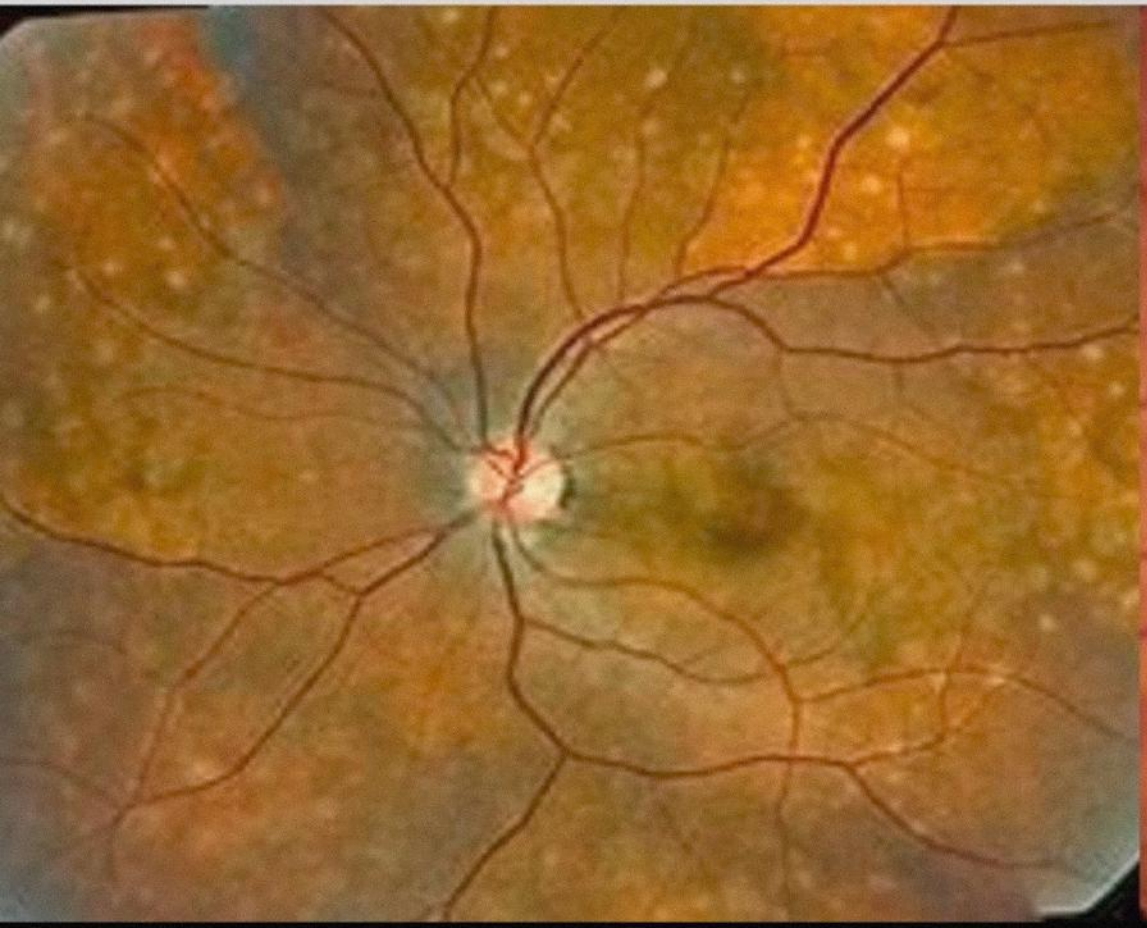
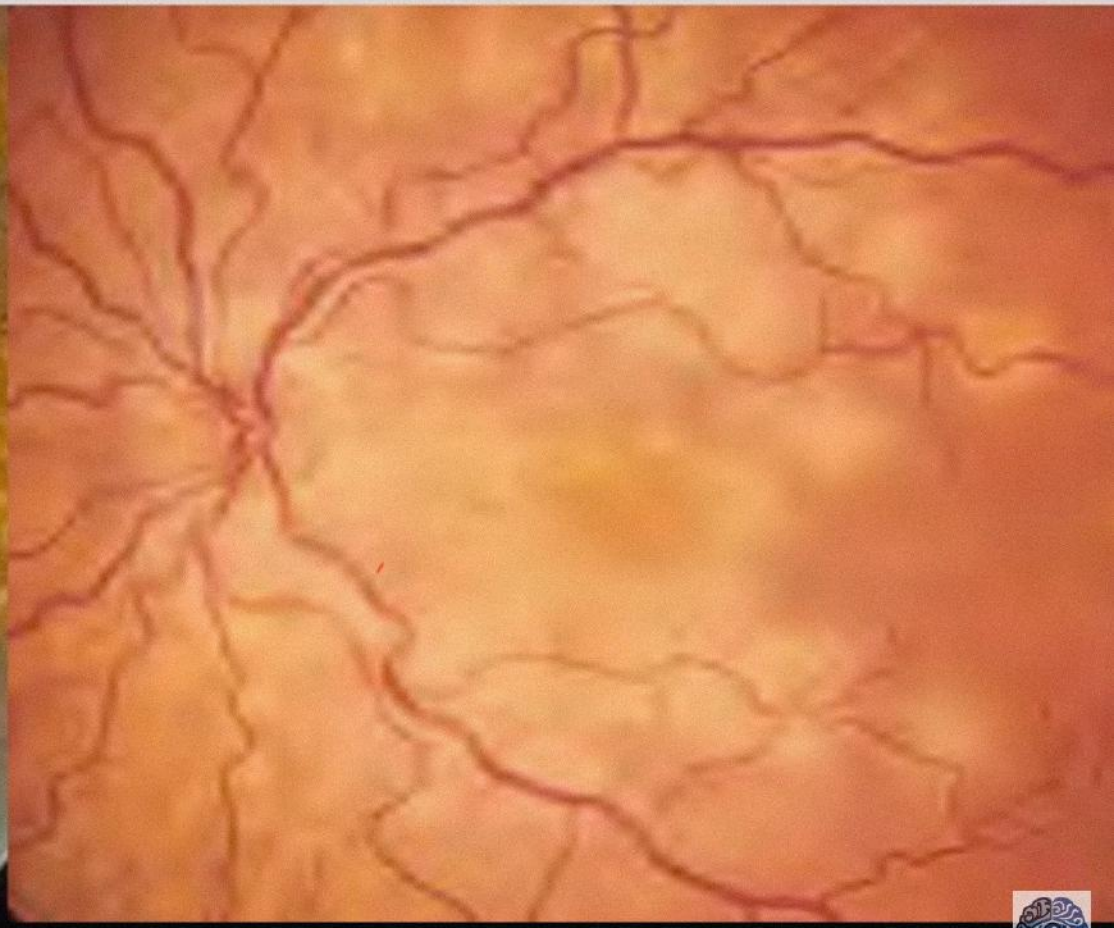
WHITE DOT SYNDROMES

MEWDS V/S APMPPE



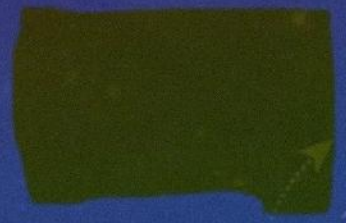


WHAT ARE WHITE DOT SYNDROMES ?

- ***IDIOPATHIC inflammatory*** multifocal disorders principally involving the posterior segment.

- Involves the **Outer retina, RPE and the Choroid**
- Uncommon

- Usually transient and do not cause a long term handicap



LETS ENUMERATE

1. Multiple evanescent white dot syndrome (MEWDS)
2. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
3. Birdshot chorioretinopathy
4. Punctate inner choroidopathy (PIC)
5. Serpiginous choroidopathy
6. Multifocal choroiditis and panuveitis
7. Subretinal fibrosis and uveitis

Many cases of uveitis, including

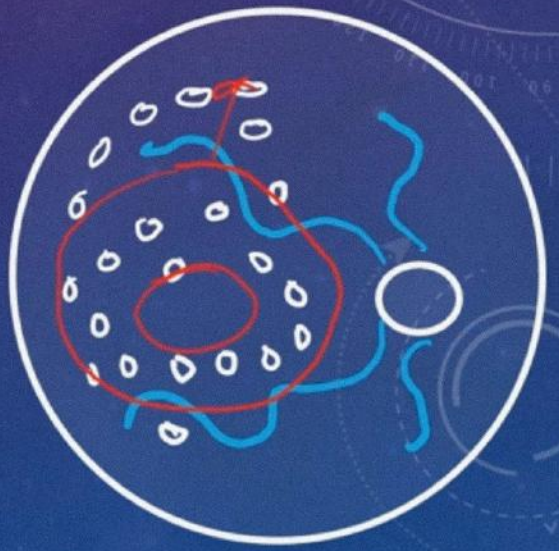
1. Multifocal choroiditis,
2. Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
3. Multiple evanescent white-dot syndrome (MEWDS)

