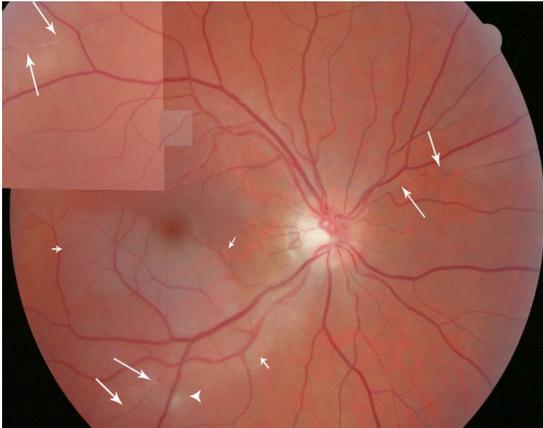


Syndrome de Susac

Small Infarctions of Cochlear Retinal and Encephalic Tissue
Retinopathy Encephalopathy and Deafness - Microangiopathy

Le point de vue de l'ophtalmologiste



Dominique Monnet



Hôpital Cochin –
Hôtel Dieu



Université
Paris
Descartes

Susac syndrome

- Rare mais sous diagnostiqué
- Prévalence : 0.148 per 100,000 (Europe centrale)
- Occlusions petits artères :
 - Triade : Encéphalopathie aiguë (72%), Surdit  (20%), OBAR (24%).
 -  tiologie inconnue : Endoth liopathie immune.
- Age : 7   77 ans : moyenne 30.5 ans
- H < F , M/F Ratio 1 : 3.5
 - plus s v re Hommes
 - 12 publications Susac contexte grossesse

Susac

Evidence Based Medicine ?

≥ 100 « *case reports* »

Case series of Susac syndrome.

Author	Year	Number of cases	Initial clinical presentation	% of patients with full triad at presentation	% of patients with full triad at the end of follow-up	Final outcome
Jarius et al ^[15]	2014	25	Encephalopathy (72%) Hearing loss (20%) Visual disturbances (24%)	0	80	Neurological deficits (64%) Auditory deficits (84%) Visual deficits (75%)
Mateen et al ^[17]	2012	29	Encephalopathy (90%) Hearing loss (83%) Visual disturbances (83%)	55	Not reported	Not reported
Hung do et al ^[30]	2004	4	Encephalopathy (100%) Hearing loss (100%) Visual disturbances (100%)	100	No change	Neurological deficits (50%) Visual deficits (25%)
Snyers et al ^[31]	2006	4	Encephalopathy (75%) Hearing loss (100%) Visual disturbances (100%)	75	No change	Neurological deficits (25%)

Triade

- Présentation initiale : 13 à 20% des cas
- Diagnostic difficile :
 - Suspecté :

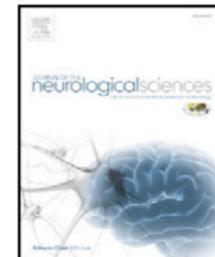
1 manifestation triade + Absence de FDR pour artériosclérose ou coagulopathie + + 1 FDR maladie Femme 20-40 ans, ≤ 1 an grossesse, IRM+ corps calleux ou lésions périventriculaires
 - Incomplet : 2 manifestations / triade
 - Complet : Triade



Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns



Short communication

Skin involvement in Susac's syndrome

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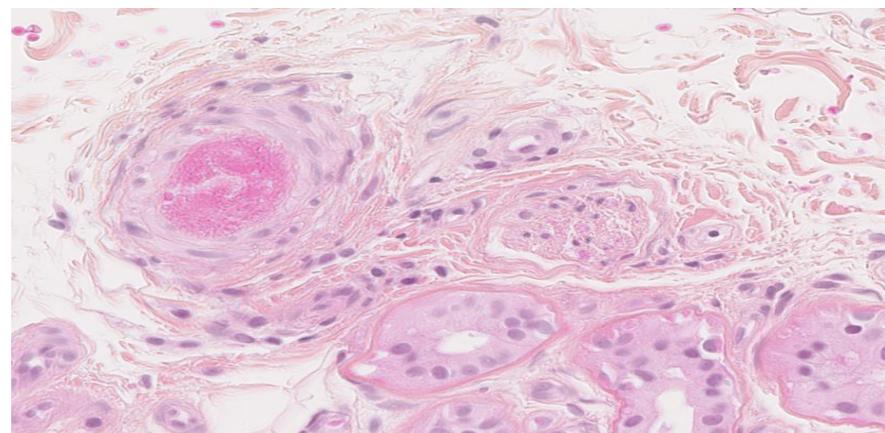
^b Université Paris Descartes, Department of Ophthalmology, Hôpital Cochin, APHP, Paris, France

^c Université Paris Descartes, Department of Dermatology, Hôpital Cochin, APHP, Paris, France

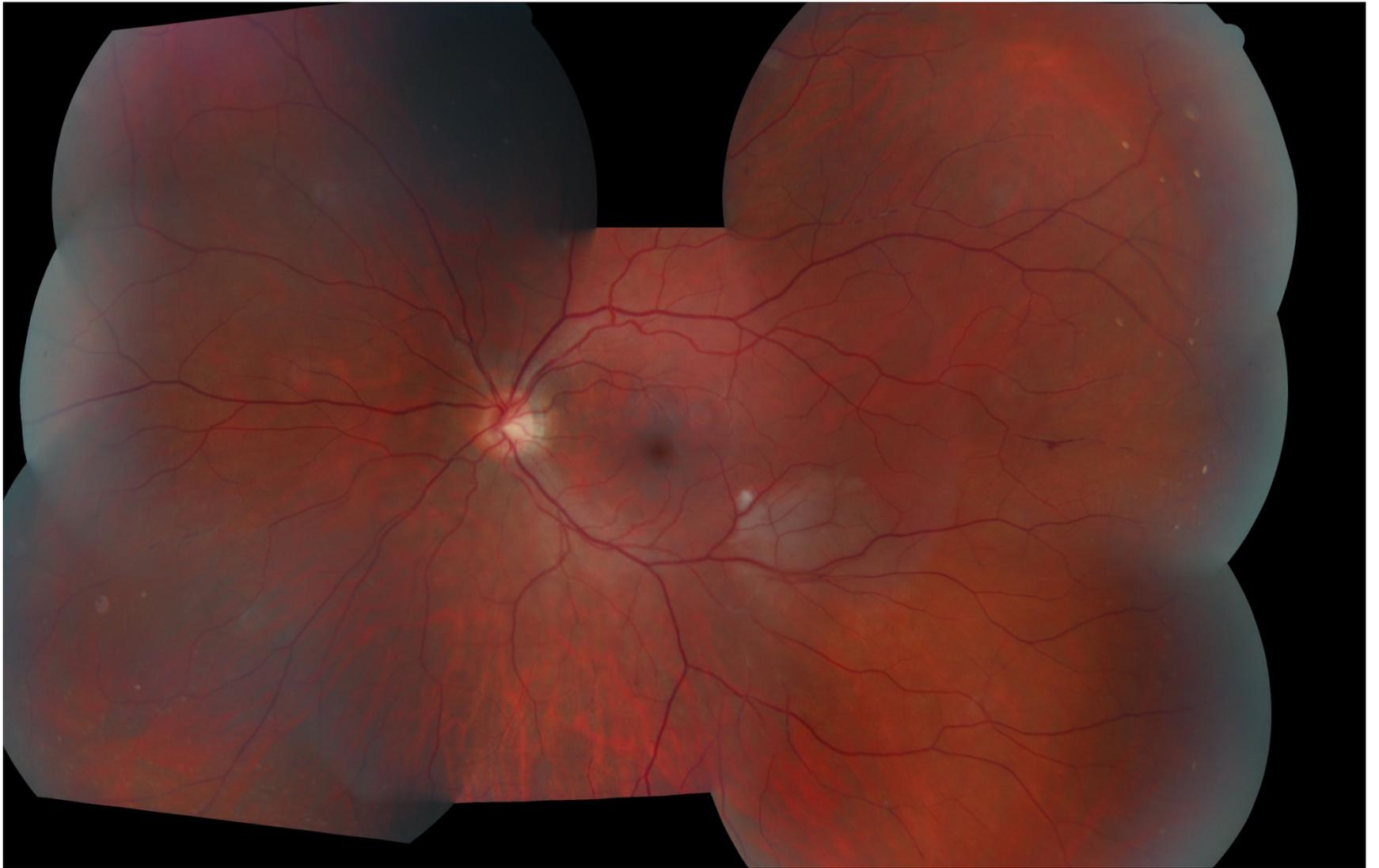
^d Université Paris Descartes, Department of Pathology, Hôpital Cochin, APHP, Paris, France

