



Macular Diseases

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The macula (anatomically fovea), situated 3 mm lateral to the optic disc, is the most sensitive part of visual acuity. The foveola is a central 0.35 mm wide zone in the macula. The inner retinal layers in the margins of the pit are displaced laterally. Figure 3.1 displays the anatomical schematic diagram of the macula.

The macular diseases mainly involve congenital anomaly (congenital macular coloboma), central serous chorioretinopathy, vitreomacular traction syndrome, vitreous hemorrhage beneath the inner limiting membrane, macular edema, macular hole, macular atrophy and proliferative diseases of the macula, such as retinal angiomatous proliferation (RAP), choroidal neovascularization (CNV), and polypoidal choroidal vasculopathy (PCV).

Stereoscopic photography plays an important role in macular diseases [1–3]. In the past, stereoscopic slide film photography of the retina is the standard with which other imaging modalities have been compared when identifying age-related macular degeneration (AMD). The Age-Related Eye Disease Study, a multicenter prospective cohort study of 4757 participants designed to access the clinical course, prognosis, and risk factor for age-related macular degeneration and cataract, uses stereoscopic color fundus photographs in a standardized fashion by certified photographers [4, 5]. In ETDRS, besides the grading diabetic retinopathy severity by stereoscopic retinal photography, the clinically significant macular edema (CSME), which is one of the key factors affecting the visual acuity, is also diagnosed correctly by stereoscopic digital fundus photography. The kappa (κ) values

among contact lens biomicroscopy (CLBM), slit-lamp biomicroscopy with 90D/78D or by stereoscopic pairs are more than 0.6. So, in most cases, the stereoscopic photography can be used as a diagnosis tool, especially for screening and telemedicine [4–6].

Abnormalities in macular diseases are various [3, 7, 8]. Drusen are yellow-white deposits within Bruch's membrane underlying the RPE and vary greatly in appearance, ranging from small round, flat spots in size and shape to large deposits even confluent with adjacent drusen. Geographic atrophy may show a sharply demarcated, usually circular zone of partial or complete depigmentation of RPE and exposure of underlying large choroidal blood vessels. The three kinds of choroidal neovascularization (type I, II, and III), which are correspondent with occult CNV, classic CNV, and retinal angiomatous proliferation (RAP), are more vividly and comprehensively shown on stereoscopic pairs than monoscopic and even OCT scans. The retina will be dome-shaped with intra-retinal exudates and cyst, accompanying by intra-retinal, sub-retinal and sub-RPE hemorrhages, sub-retinal and sub-RPE neovascularies, etc. The anastomosis between retina and choroid will be shown clearly on stereoscopic FFA pairs [7]. Macular holes are well-defined defects in the middle of the macula in various sizes. Sometimes, there is a thin membrane above the posterior retina called epi-retinal membrane (ERM).

Generally speaking, with the combination of stereoscopic photography and other high-tech tools such as OCT, more and more macular signs and characteristics will be explored.

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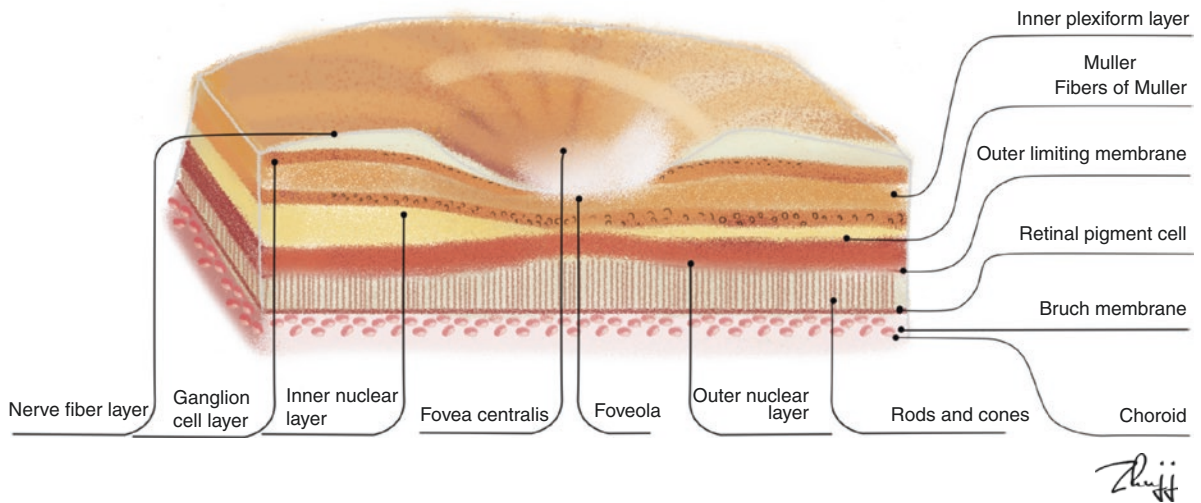