} associated with a VIRAL ETIOLOGY

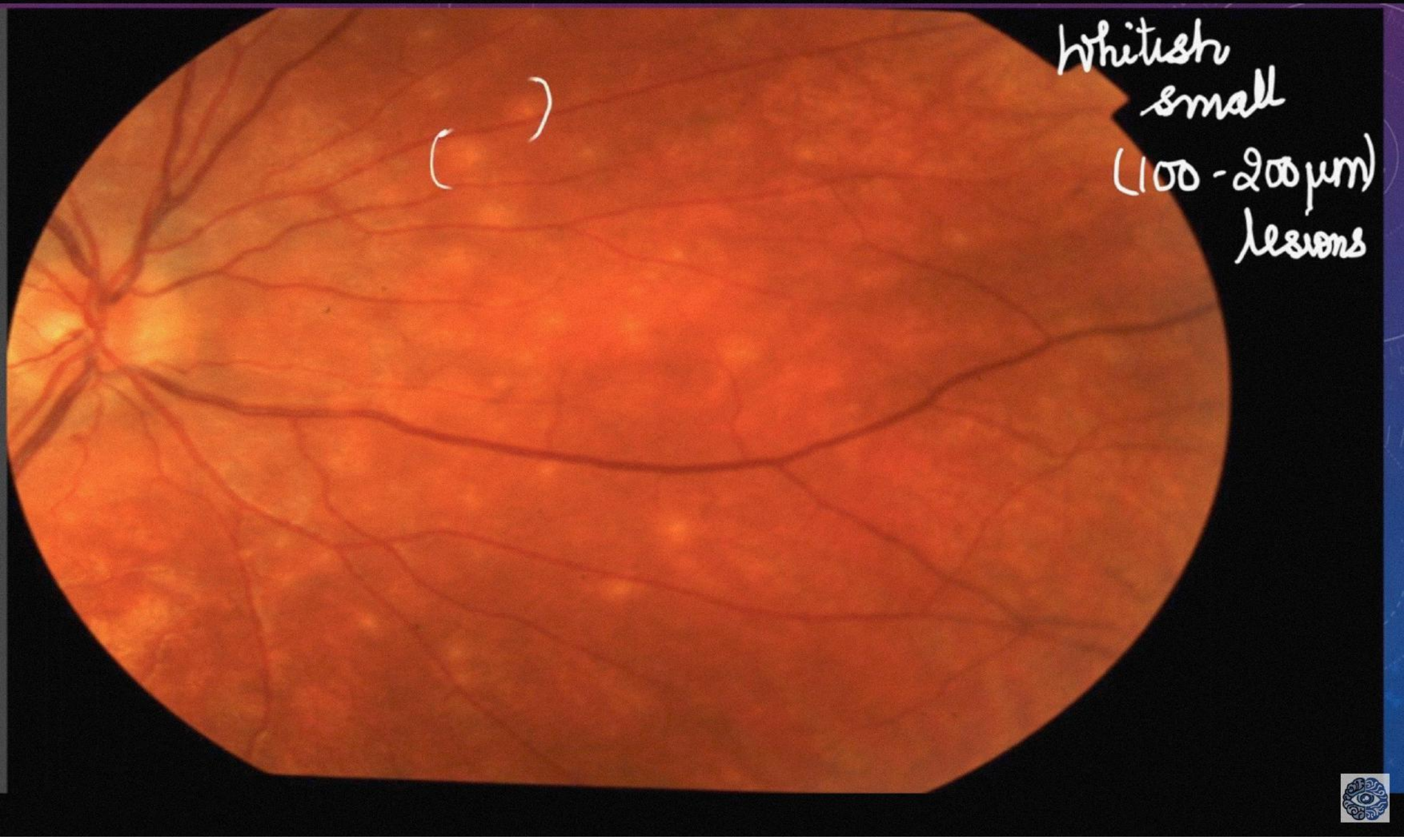
Multiple evanescent white dot syndrome (MEWDS)

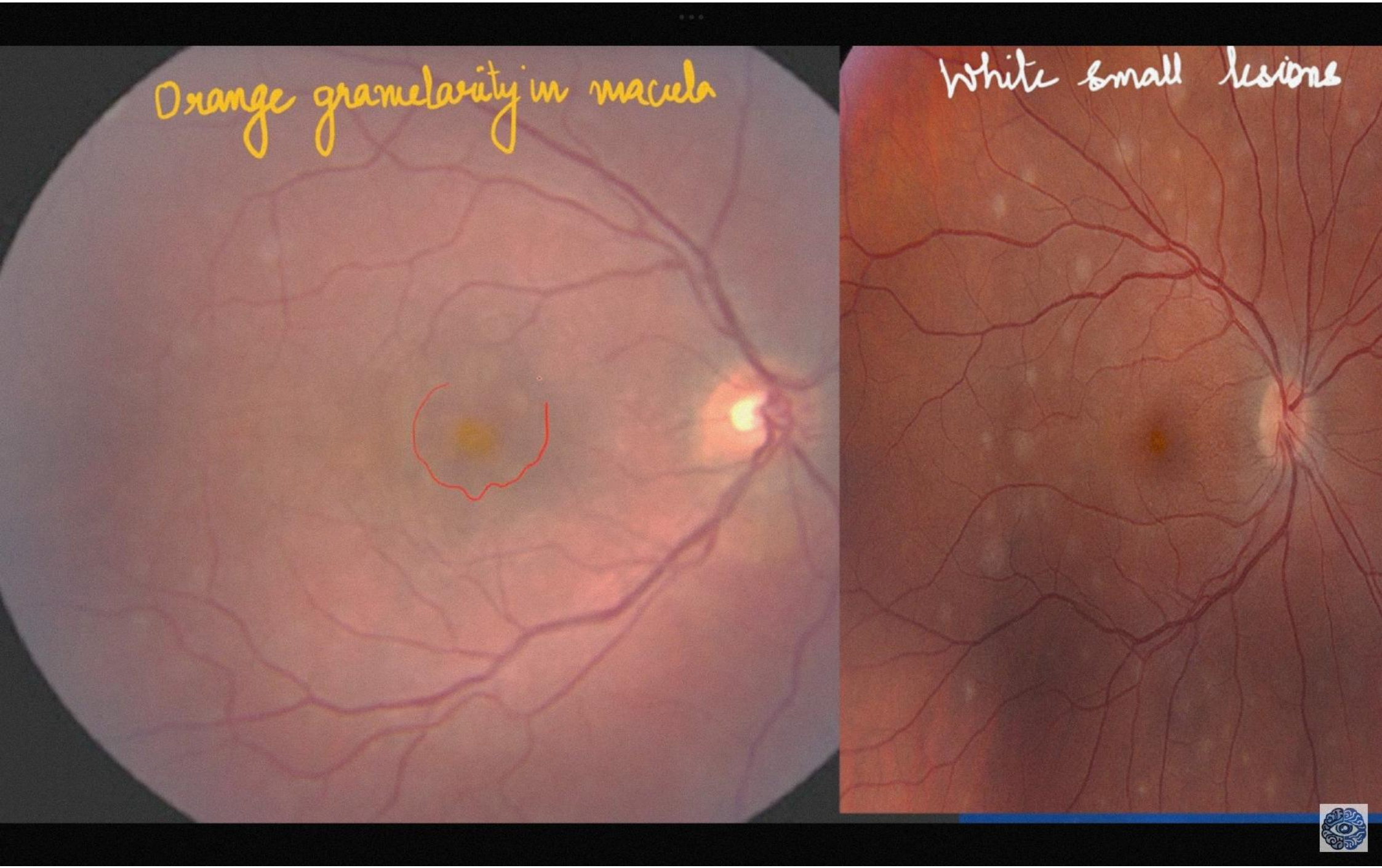
MEWDS FEATURES

- **MULTIPLE SMALL (100–200 µm),** discrete white lesions are noted deep in the retina or at the level of the retinal pigment epithelium (RPE)
- Appear in the posterior pole and extend to the midperiphery.
- Concentrated in the perifoveal region, but seem usually to spare the fovea itself.
- There is often a **granular appearance** to the macula.



- The granularity→ the appearance of tiny white or orange specks, which do not approach the size of the deeper circular lesions


Whitish
small
(100-200 μm)
lesions


Orange granularity in macula
White small lesions

WHY THEY ARE CALLED EVANESCENT ?

- The white lesions and macular granularity will fade with time, but subtle RPE alterations can be noted.
- Fade with time on their own **in 2-6 weeks**

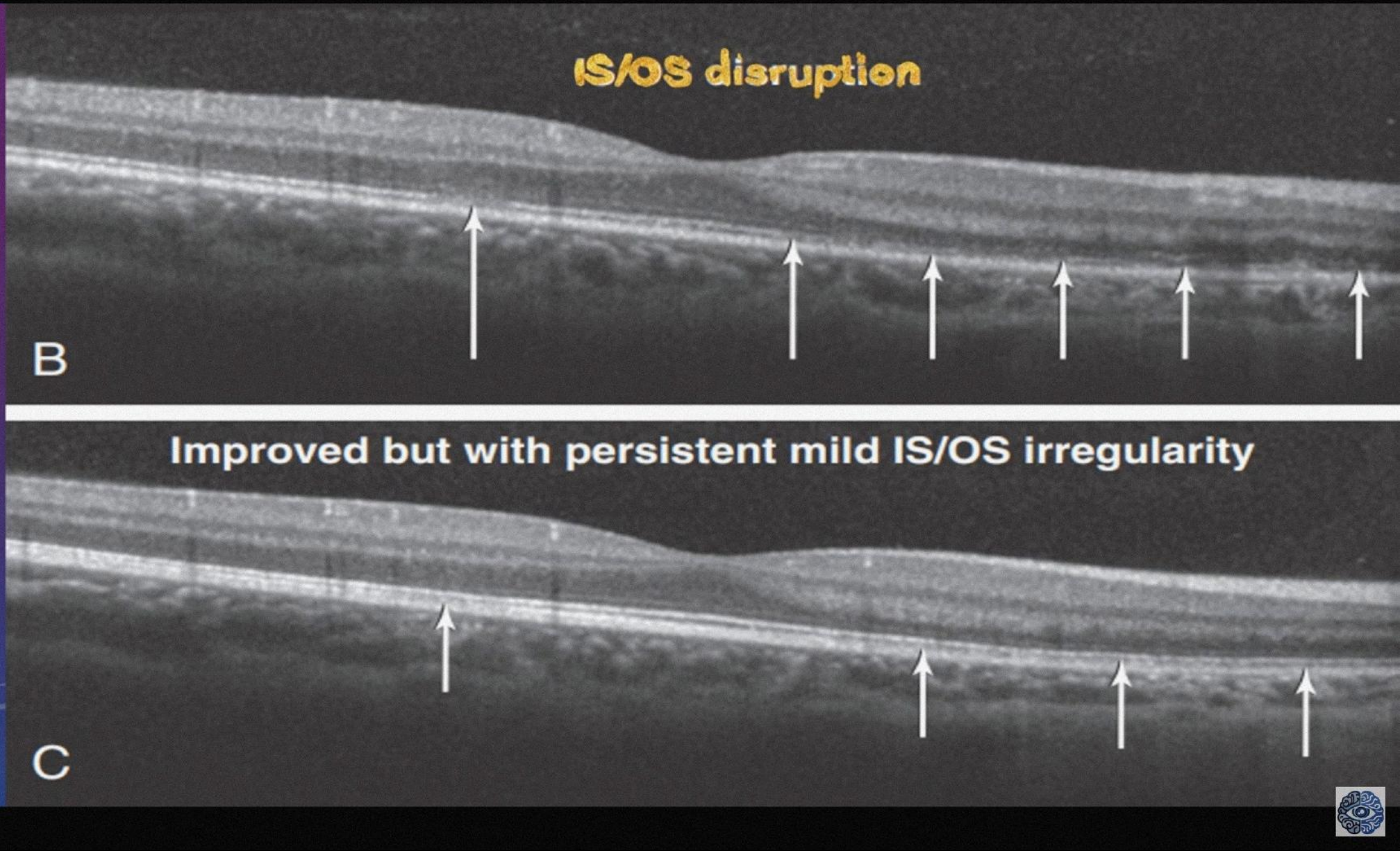
WHO IS AFFECTED ?

