10th Evolving Practice Of Ophthalmology Middle East Conference
EPOMEC
EXPANDING VISION

Vascular Tango: A rare association of paracentral acute middle maculopathy

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No financial disclosures

Aim:

- To report a rare association of paracentral acute middle maculopathy (PAMM) in an adult female.

History & Chief Complaints

Sudden onset scotoma in left eye x few hours

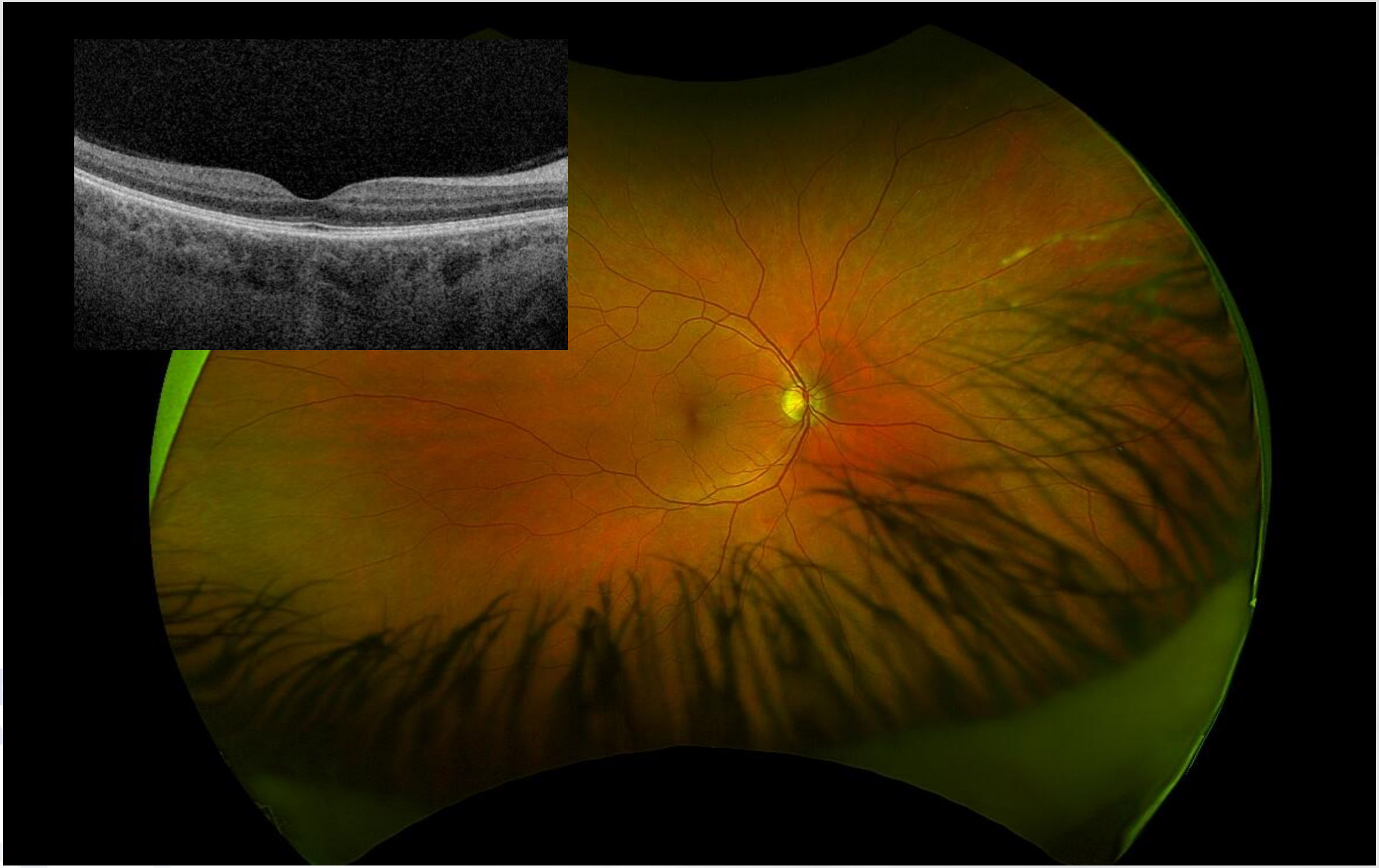
Visual Acuity

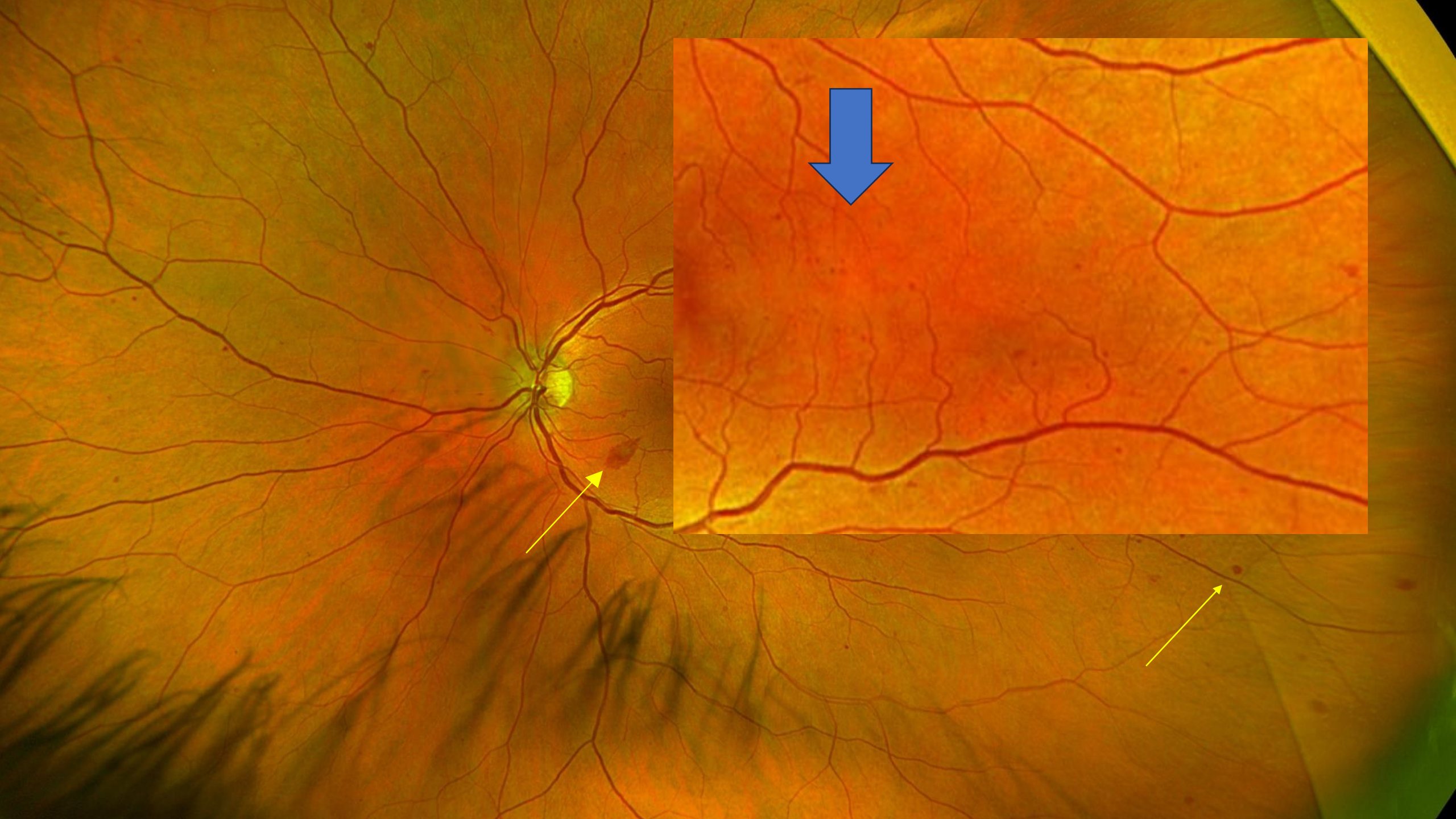
	OD	OS
Uncorrected visual acuity	20/20	20/20
Near vision	N6	N6

