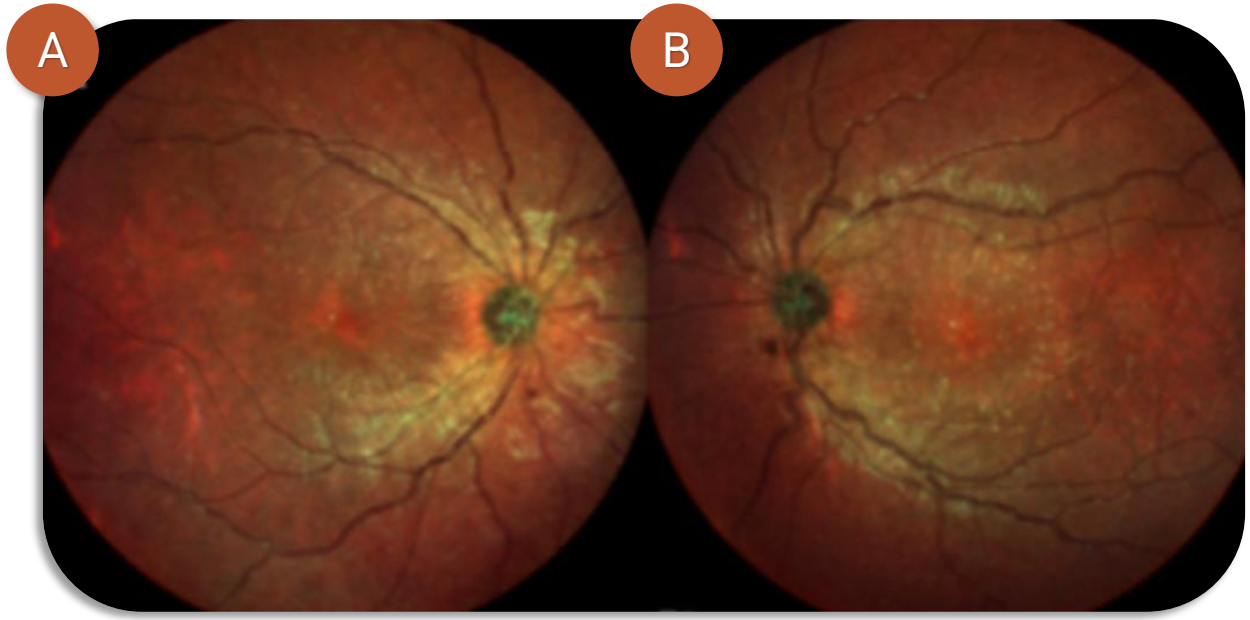


VKH: OCTA

Multifocal well-defined areas of flow void seen in the choriocapillaris segmentation corresponding to hypocyanescent areas seen on ICGA

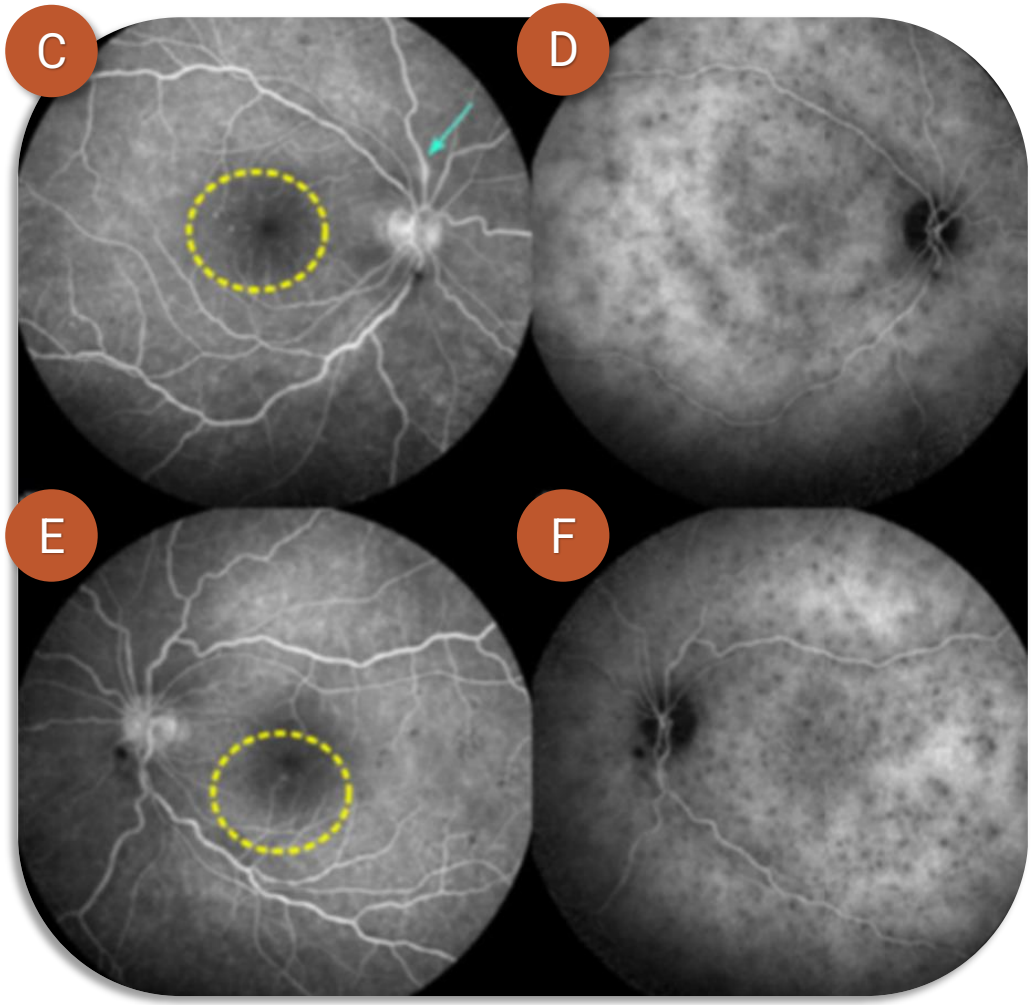
Multiple evanescent white dot syndrome (MEWDS)