- Young female (10-47 yrs.)
- MYOPIC females
- COMMONLY present as **painless monocular blurring (6/9–6/60**) and photopsia.
- LESS COMMONLY as floaters and scotomata

INVESTIGATIONS IN MEWDS

OCT

May show inner-segment/outer-segment junction disruption.

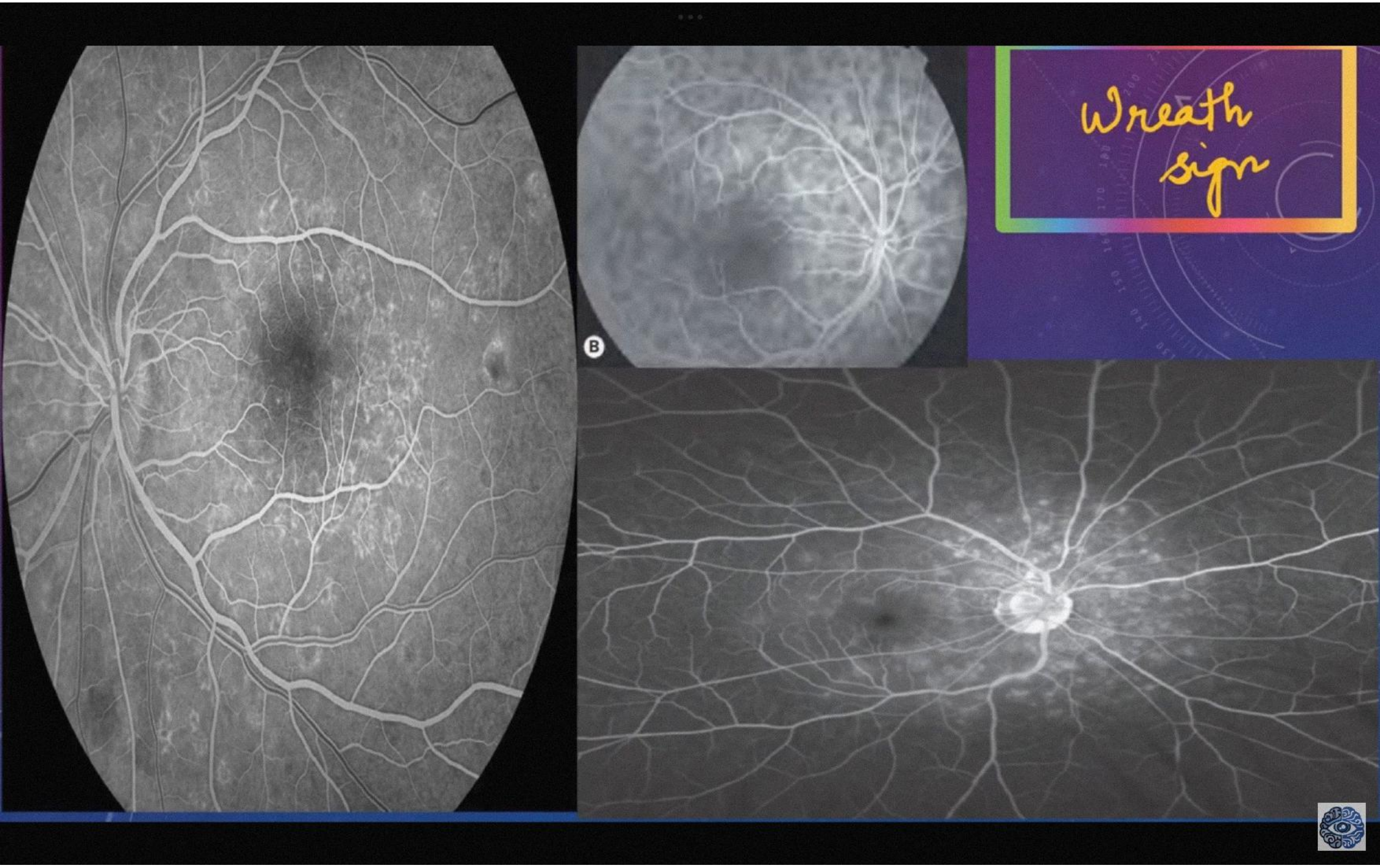

IS/OS disruption
B
Improved but with persistent mild IS/OS irregularity
C

FFA (FUNDUS FLUORESCEIN ANGIOGRAPHY)

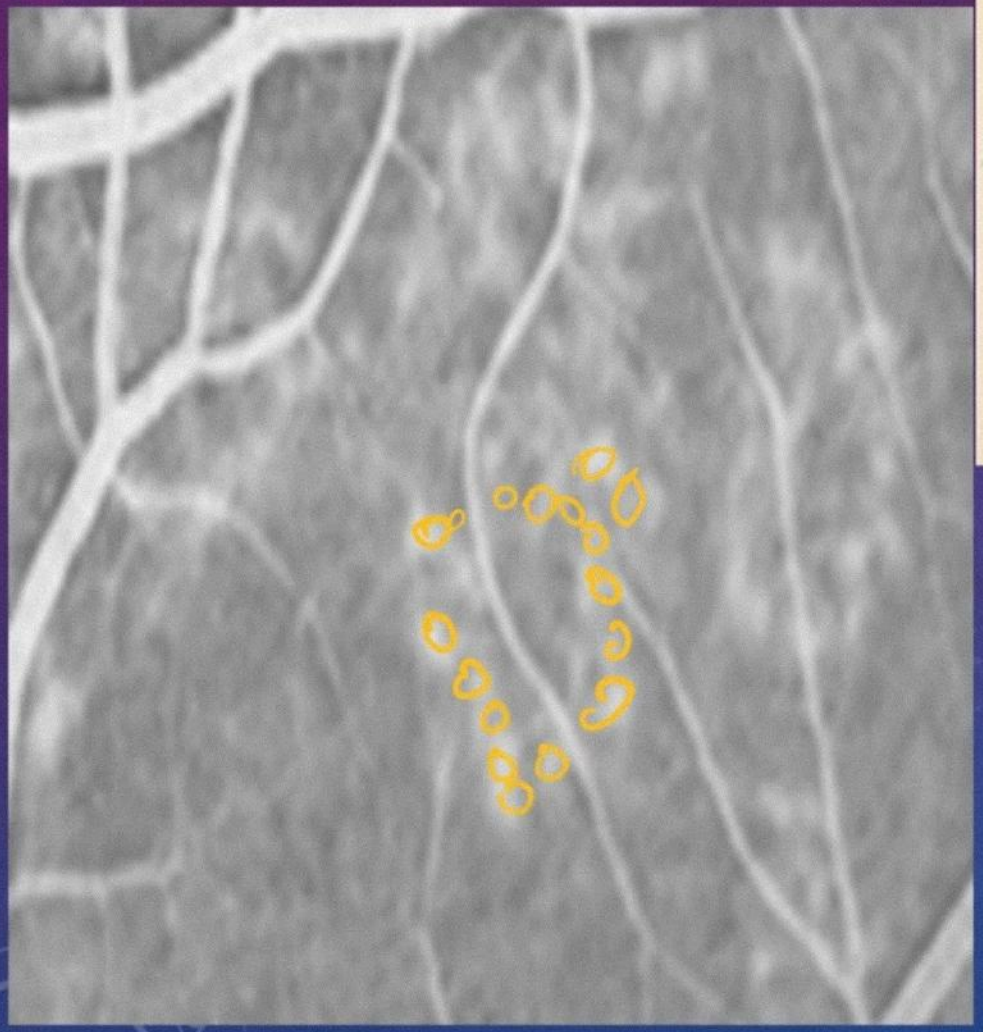
- FA reveals **early punctate hyper fluorescence** in a ***wreath-like pattern***
- **Late staining**, in areas corresponding to the white dots.

- This hyper fluorescence may be due to dilated retinal microcirculation in middle or deep retinal capillary plexus.

- Retinal vascular sheathing and optic nerve staining may be seen in some patients with MEWDS.