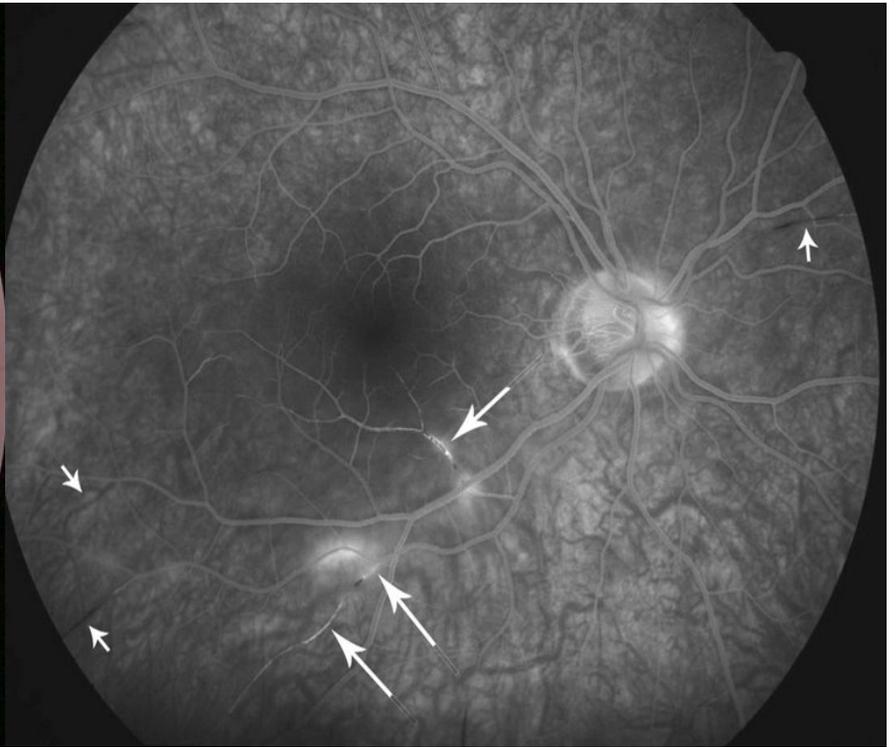
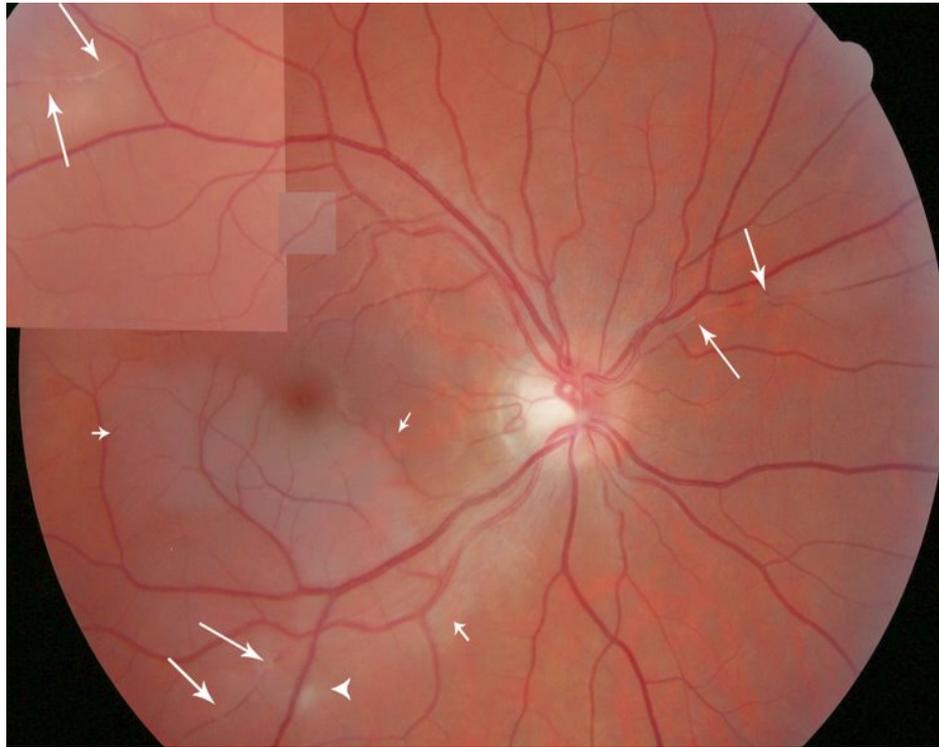