Fig. 3.1 Anatomical schematic diagram of the macula

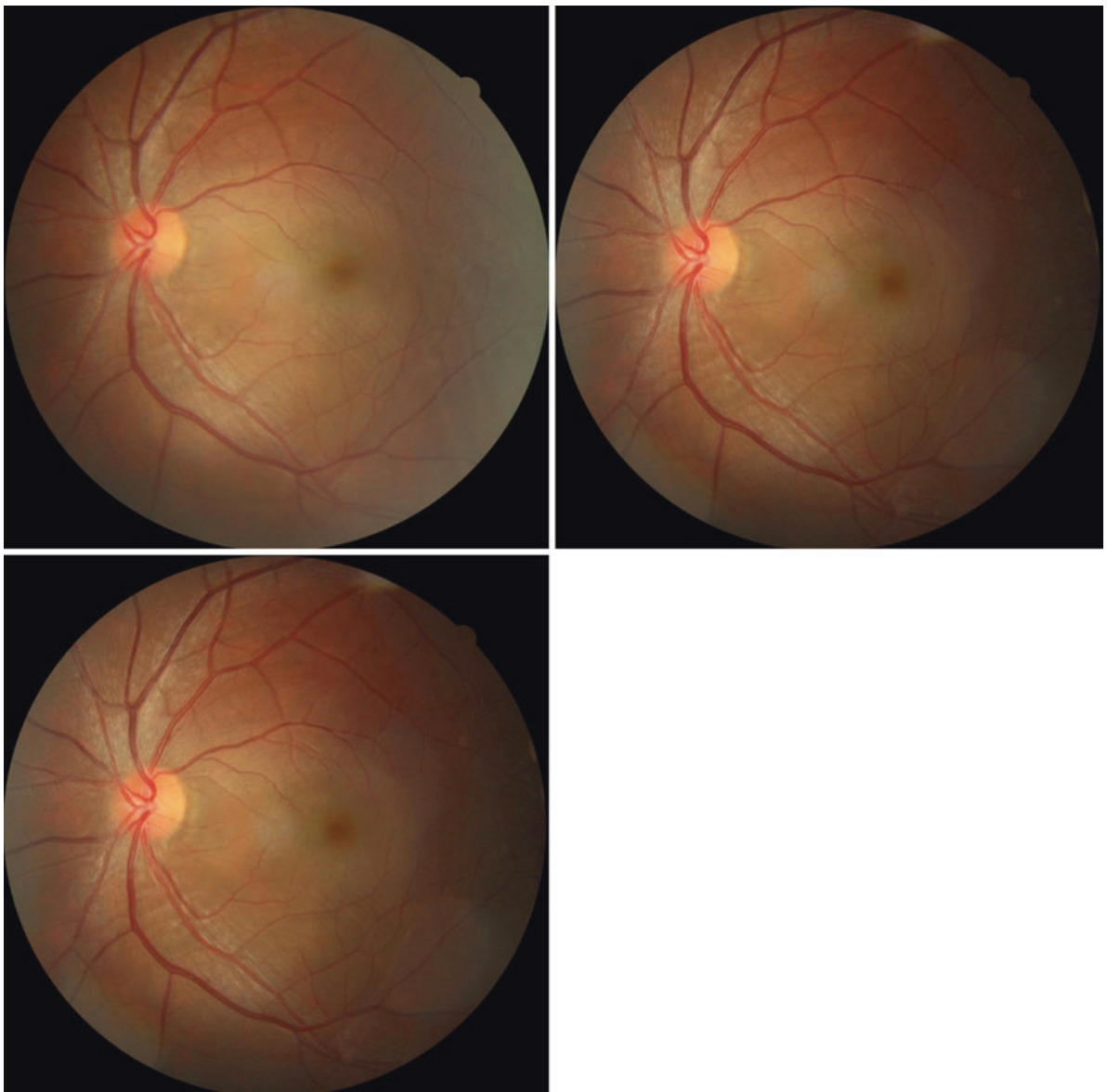


Fig. 3.2 Central serous chorioretinopathy
I. The central reflex was disappeared