Wreath sign
B

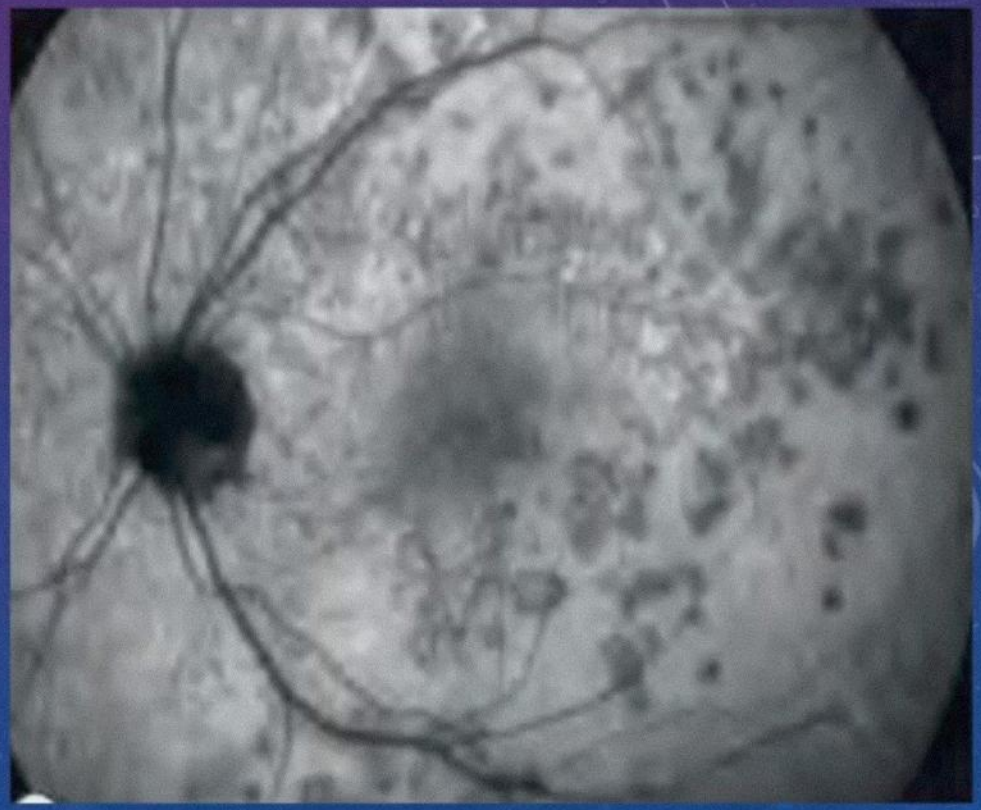
Wreath sign



HYPER ⟶ HYPER

ICG (INDOCYANINE GREEN ANGIOGRAPHY)

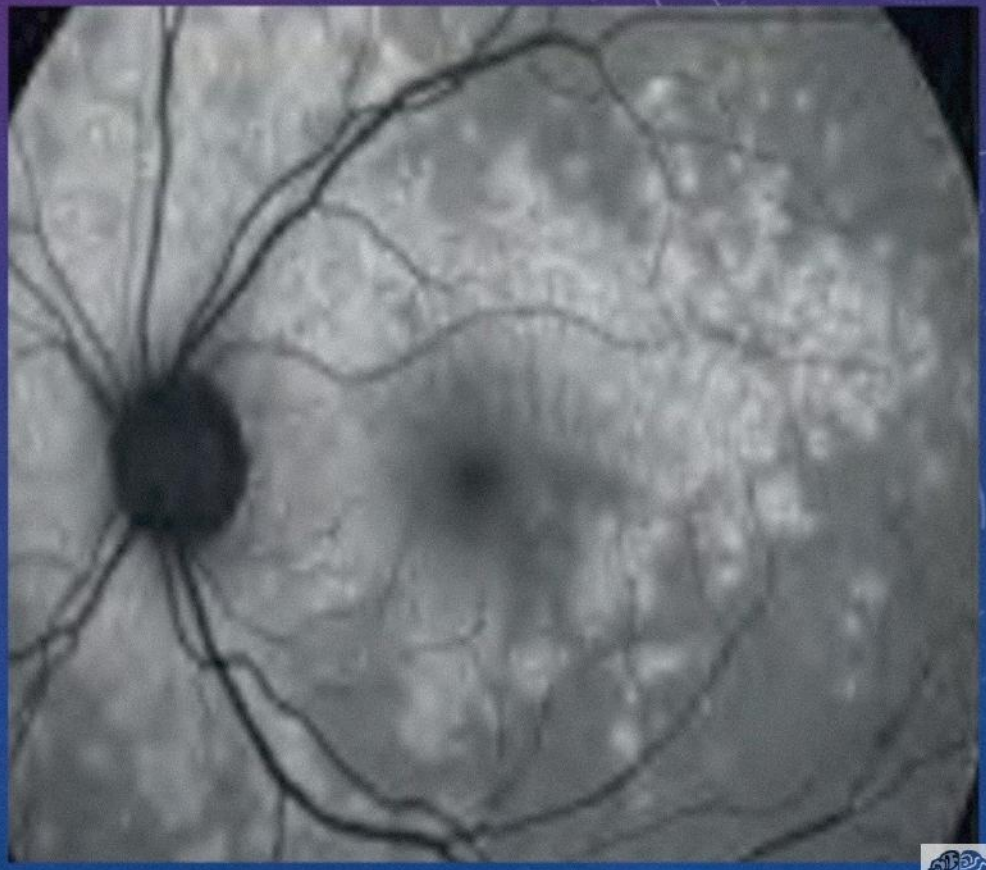
- ICGA shows **hypo cyanescent** spots that are often more numerous than visible clinically or on FA.



FAF (FUNDUS AUTOFLOURESCENCE)

- **Hyperautofluorescent spots** corresponding to the macular lesions are visible during active inflammation.

- FAF has been used to demonstrate subclinical lesions in patients with only foveal granularity



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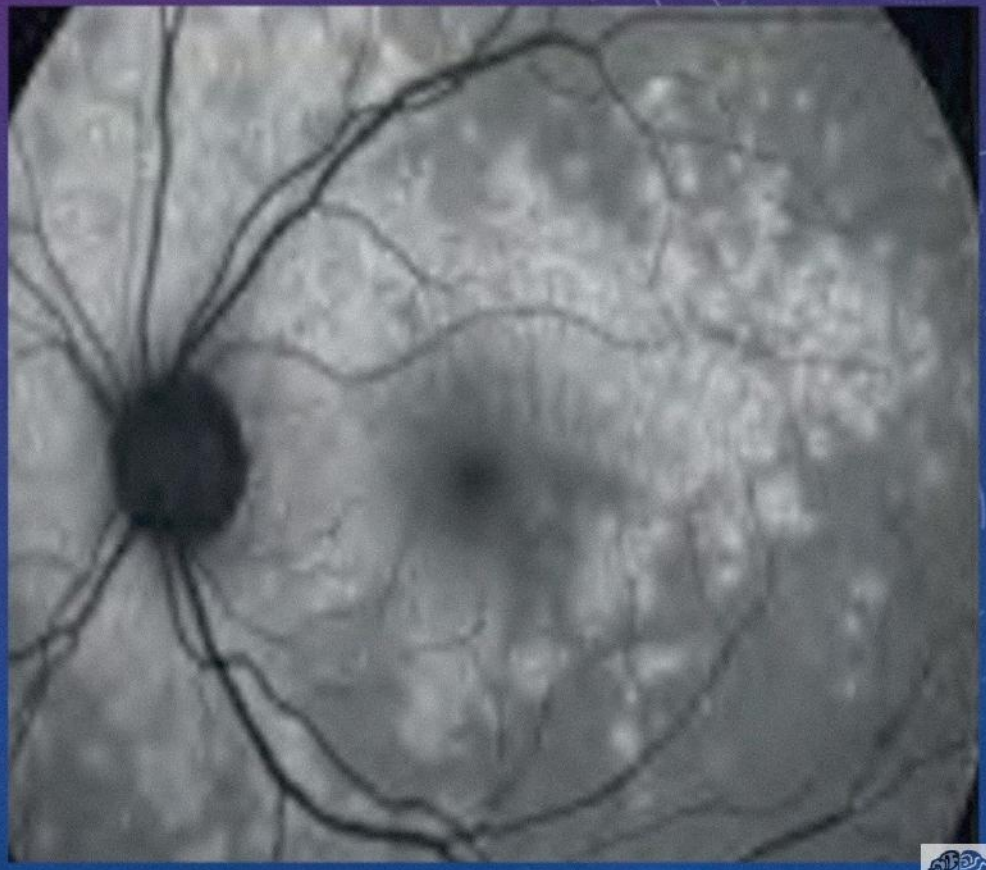
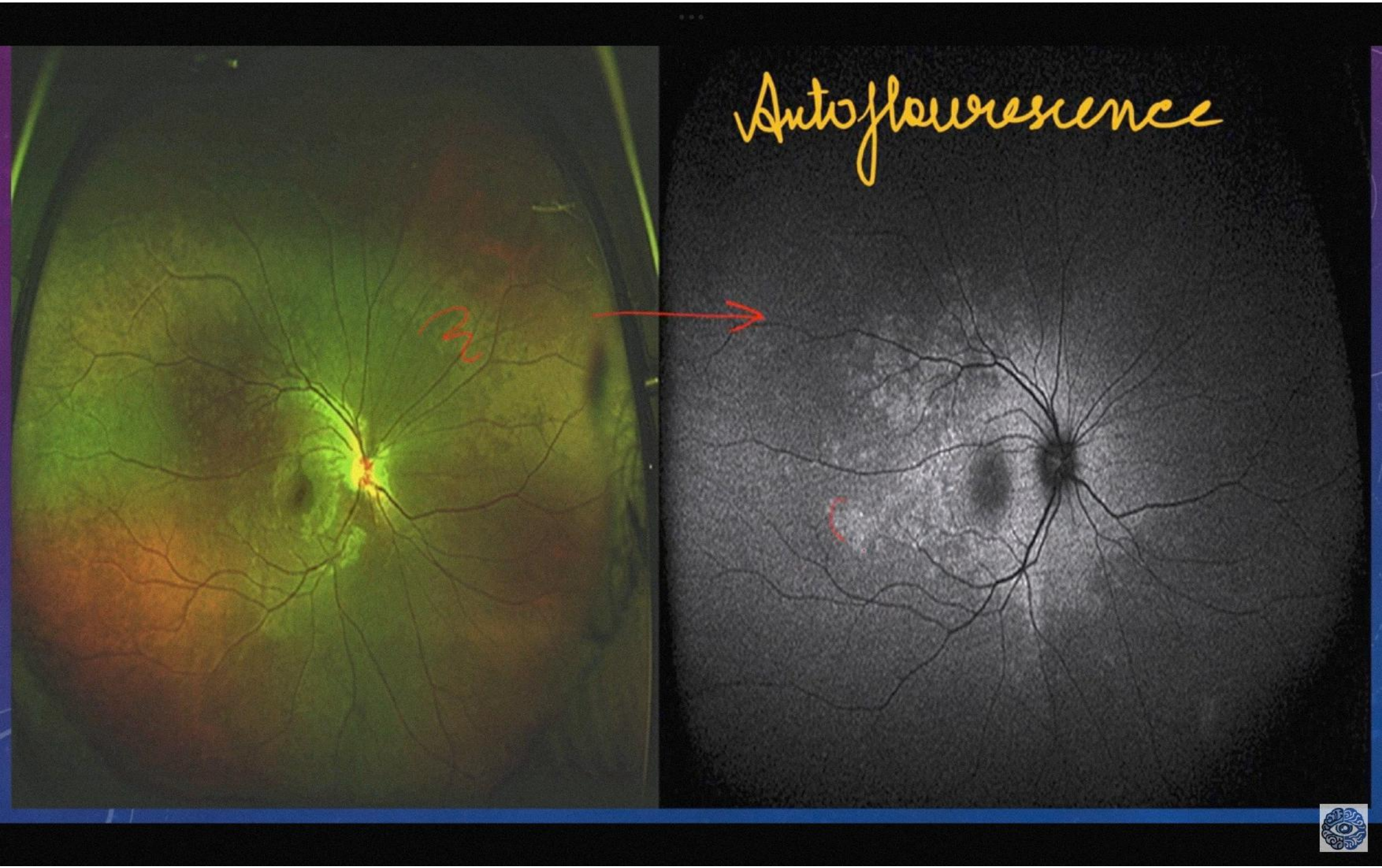
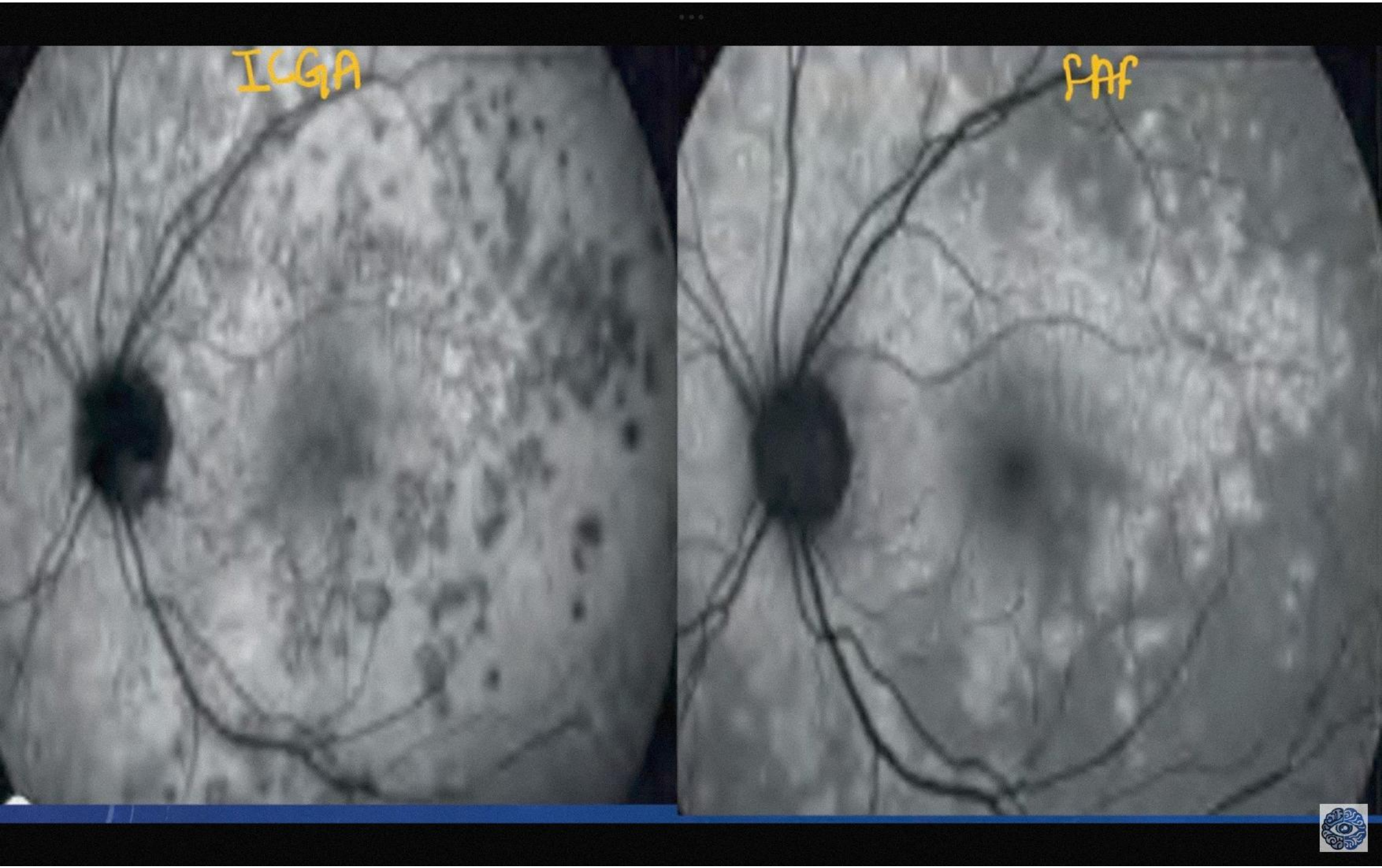