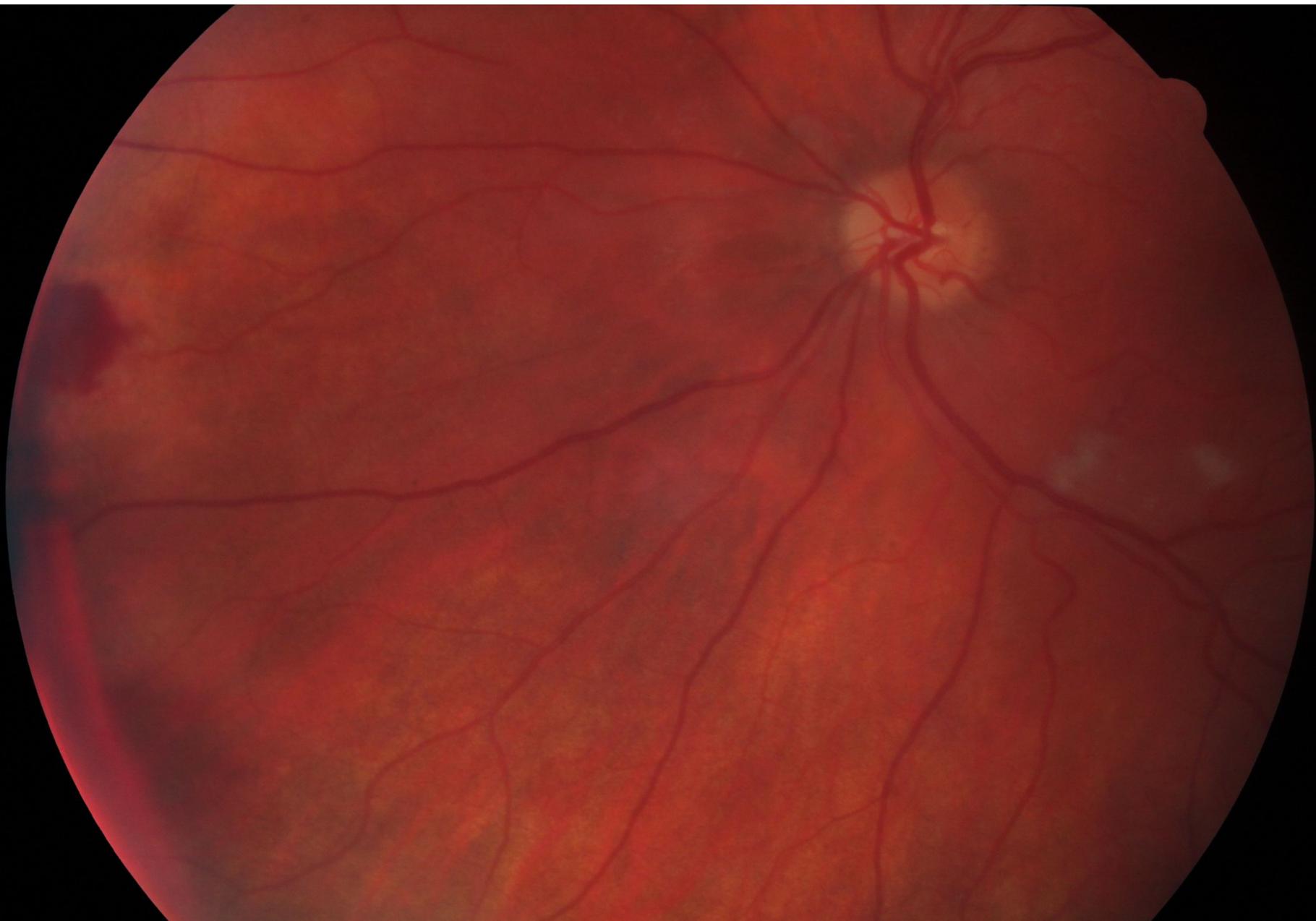
A – Livedo racemosa (right foot)