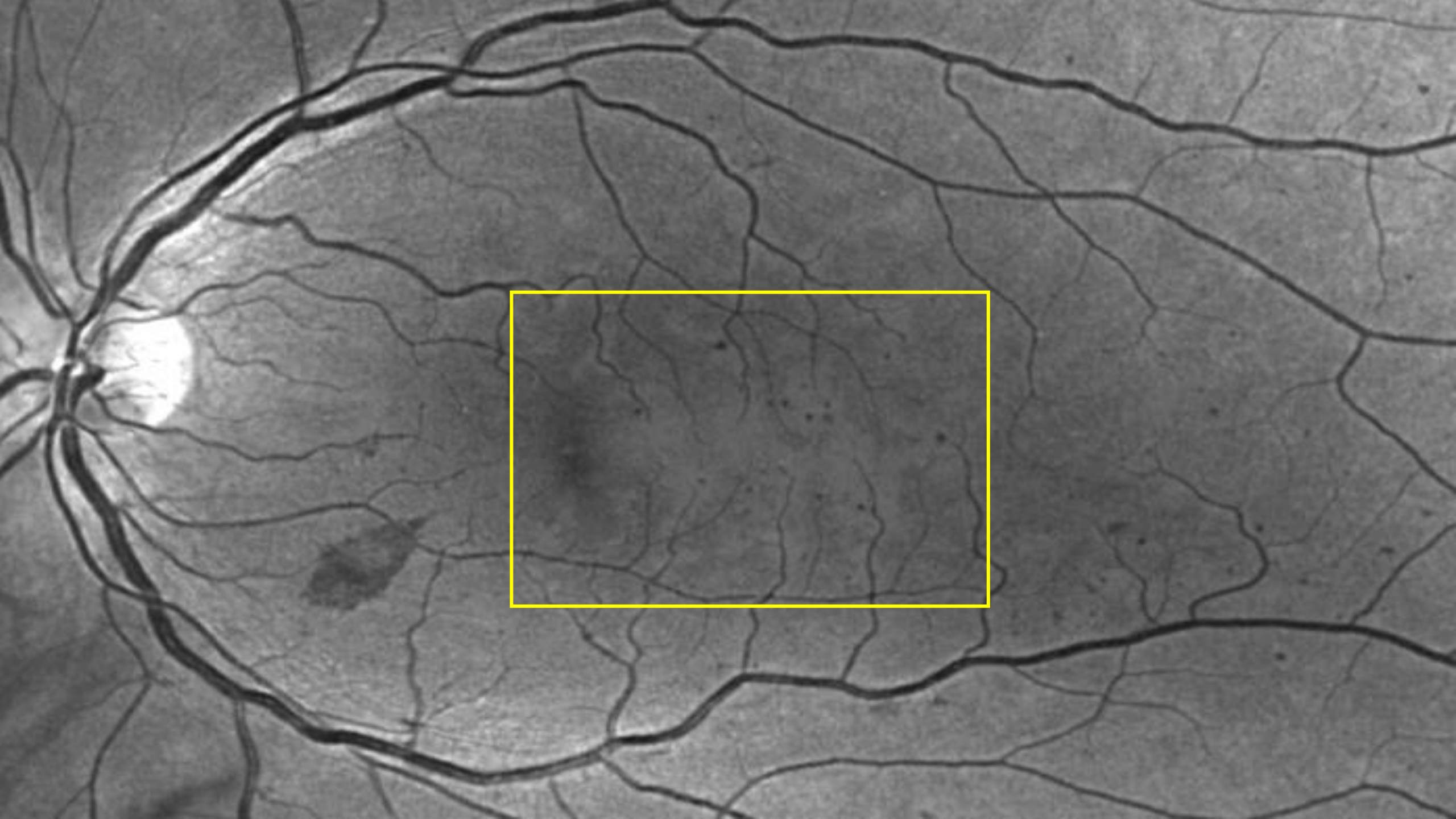
Anterior Segment Examination

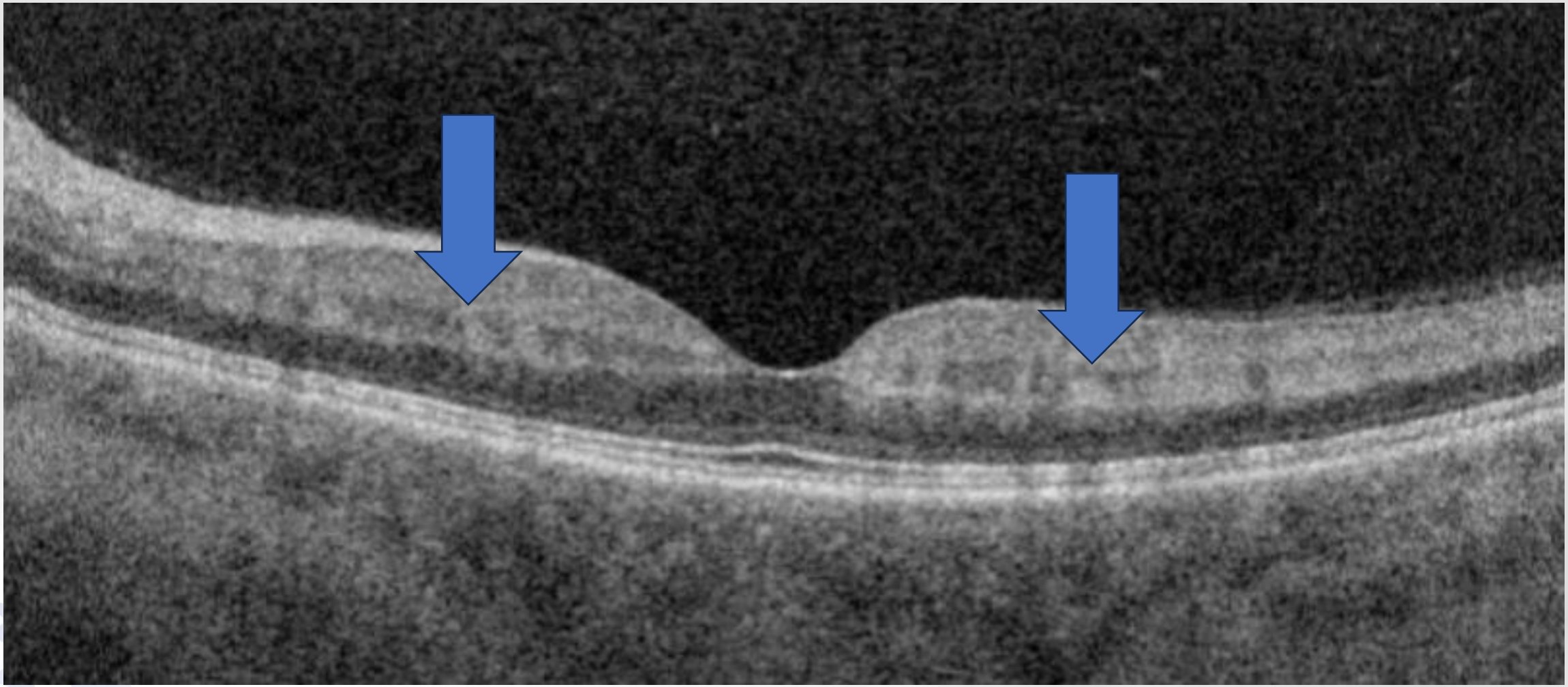
	OD	OS
Lids	Normal	Normal
Conjunctiva	Normal	Normal
Cornea	Clear	Clear
Sclera	Normal	Normal
Anterior Chamber	Quiet and Deep	Quiet and Deep
Iris	Normal	Normal
Lens	Clear	Clear
Anterior Vitreous	Normal	Normal