Autofluorescence


ICGA
FAF

- **Visual fields** : - The blind spot is commonly enlarged, with a temporal field defect.
- **ERG** shows a transiently **reduced a-wave amplitude.**
- Electrooculography (EOG) and visual evoked response (VER) abnormalities may be present.

TREATMENT

- This is generally not required as the symptoms and signs start to improve spontaneously in most cases by 2–6 weeks.

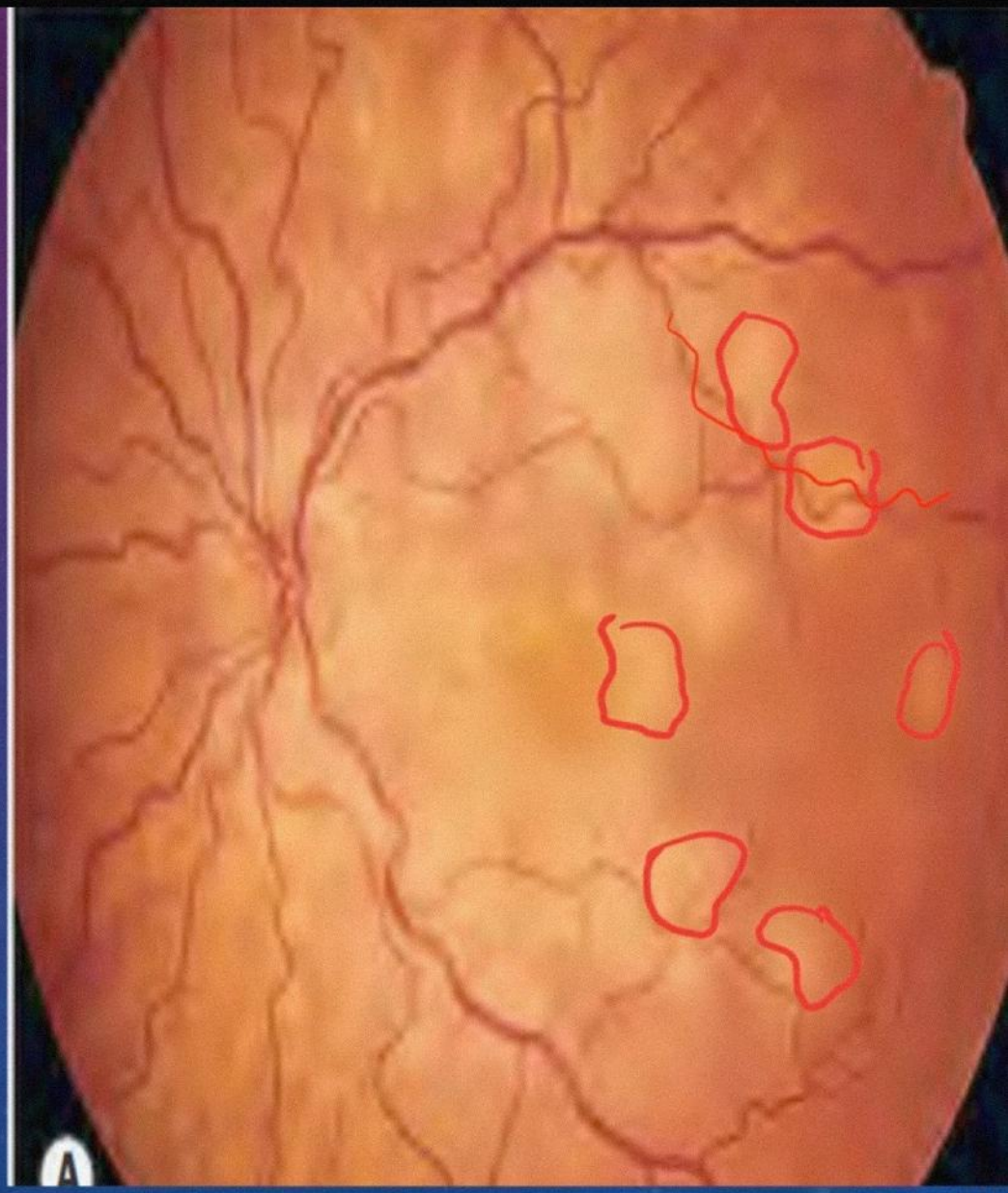
- In rare cases treatment is needed for choroidal neovascularization

APMPPE : ACUTE POSTERIOR MULTIFOCAL PLACOID PIGMENTED EPITHELIOPATHY

CLINICAL FEATURES

- APMPPE is a term first used by Gass in 1968 to describe a syndrome of **MULTIPLE LARGE PLAQUE-LIKE LESIONS** at the level of the RPE associated with temporary visual loss.
- Younger patient (age 20 to 50),
- No gender predominance,
- Viral prodrome usually present

- Bilateral and appear simultaneously or sequentially
- The characteristic clinical finding is **multiple flat yellow-white (cream-colored) plaques** at the level of the RPE .
- **Big lesion (>0.5 DD)**
- These vary in size and are clearly defined.
- The placoid lesions typically begin in the macula or posterior pole, with later-developing lesions noted more peripherally.
- The lesions do not extend beyond the equator.