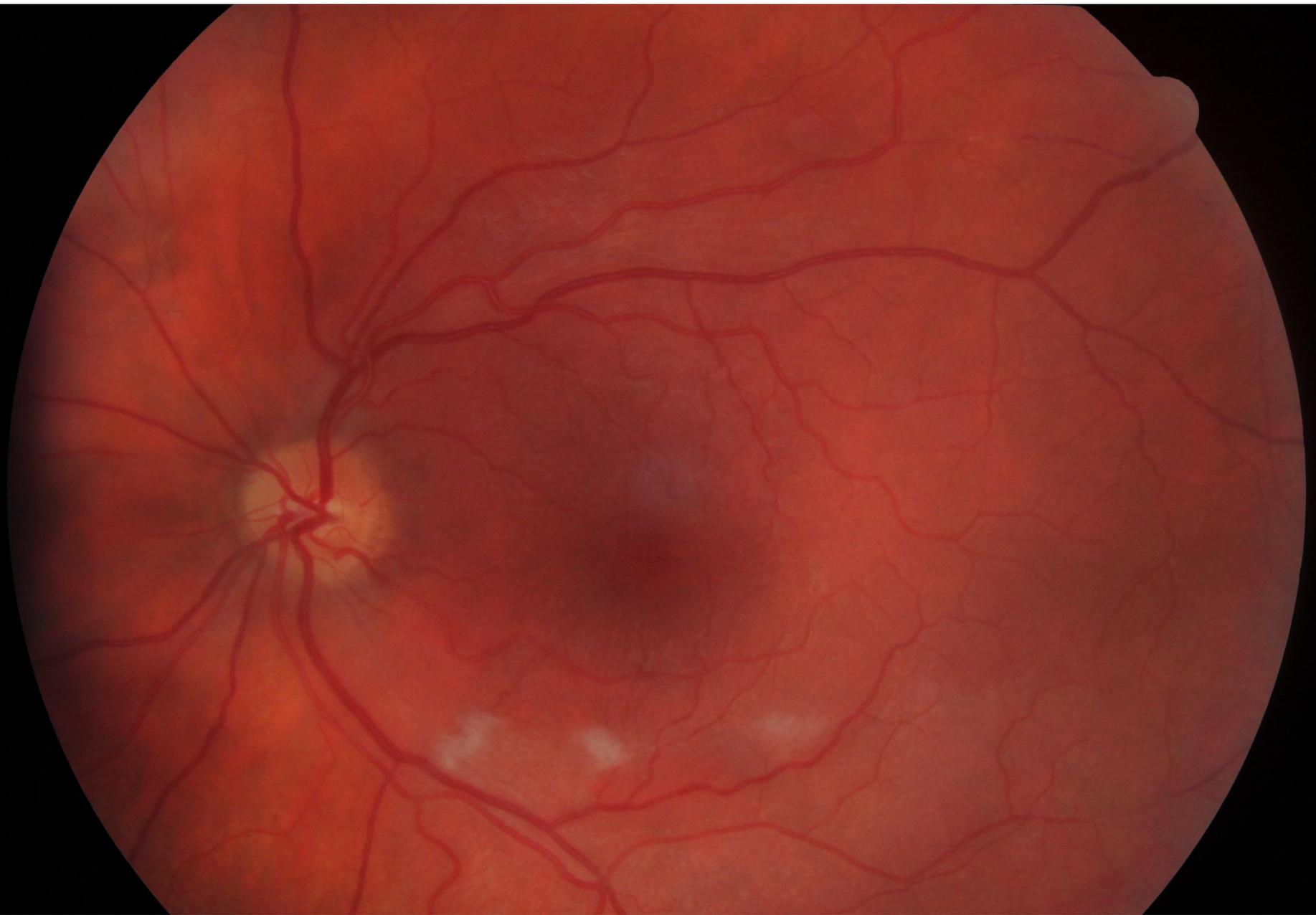


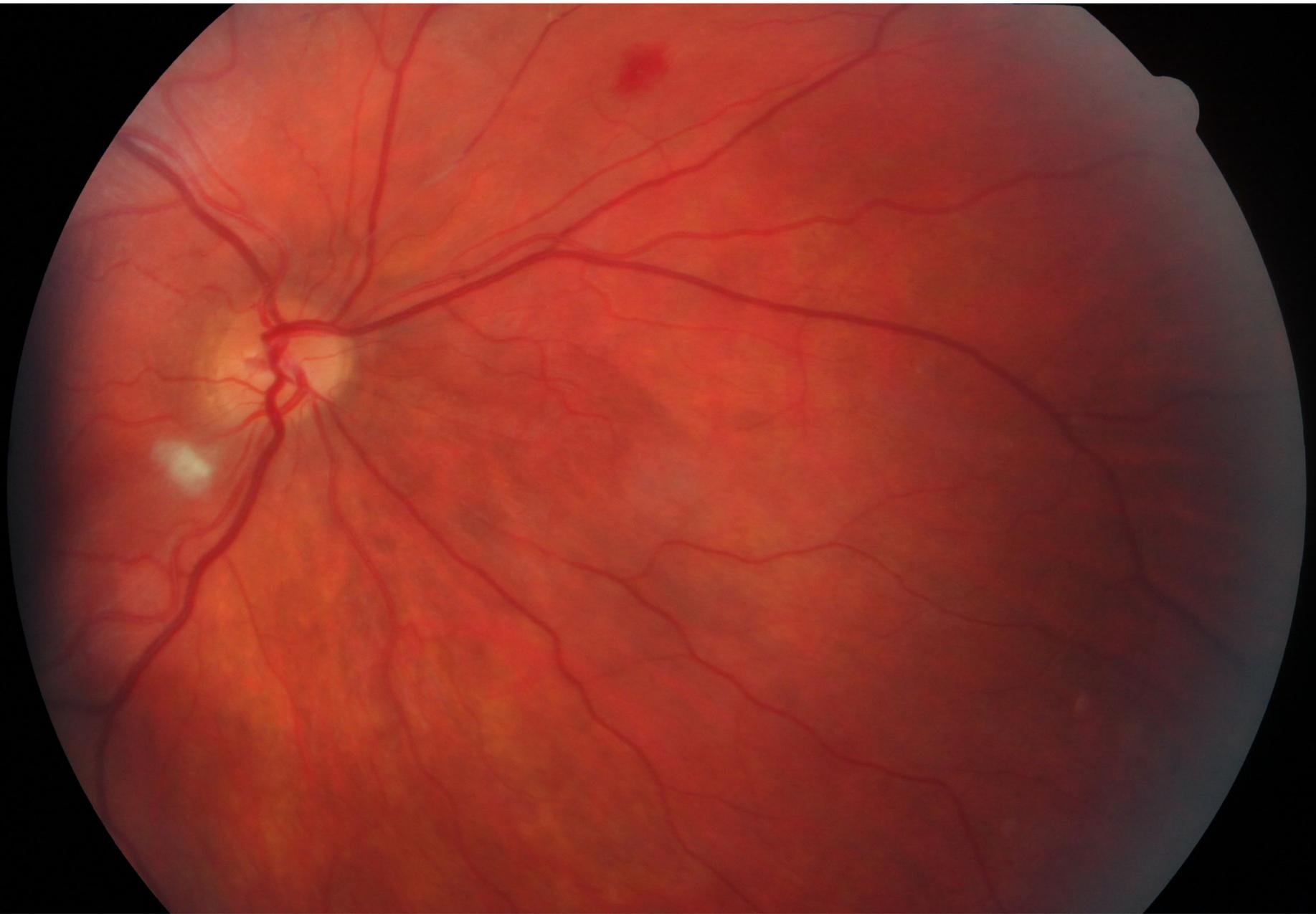
B – Skin biopsy (HES) : Occlusion of dermal peri-sudoral vessel without signs of inflammation