- Rare
- Idiopathic
- Inflammatory disorder
- Unilateral, multiple, small, deep retinal white dots in the posterior pole
- Edematous optic disc
- Young myopic women
- Sudden onset of decreased vision
- Photopsias
- Enlarged blind spot
- Spontaneous and complete recovery of visual function within weeks



MEWDS: FA/ICGA

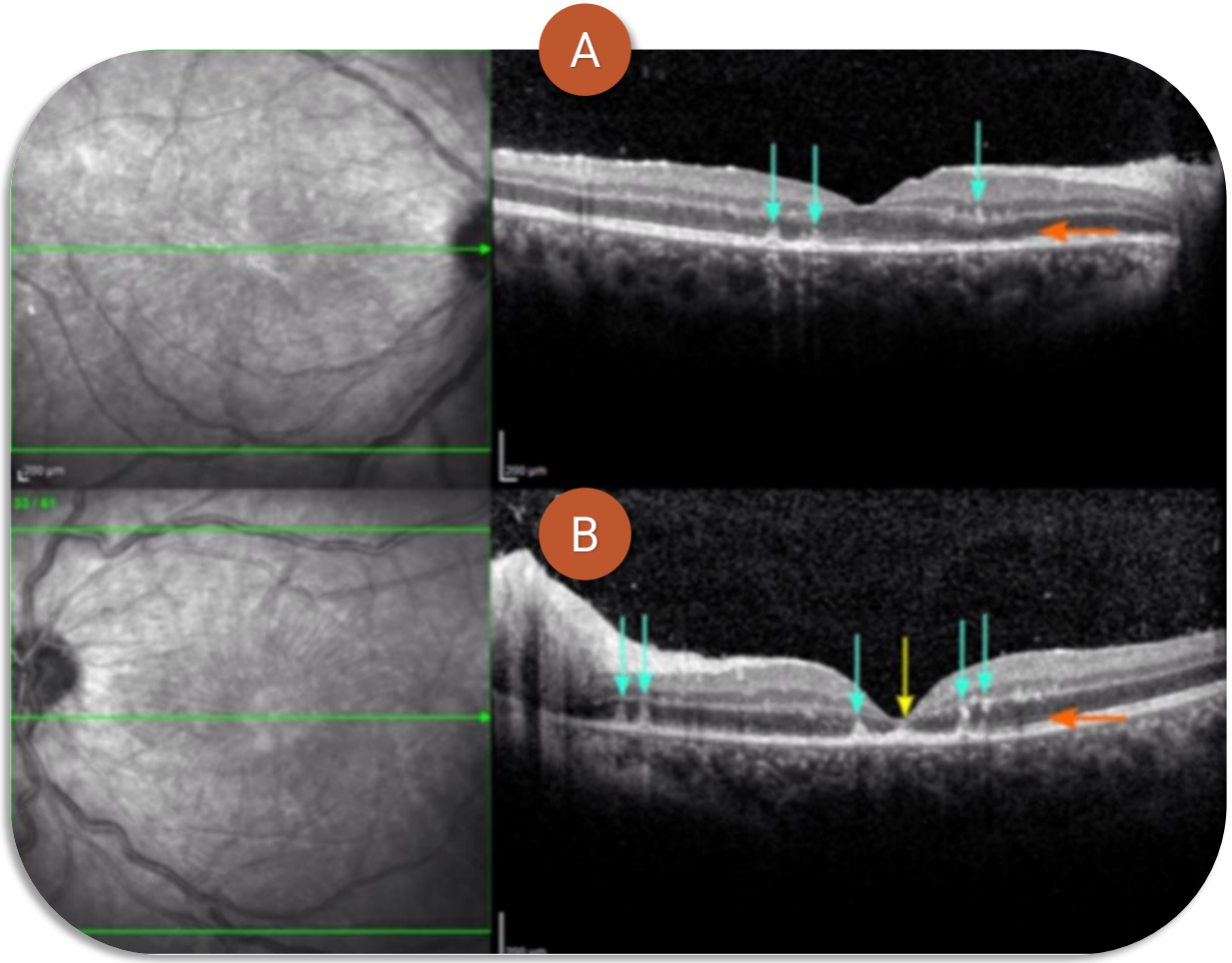
- **FA → early** patchy or wreath-like punctate **HYPERFLUORESCENCE** corresponding to **DOTS** and **late staining** of lesions corresponding to **SPOTS**
- **DOTS** (~100 microns): outer retina and RPE → stretching of the RPE cells by inflammatory exudates → **WINDOW DEFECT**
- **SPOTS** (> 200 microns): RPE and inner choroid → inflammatory exudates → **LATE STAINING**
- **ICGA → HYPOCYANESCENT** spots in the early or intermediate phase, becoming **more prominent in the late phase** → more numerous than seen clinically or on FA
- Hypocyanescent spots → inflammatory lesions in the choroid → narrowing of the precapillary arterioles and thickening of the choriocapillaris wall → DECREASED CIRCULATION