SYMPTOMS

- Subacute moderate painless decrease in vision up to 20/400
- Central/paracentral scotomata and photopsia.
- The fellow eye is affected within a few days or weeks.
- **Headache** and other neurological symptoms are common and can commence many months after ocular disease onset.

SYMPTOMS

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- The fellow eye is affected within a few days or weeks.
- **Headache** and other neurological symptoms are common and can commence many months after ocular disease onset.
- Anterior uveitis and vitritis are usually very mild

PATHOGENESIS OF APMPPE

- **Cell-mediated immunity** to viral antigen.
- **Choroidal hypoperfusion hypothesis**

inflammation → hypoperfusion → ischemia of RPE & photoreceptors
of
choriocapillaries

ASSOCIATION

- **APMPPE** typically occurs in otherwise healthy persons
- Can be associated with diseases with significant **vasculitis** component eg ; **WEGENERS GRANULOMATOSIS** and **SCLERITIS**
- Can be associated with cerebral vasculitis.
- Erythema nodosum and other systemic manifestations of vasculitis have been reported.
- The clinical picture of APMPPE can be mimicked by other entities such as **SARCOIDOSIS AND TUBERCULOSIS**

ASSOCIATION

- **HLA-B7** and **HLA-DR2** are associated in a substantial proportion of patients.

RESOLUTION

- The fundus lesions resolve, they lose their cream-colored appearance.
- Older lesions may resolve while new lesions are still appearing.
- There is often a residual RPE stippling, mottling, and depigmentation

- In a subset of patients symptoms **recurs**
- In 25% visual recovery is limited to 6/15 or worse as a consequence of RPE and photoreceptor damage to the fovea

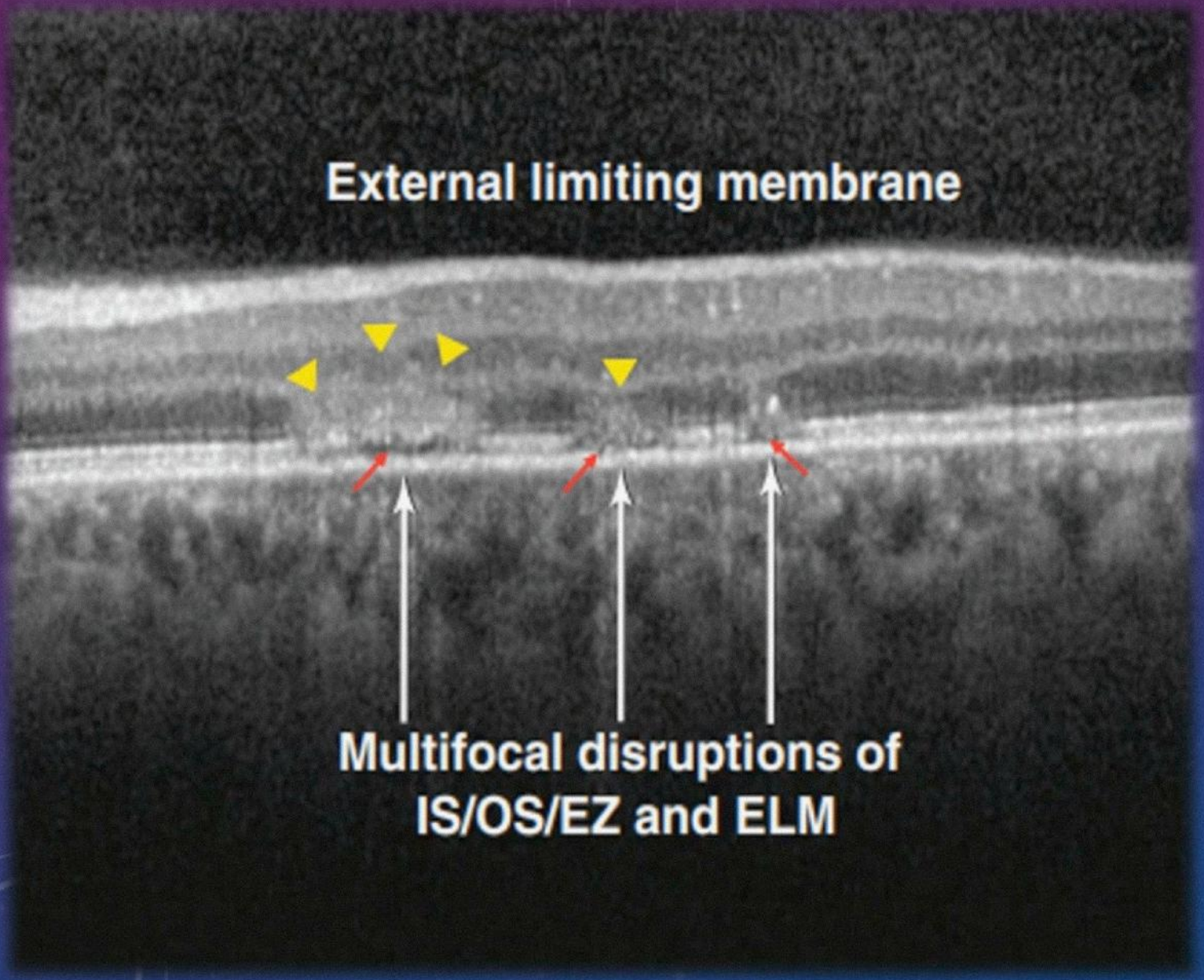
HOW TO INVESTIGATE FOR APMPPE?

INVESTIGATIONS

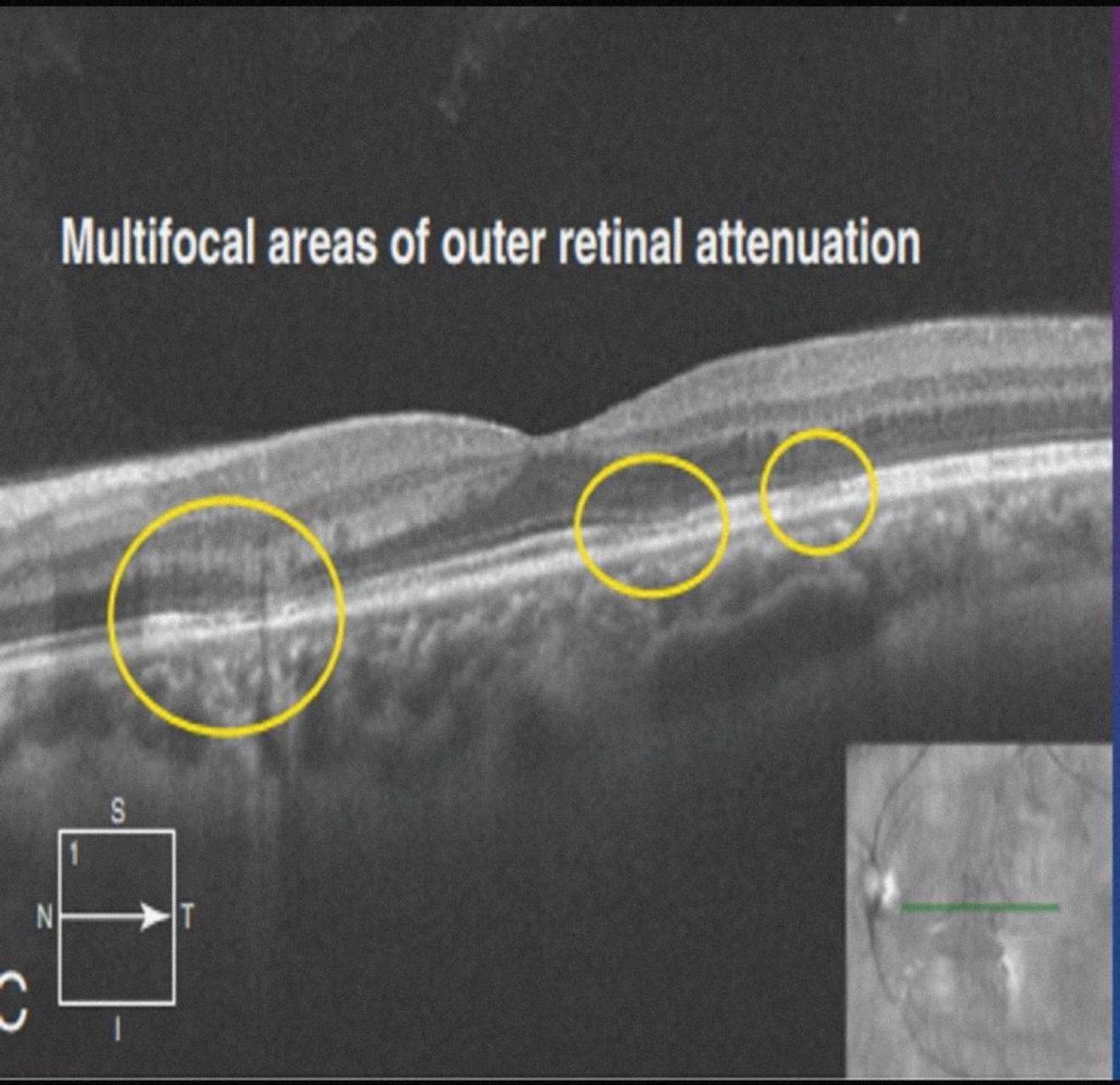
- Alternative diagnoses should be excluded.
- **OCT** of the macula.
- **FFA**
- **ICGA**
- **CNS imaging** and lumbar puncture should be performed in patients with neurological symptoms.(multifocal white lesion on MRI and CSF pleocytosis)

OCT FINDINGS

- Acute phase show **hyperreflectivity** & involvement from the outer nuclear layer to to the RPE, which includes the photoreceptors.
- OCT findings in the resolution phase may normalize or leave permanent defects in the **outer nuclear layer, IS/OS/EZ, and RPE**
- *Atypical OCT findings* include intraretinal fluid, significant subretinal fluid, and localized thickening of the inner retina