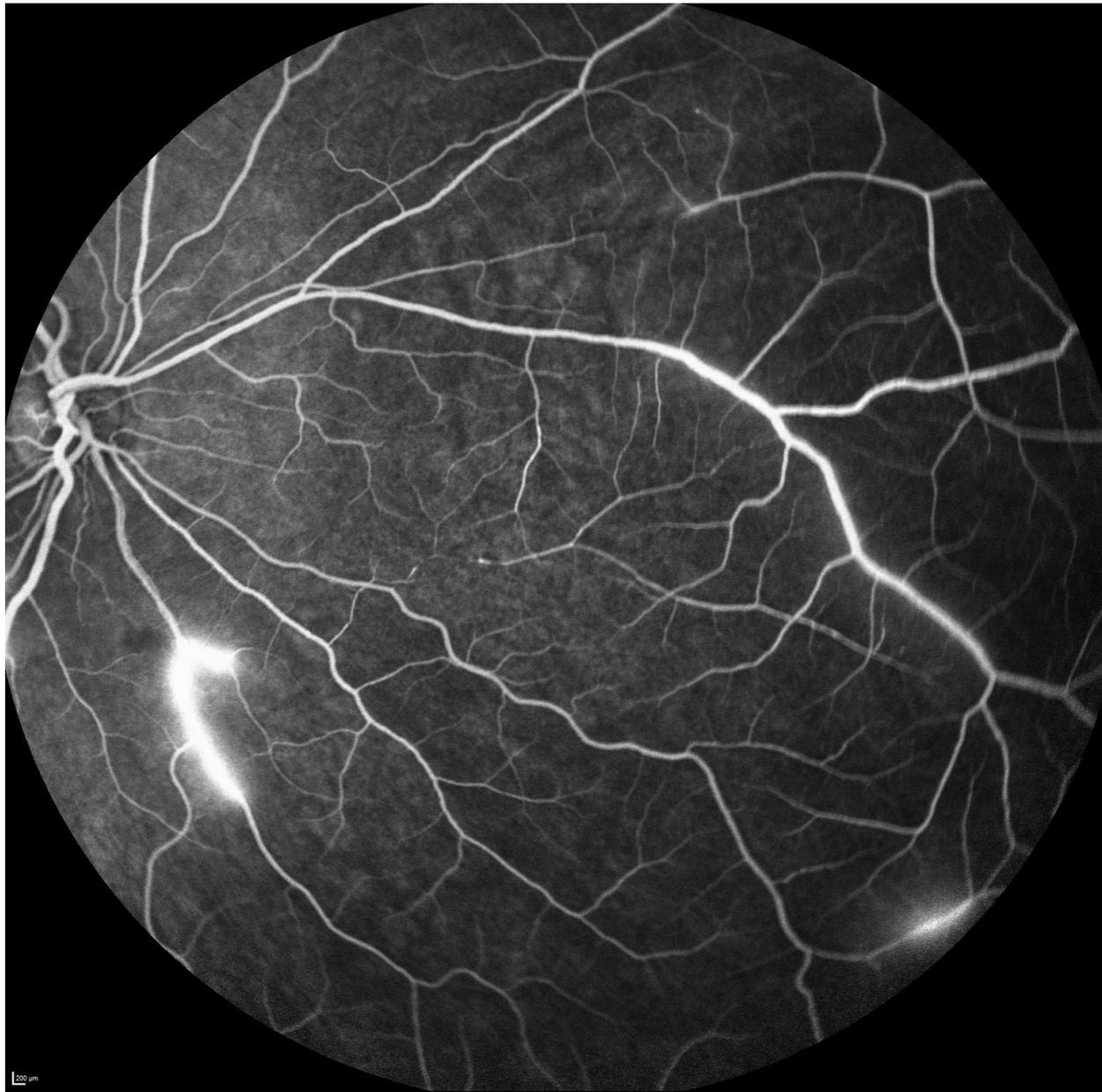












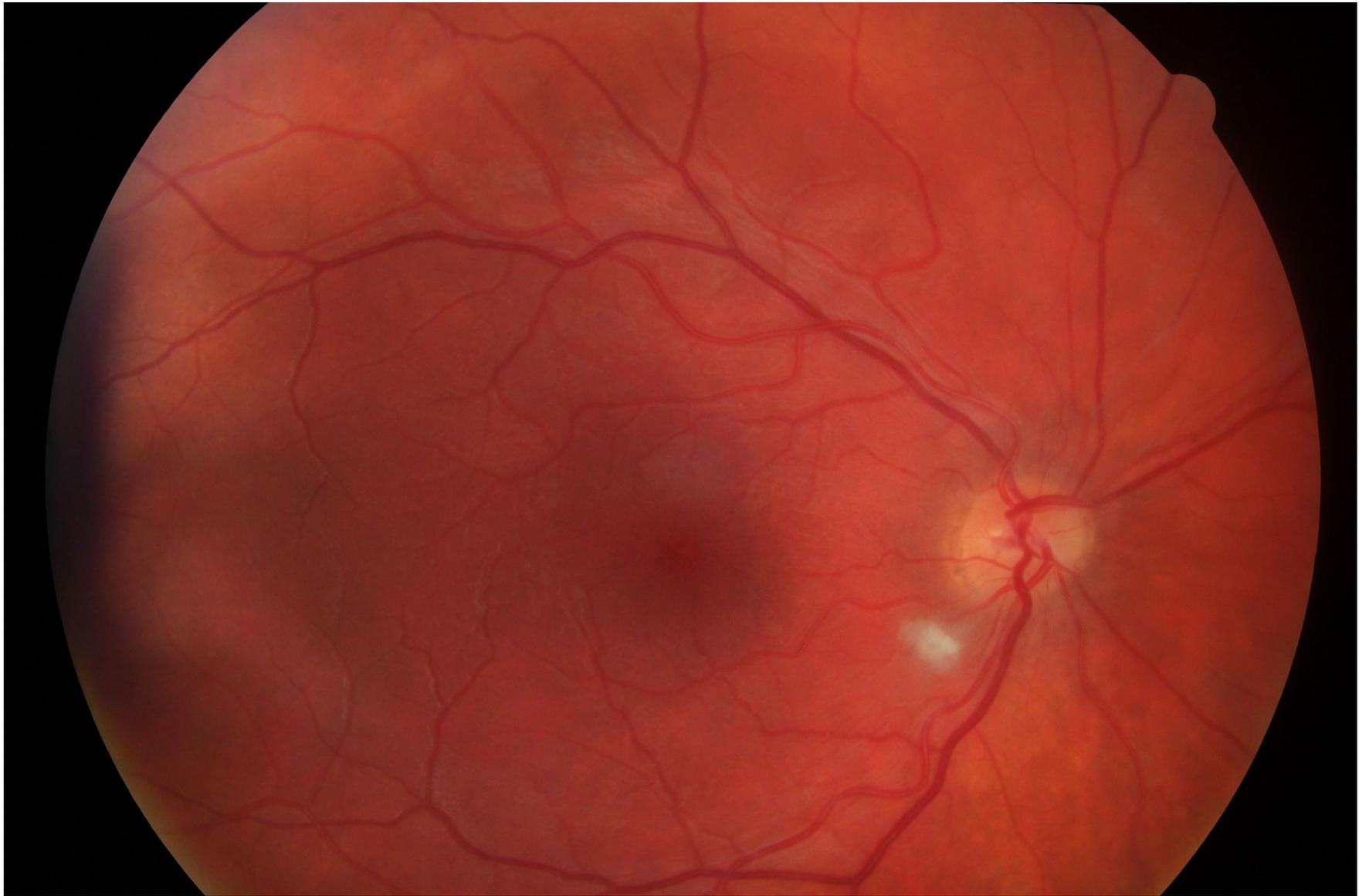
200 µm

22/07/2016, OD
FA 6:30.29 55° ART(5) [HR]