II. The apex of sensory retinal detachment
III. Fold of ILM

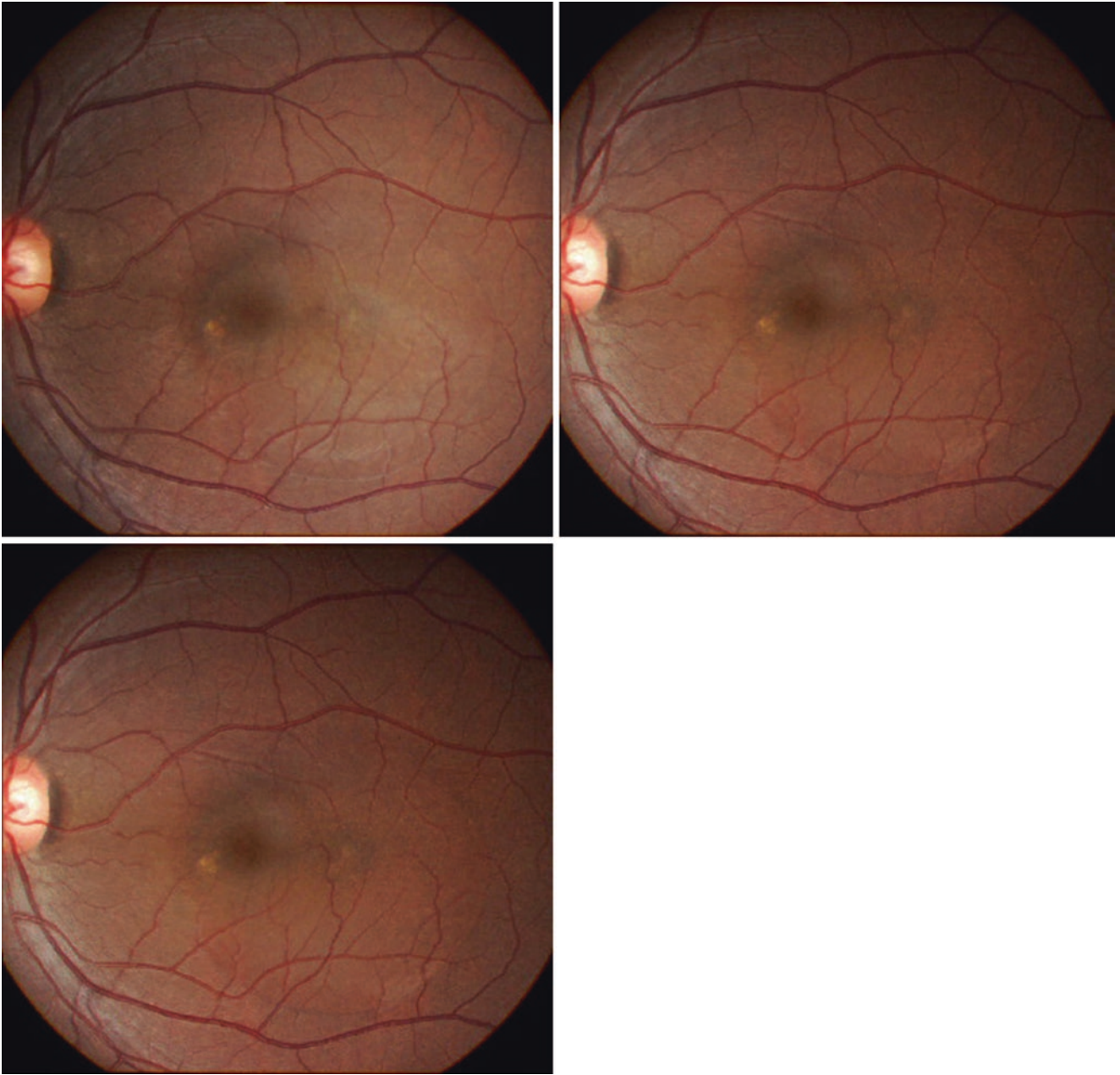


Fig. 3.3 Central serous chorioretinopathy
I. Leakage from venules inferior-nasal to the macula