Multiple focal disruptions of the retinal pigment epithelium (RPE), IS/OS/EZ, and external limiting membrane (ELM), with overlying hyperreflectivity of the outer retina limited to the outer nuclear layer and outward layers



MEWDS shows focal irregularities in the external limiting membrane (ELM) and the ellipsoid zone (EZ)

APMPPE shows a hyperreflective area above the RPE corresponding to the placoid lesions, with disruption of the outer retina and rarely the presence of subretinal or intraretinal fluid

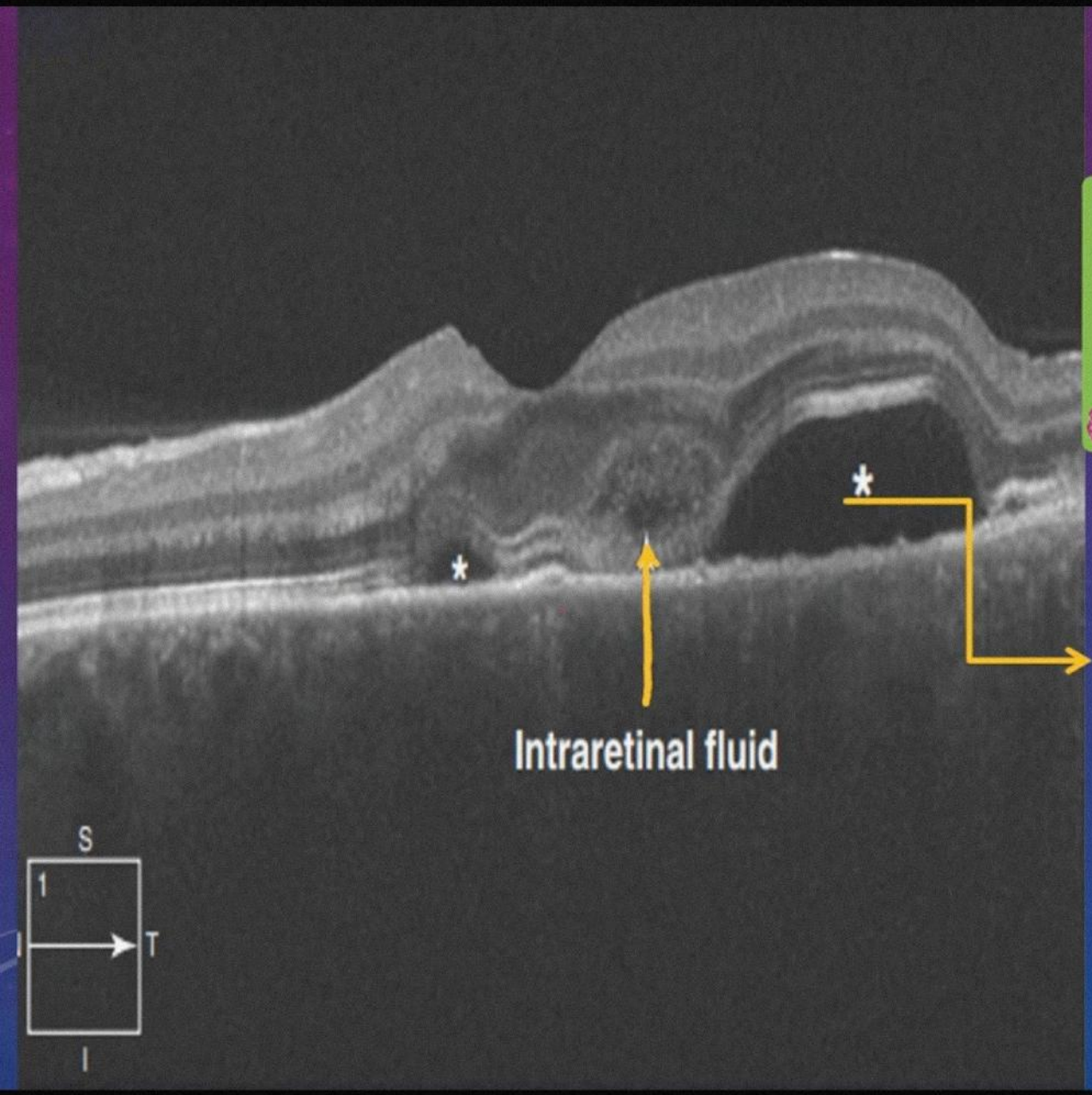


FIG. 3. Atypical case of APMPPE shows significant subretinal fluid *(asterisks)* and localized cystic thickening of the outer retina. These features can resemble findings in Vogt-Koyanagi-Harada syndrome.