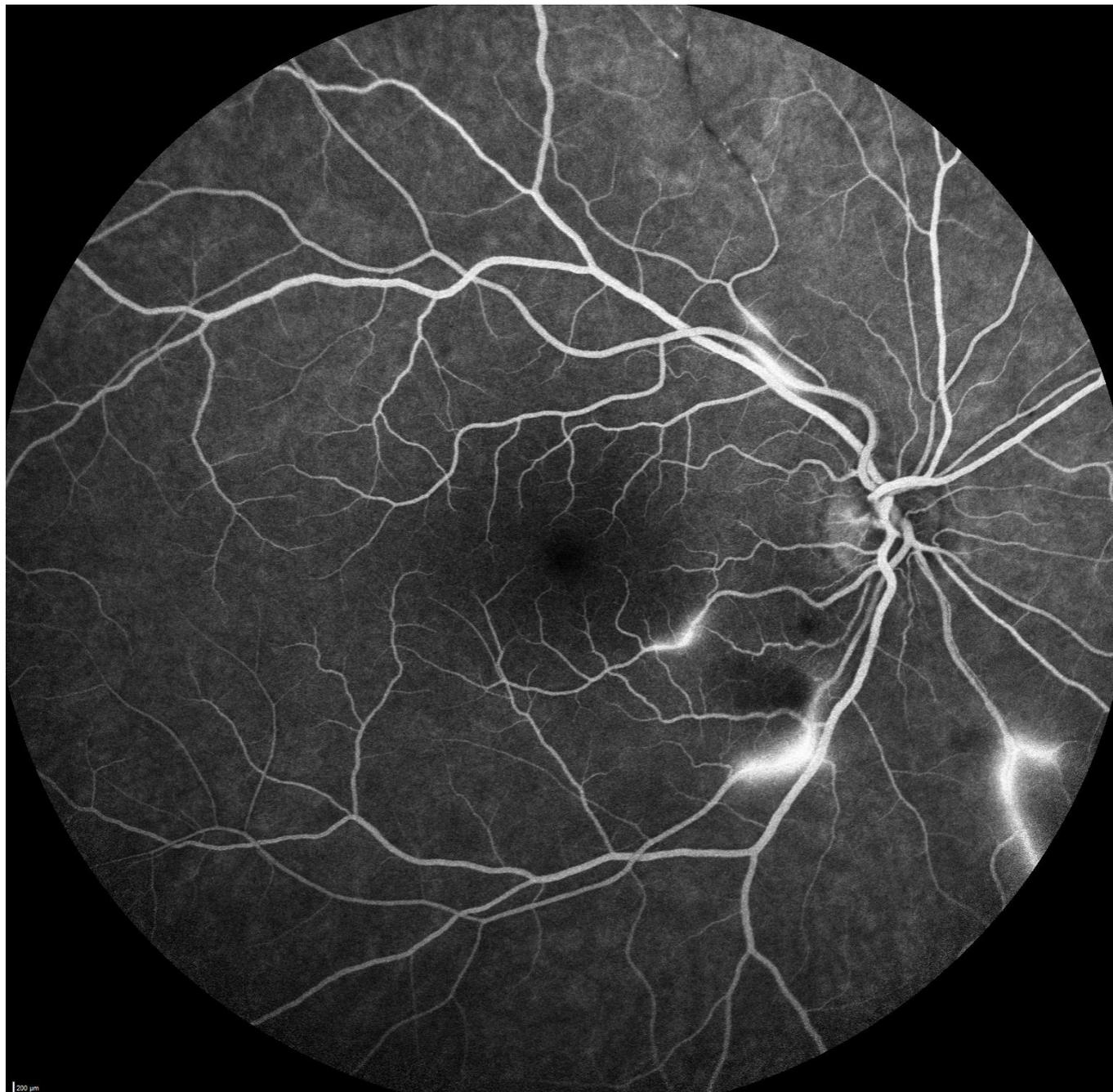
200 μ m

22/07/2016, OS
FA 6:01.33 55° ART(3) [HR]