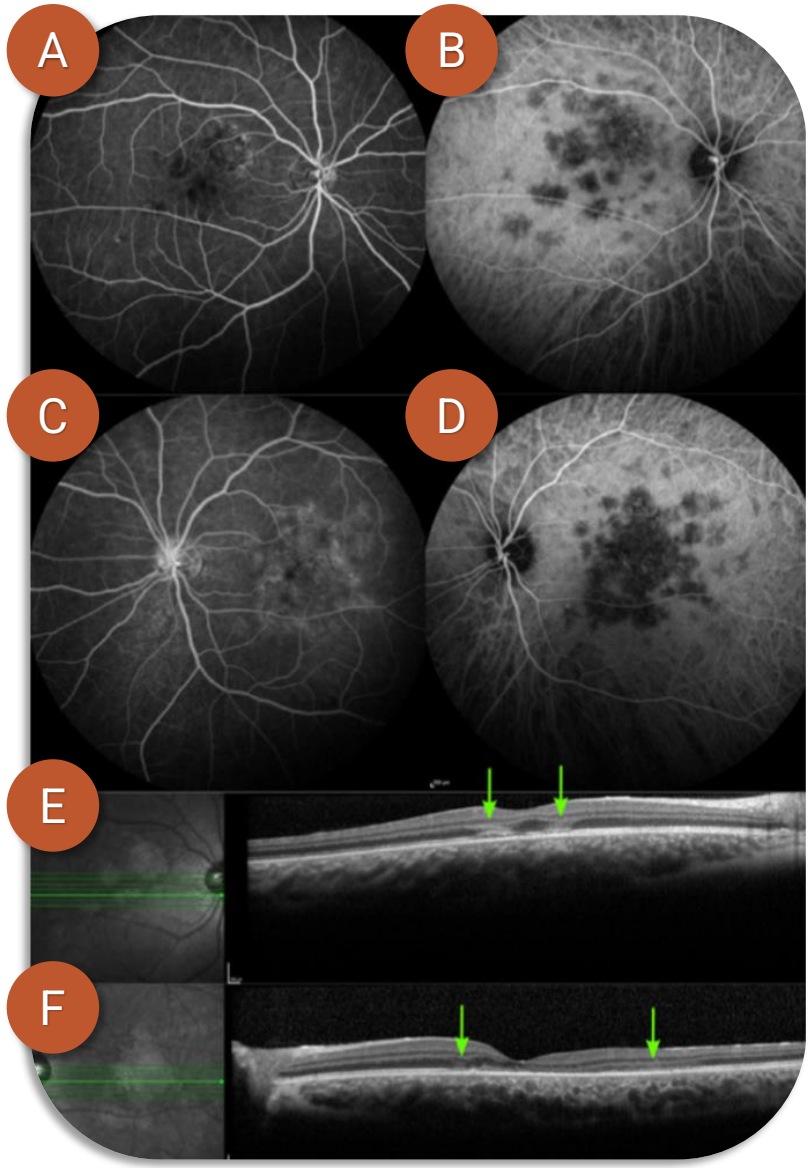


MEWDS: OCT

1. **Disruption of the ellipsoid zone**, often involving the interdigitation zone and ELM
2. **Hyperreflective dots in the outer nuclear layer (ONL)**
3. **Hyperreflective material on the inner aspect of RPE**

- **RPE**: key tissue in the inflammatory process → primary epitheliopathy
- **OCTA:** areas of flow void in the choriocapillaris layer, less numerous than seen clinically or OCT





Conclusion

- Advancements in **choroidal OCT** provide near histological images of the retina and choroid → **noninvasive study of choroidal morphology and vasculature**
- This has **revolutioned the understanding of choroidal involvement in various inflammatory conditions** to the extent of completely refuting several hypotheses based purely on angiographic studies
- Considering its **non-invasive nature**, ease of acquisition and improving analytics → **OCTA** is becoming the **preferred** imaging modality
- ***With wider application of multimodal imaging → every imaging technique finds its place in the clinical practice to improve patient care, classification of inflammatory diseases and response to therapy***.

Thank you

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