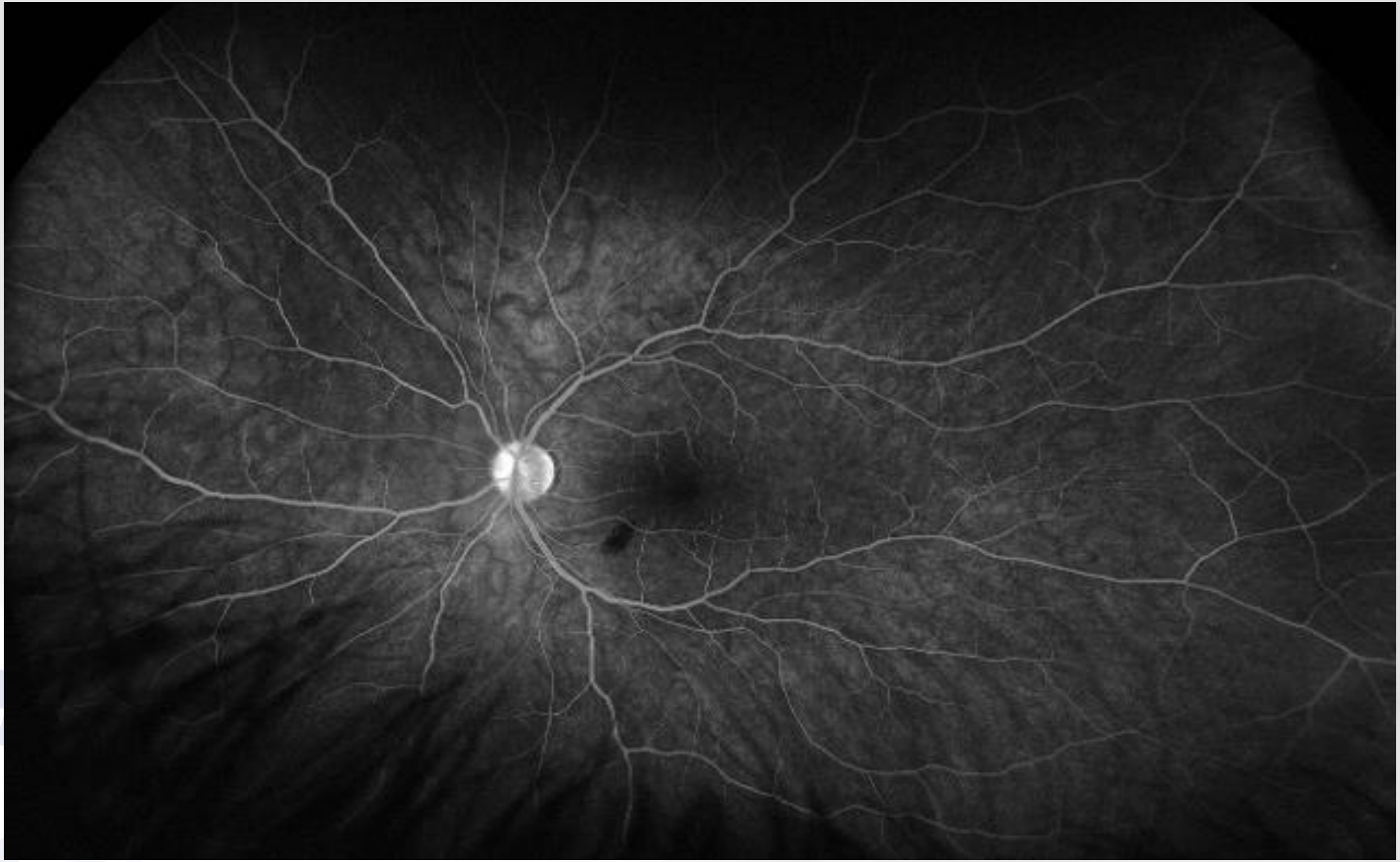
No evidence of anterior inflammation

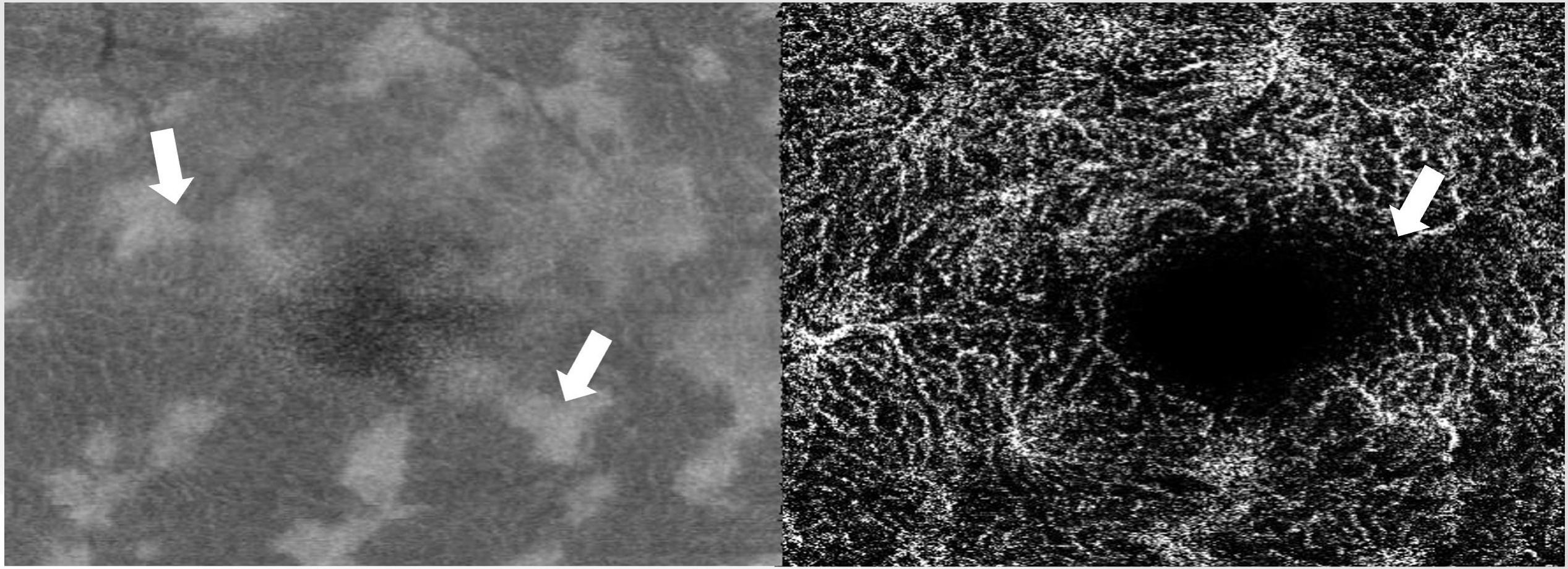












Probable Diagnosis

Paracentral acute middle maculopathy (PAMM) secondary to possible underlying systemic vasculitis

Going back in History

History of painful rashes on hand and arms associated with arthralgias

Deep painful subcutaneous rashes and nodules, painful finger tips, stiffness of the fingers and joint pain