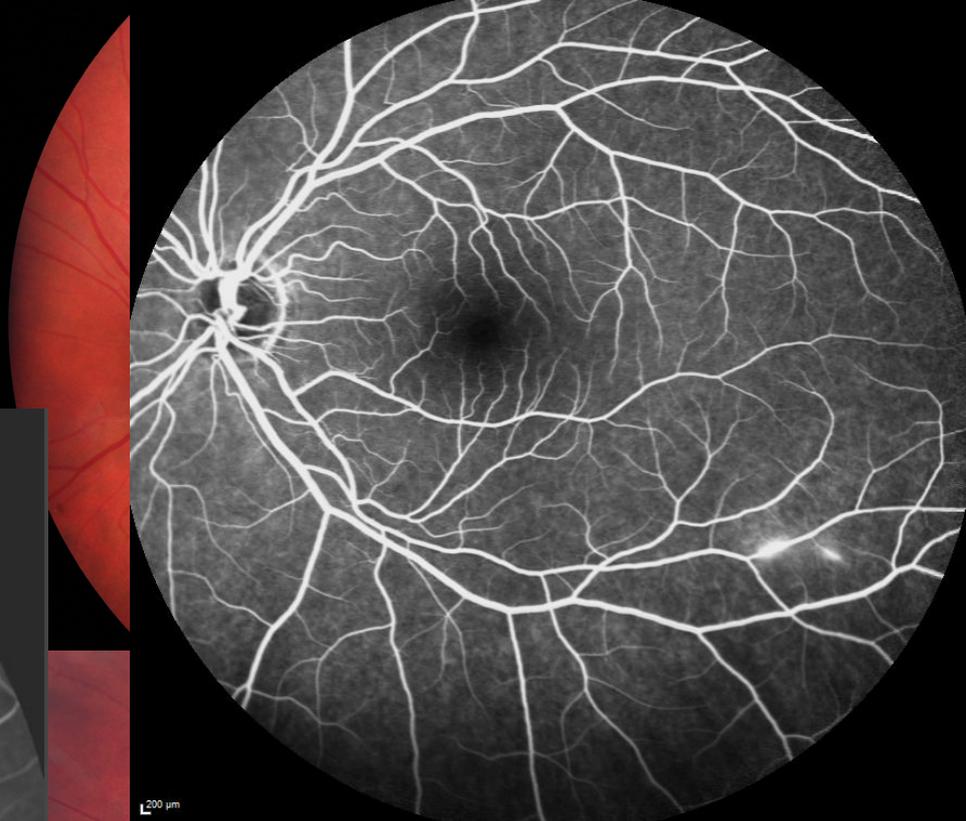




200 μ m



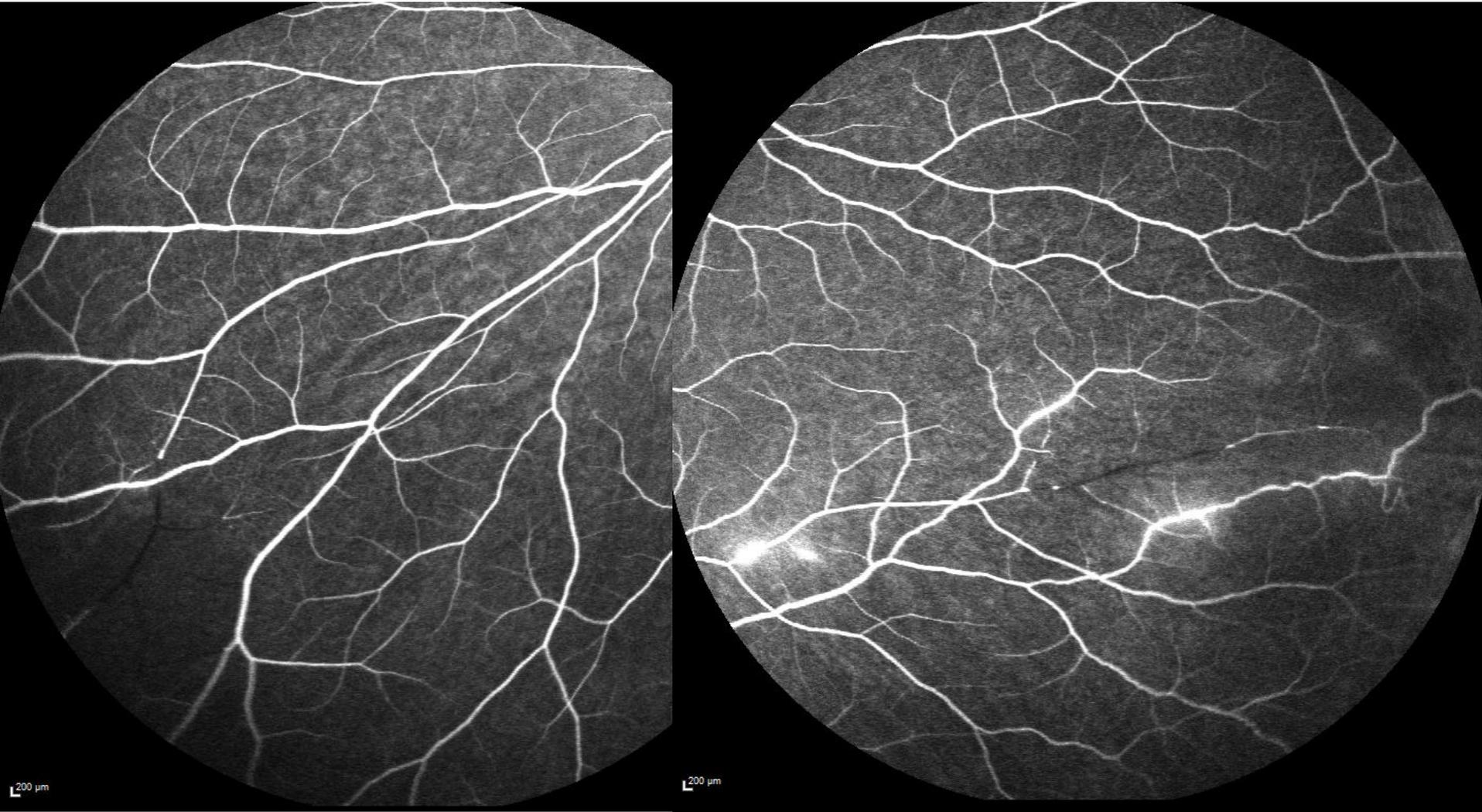
200 μ m

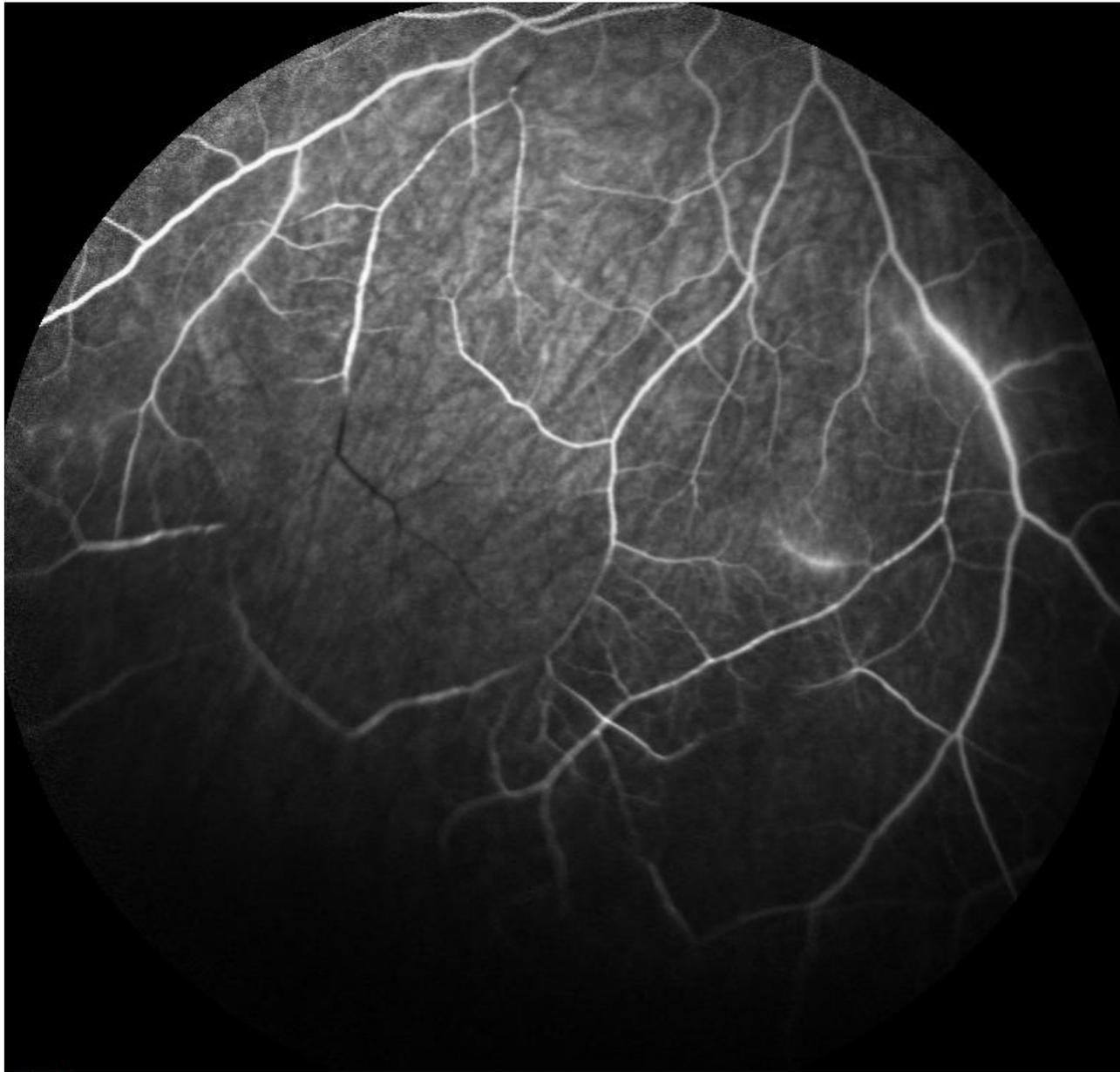


200 μm

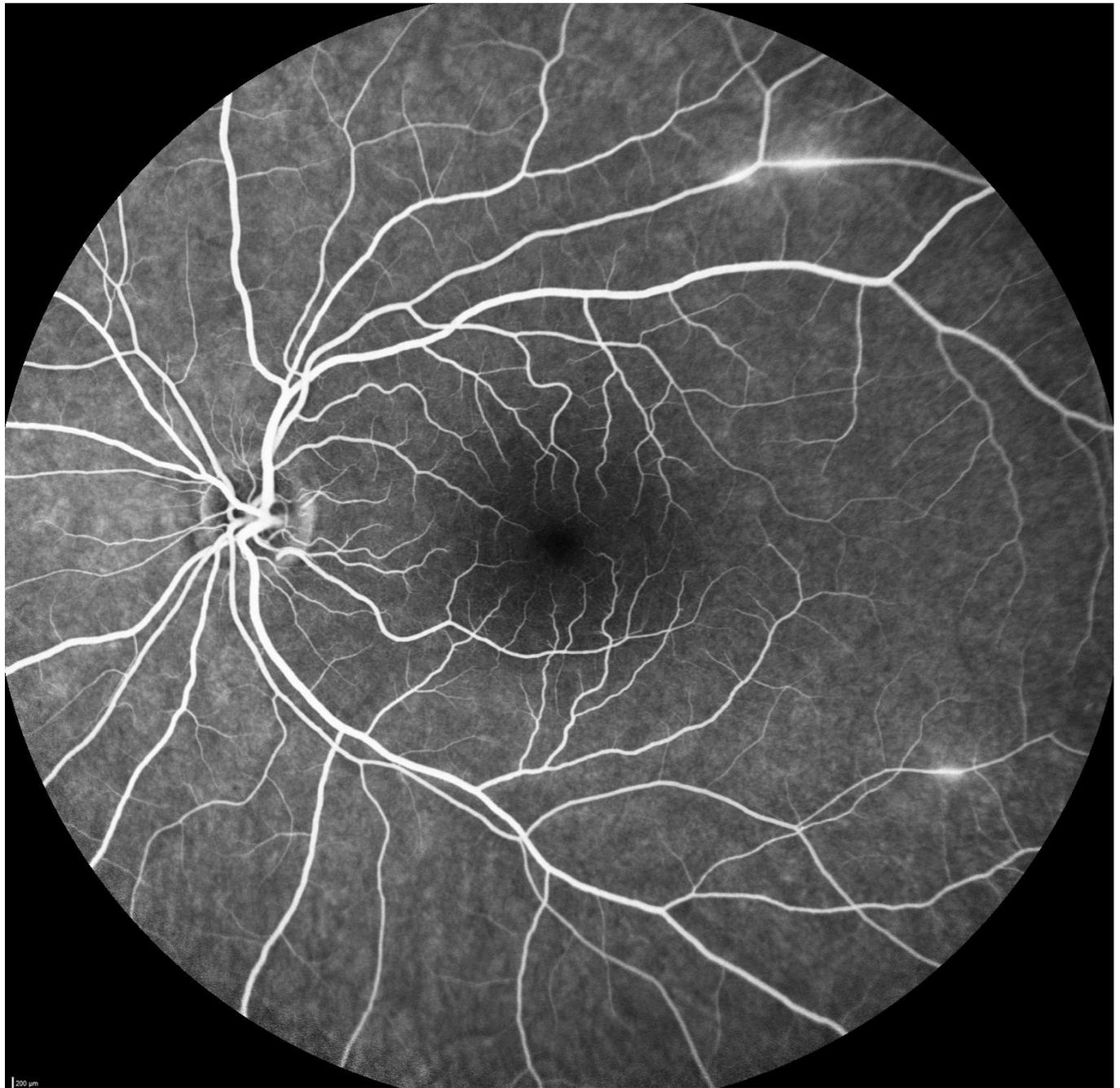
200 μm

Angiographie à la fluorescéine





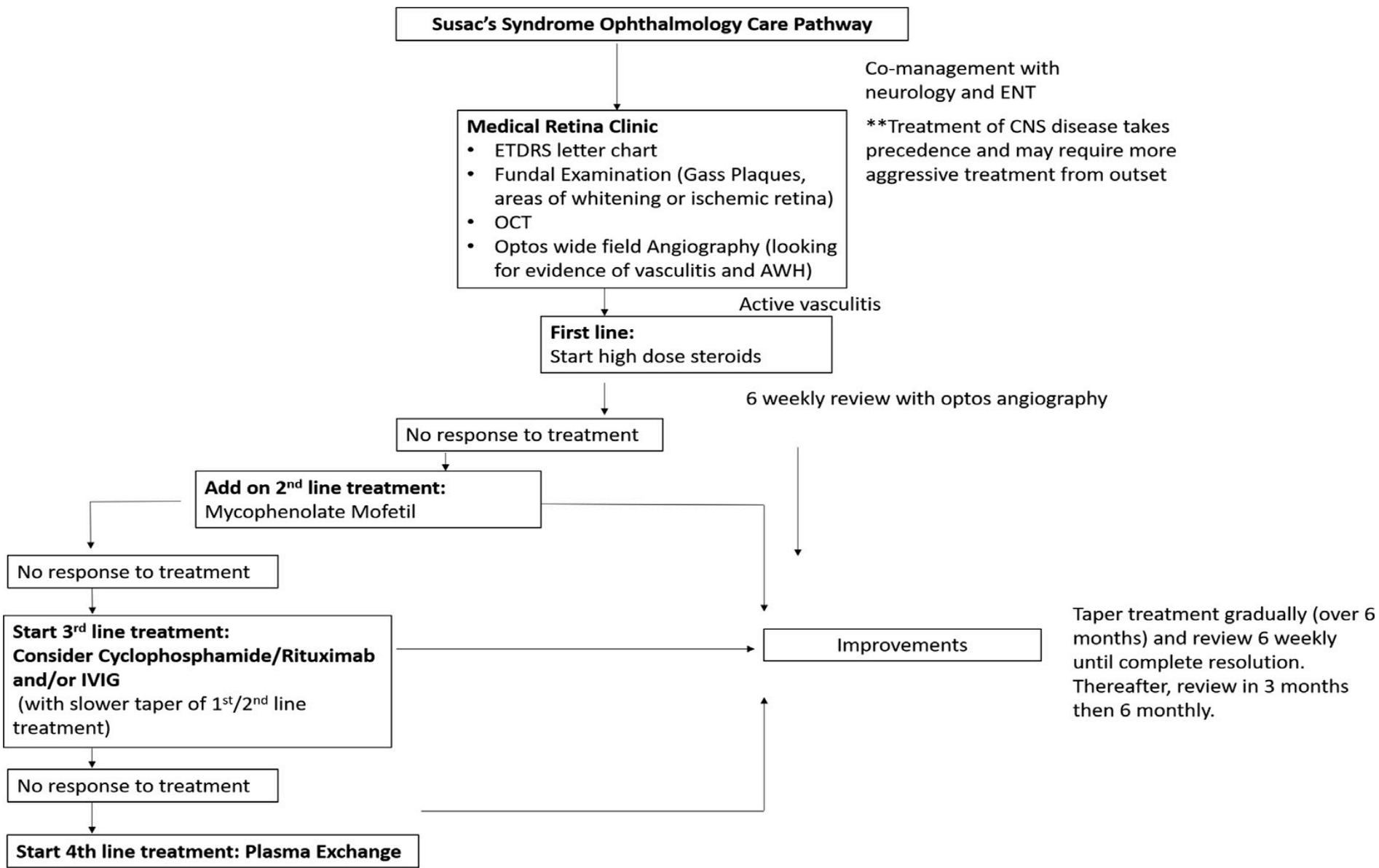
Après traitement



200 μ m

Traitement : OPH

- Aucun essai randomisé contrôlé
 - 63 case reports => Algorithme empirique
- En cas d'atteinte du SNC :
 - Bolus CT prime sur l'atteinte OPH
- Atteinte oculaire isolée : moins agressif et plus court
 - Angiographie mensuelle (3 à 6 semaines)
 - IVIG ET mycophenolate mofetil / 6 mois



Heng LZ, Bailey C, Lee R, Dick A, Ross A.

A review and update on the ophthalmic implications of Susac syndrome.

Surv Ophthalmol. 2019 Jul -Aug;64(4):477-485.



Pathologies rares = EBM possible ?

Intrication avec des atteintes extra-ophtalmologiques

Critères d'initiation de traitement :

Critères d'arrêt : encore plus difficile / définition rémission