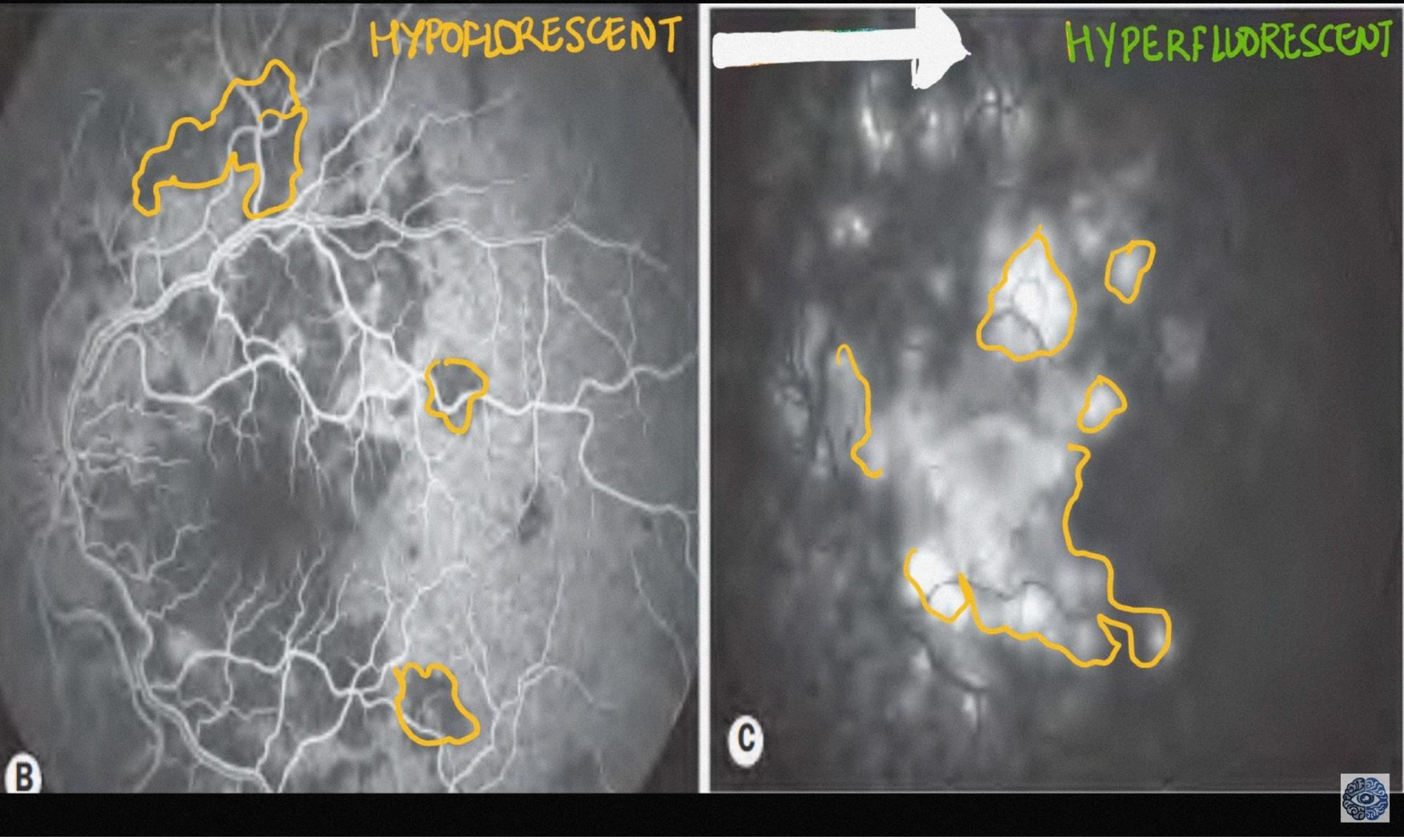
Subretinal fluid

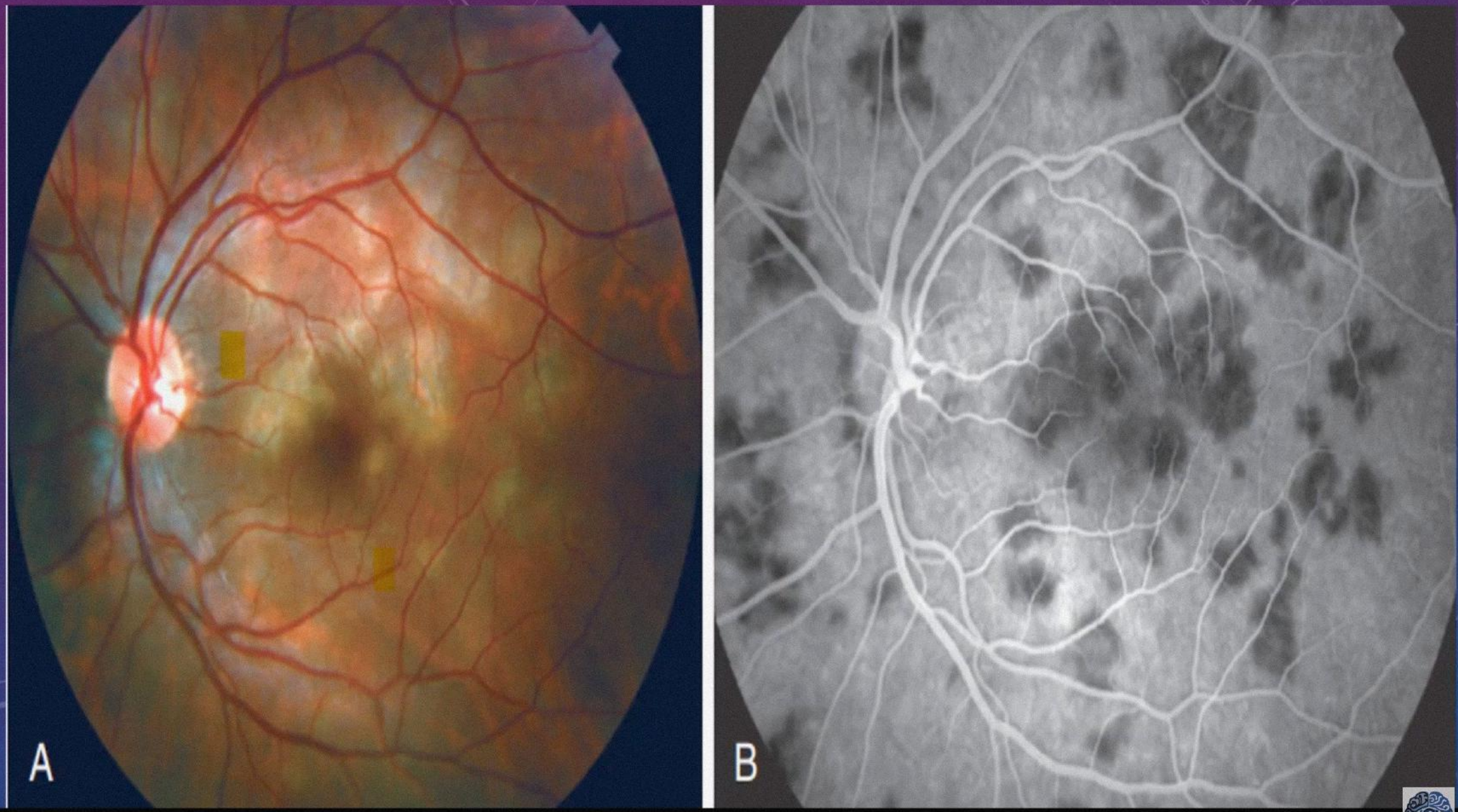
FFA

- **Acute stage** of the disease, during which there are active cream-colored lesions, the choroidal fluorescence is blocked in the early phases of the angiogram.
- In the late stages there is staining of the lesions that had previously blocked fluorescence.

- **HYPOFLUORESCENCE FOLLOWED BY HYPERFLUORESCENCE** in combination with appropriately sized and colored fundic lesions is diagnostic of APMPPE.

- **Resolved stage** demonstrate transmission defects in the RPE without leakage.

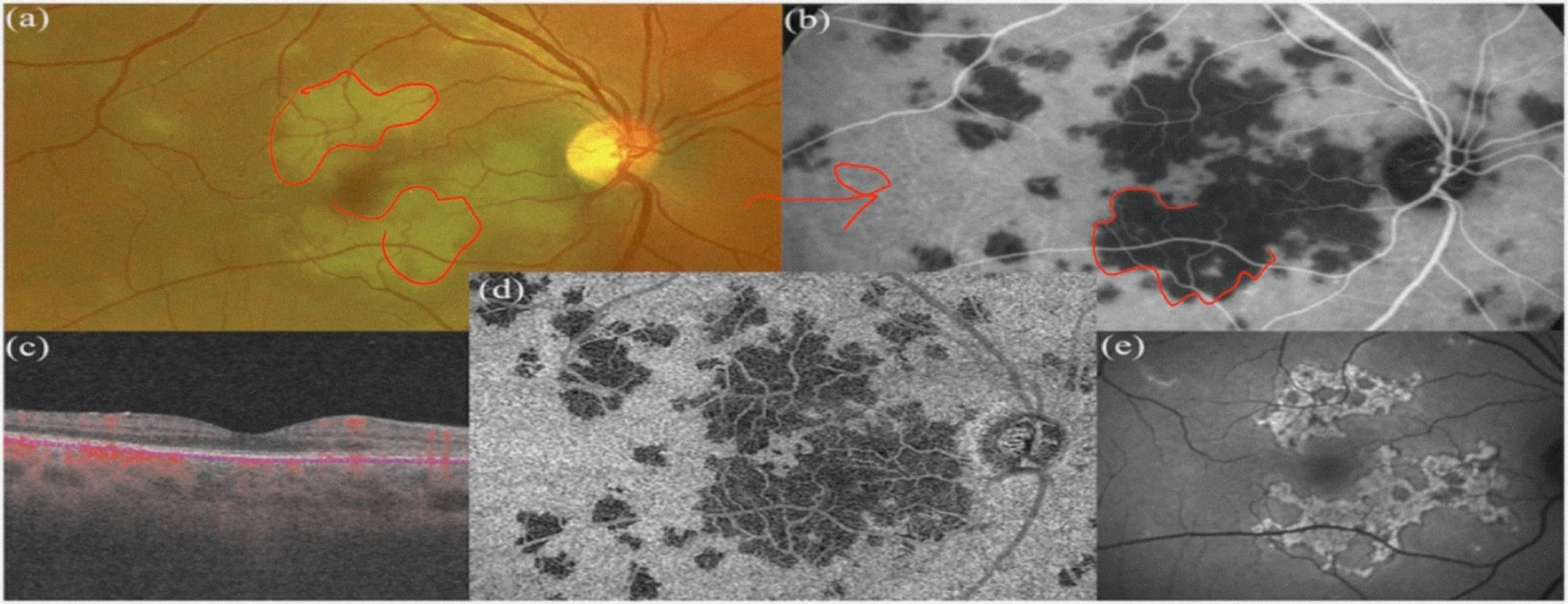

HYPOFLORESCENT
B
HYPERFLUORESCENT
C


A
B

ICGA

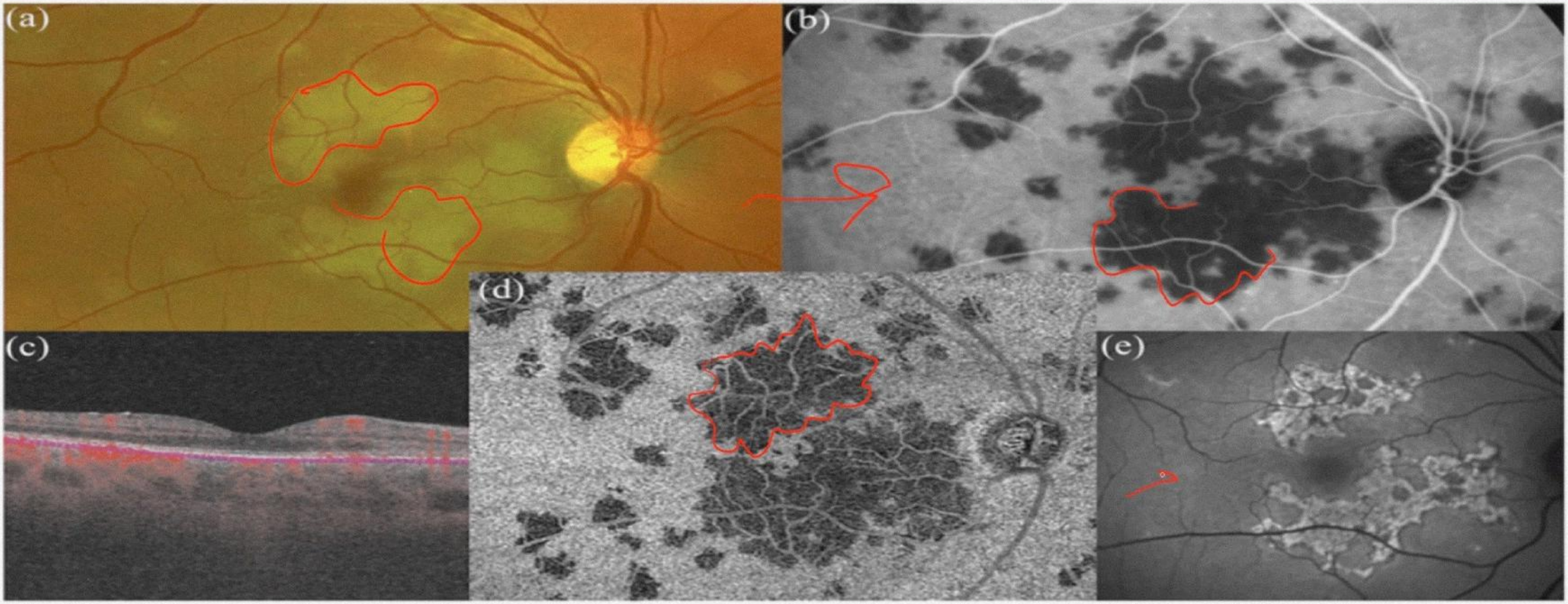
HYPOCYANESCENT LESIONS

Imaging	APMPPE	MEWDS
FA	Early hypofluorescence, late hyperfluorescence, and staining; window defects in the quiescent stage	Early and late hyperfluorescence of the white dots in a wreathlike pattern. May have optic nerve leakage
OCT	Hyperreflective area above RPE with disruption of ELM and EZ corresponding to the placoid lesions Rare: presence of subretinal or intraretinal fluid	Focal irregularities in ELM and EZ, focal thinning of the ONL, usually resolves.



Multimodal imaging of acute posterior multifocal placoid pigment epitheliopathy (APMPPE). In the active phase, (a) fundus photos show multiple yellowish placoid lesions, (b) hypocyanescent on indocyanine green angiography, while (c and d) OCT angiography well delineates the dark areas of choriocapillaris hypoperfusion. Ten days after presentation, (e) fundus autofluorescence shows hyper-autofluorescent lesions.

Credit - Researchgate.



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TREATMENT

- Treatment is not usually required, but steroids should be considered for patients with macular involvement.
- Steroids and possibly even IMMUNOSUPPRESION may be given for cerebral vasculitis.
- Patients with neurological symptoms definitely needs treatment .