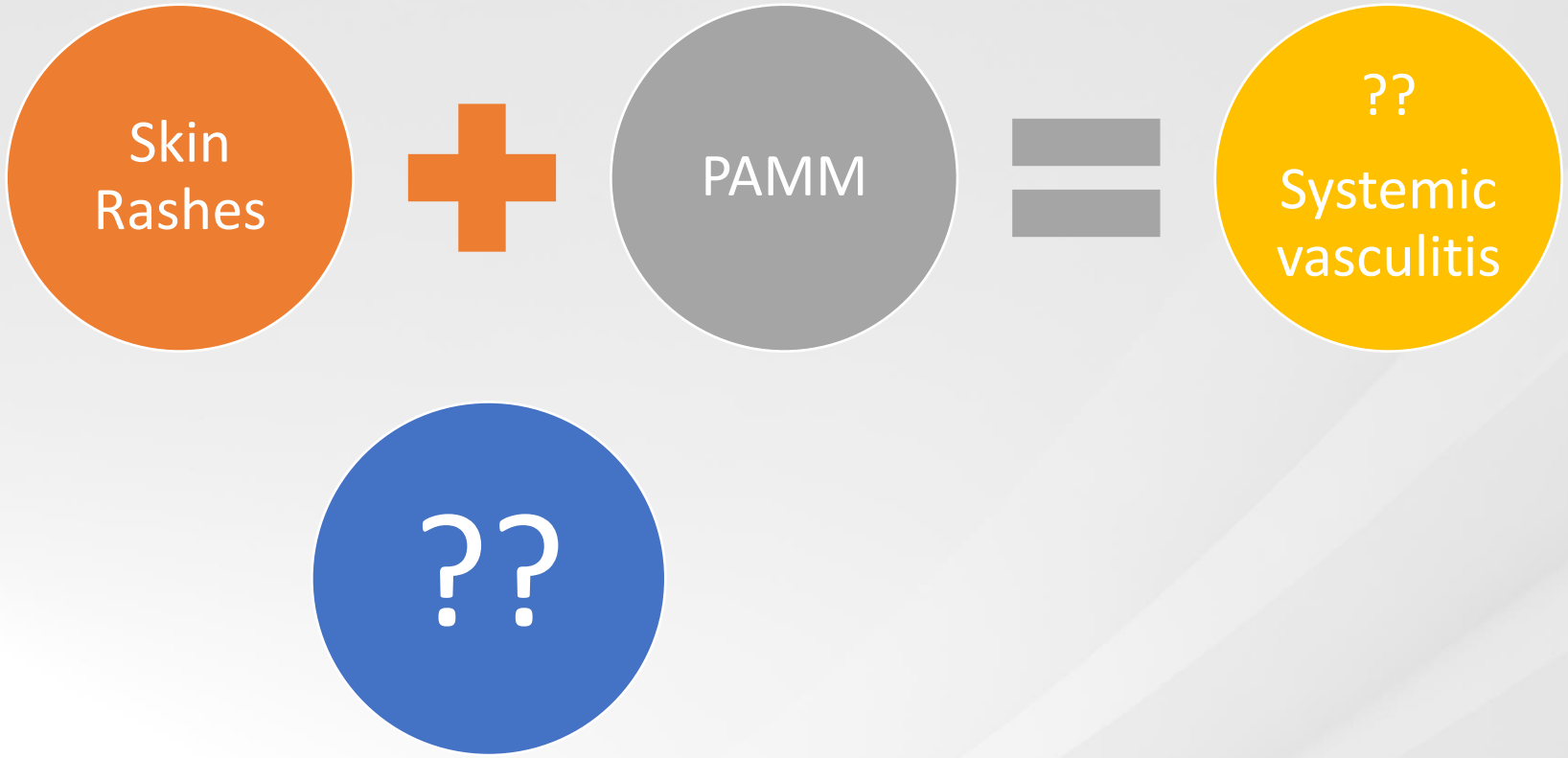
Consulted rheumatologist for the same

Rheumatology work up normal except non specific elevation of PML/SSC

Classified as undifferentiated connective tissue disorder

Started on oral steroids 20 mg/day for management of symptoms

Going back in History



Previous Health Records

Gynecologist Consultation

- [ ] Vaginal ulcers

Dentist Consultation

- [ ] Aphthous ulcers

Recurrent for past 1 year

Diagnosis


Recurrent oral and vaginal ulcers
Greek Ethnicity
Rashes
??
PAMM s/o underlying retinal vasculitis

Criteria for Behcet's Disease (BD)

Criterion	Required features
Recurrent oral ulceration	Aphthous ulceration with at least three episodes in any 12-month period
Plus any two of the following:	
Recurrent genital ulceration	Aphthous ulceration or scarring
Eye lesions	Anterior or posterior uveitis, cells in vitreous on slit-lamp examination or retinal vasculitis
Skin lesions	Erythema nodosum-like lesions; papulopustular skin lesions or pseudofolliculitis with characteristic acneiform nodules
Pathergy test	Papule 2 mm or more in size, developing 24—48 h after pricking the skin with a 20—25-gauge needle

Management

Started on oral Azathioprine 50 mg/day, oral steroids continued

HLA B 51 positive

Discussion

- Ocular involvement in BD:

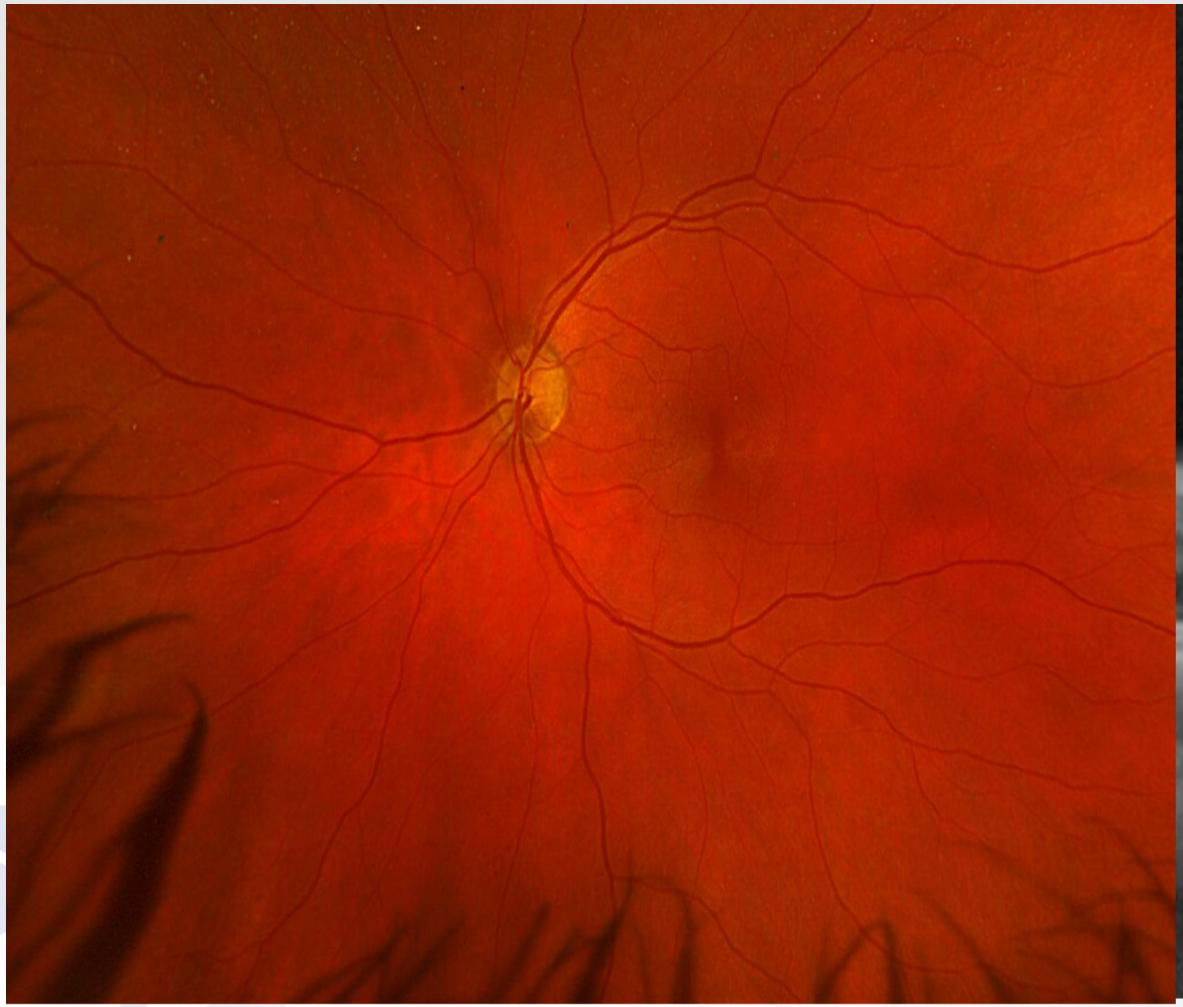
- Anterior uveitis with hypopyon

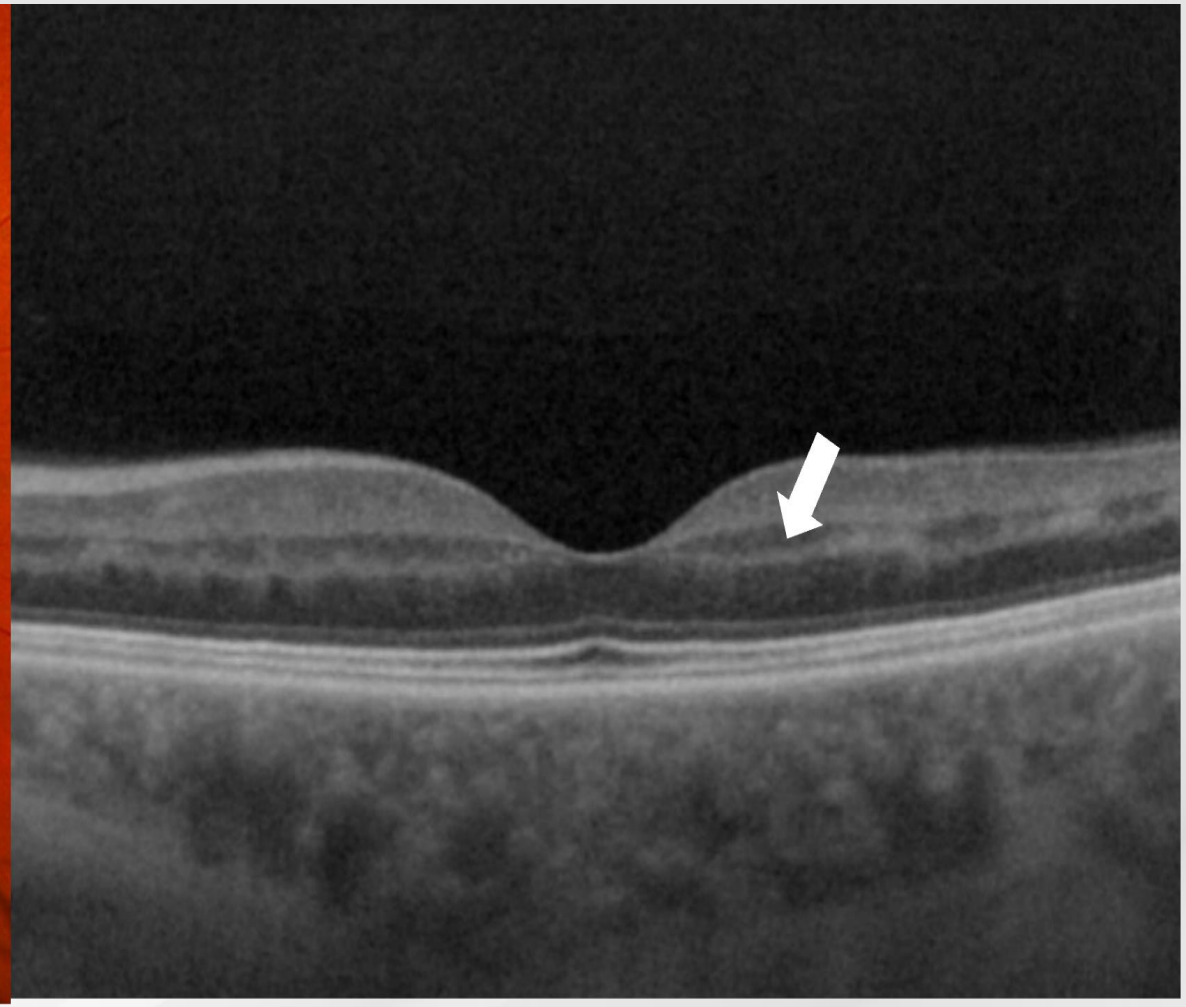
- Retinal vasculitis

- PAMM is an atypical manifestation which is very rare
- It has been reported previously but the patient also had nongranulomatous keratic precipitates and bilateral retinal phlebitis *
- This case is unique as PAMM was the sole ocular manifestation of BD.

* Song, D., Choi, D. J., & Bhatt, N. (2022). Paracentral Acute Middle Maculopathy in Behçet's Disease. *Retinal Cases and Brief Reports*.

Follow-Up: 3 weeks





Conclusion

- While the association of PAMM with underlying vasculitis is recognized, its occurrence in Behcet's disease is exceedingly rare.
- Apart from standard diagnostic methods like FFA, additional imaging techniques, such as OCT and red-free imaging, are recommended for early identification of atypical presentations like PAMM in Behçet's disease.