II. The area of sensory retinal detachment

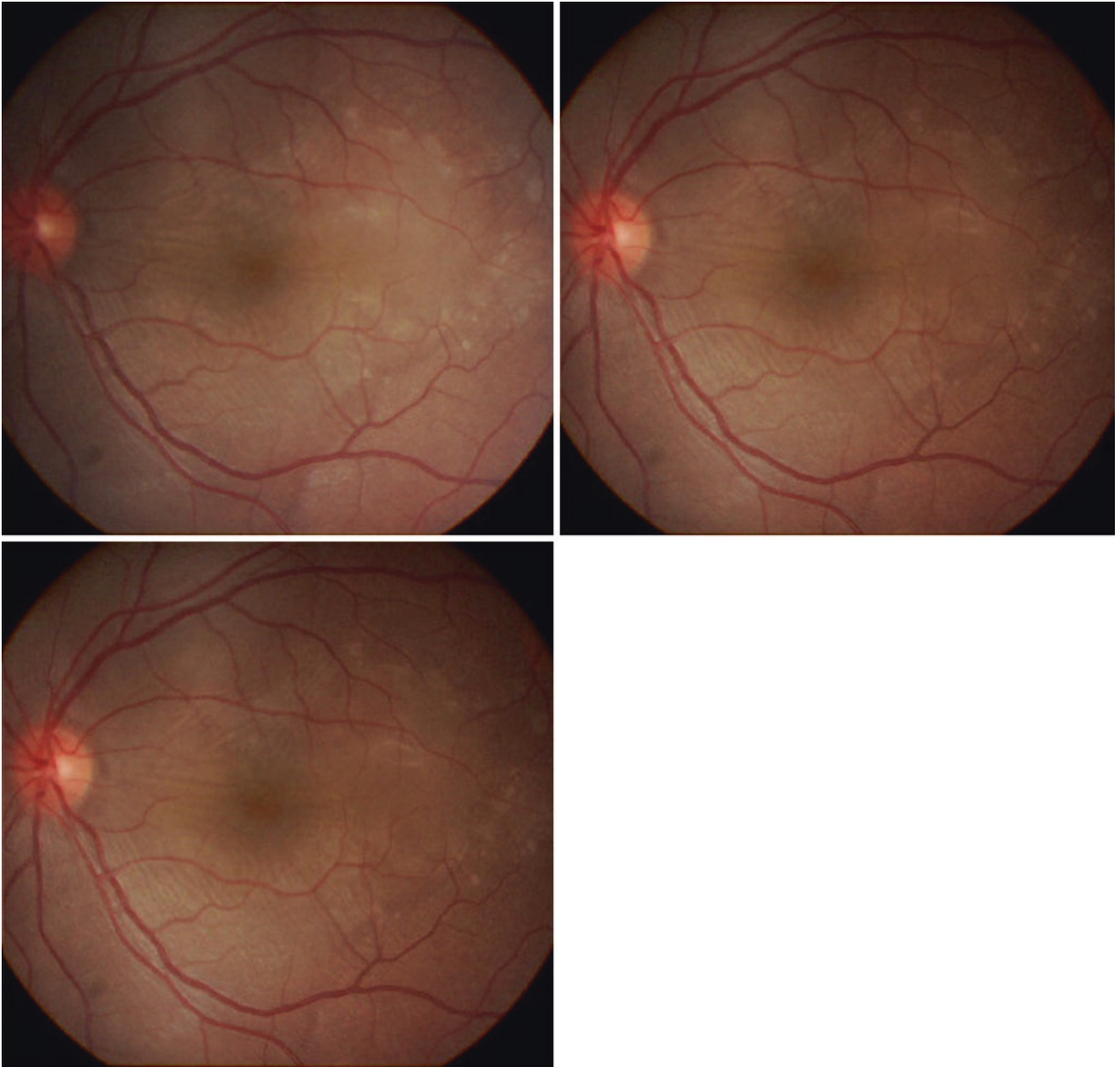


Fig. 3.4 Sensory retinal detachment
I. Apex of detachment
II. Fold of epiretinal tissue (ILM)

III. Intermediate retinal exudates
IV. Vessels located in the depressed area, which was lower than the area I

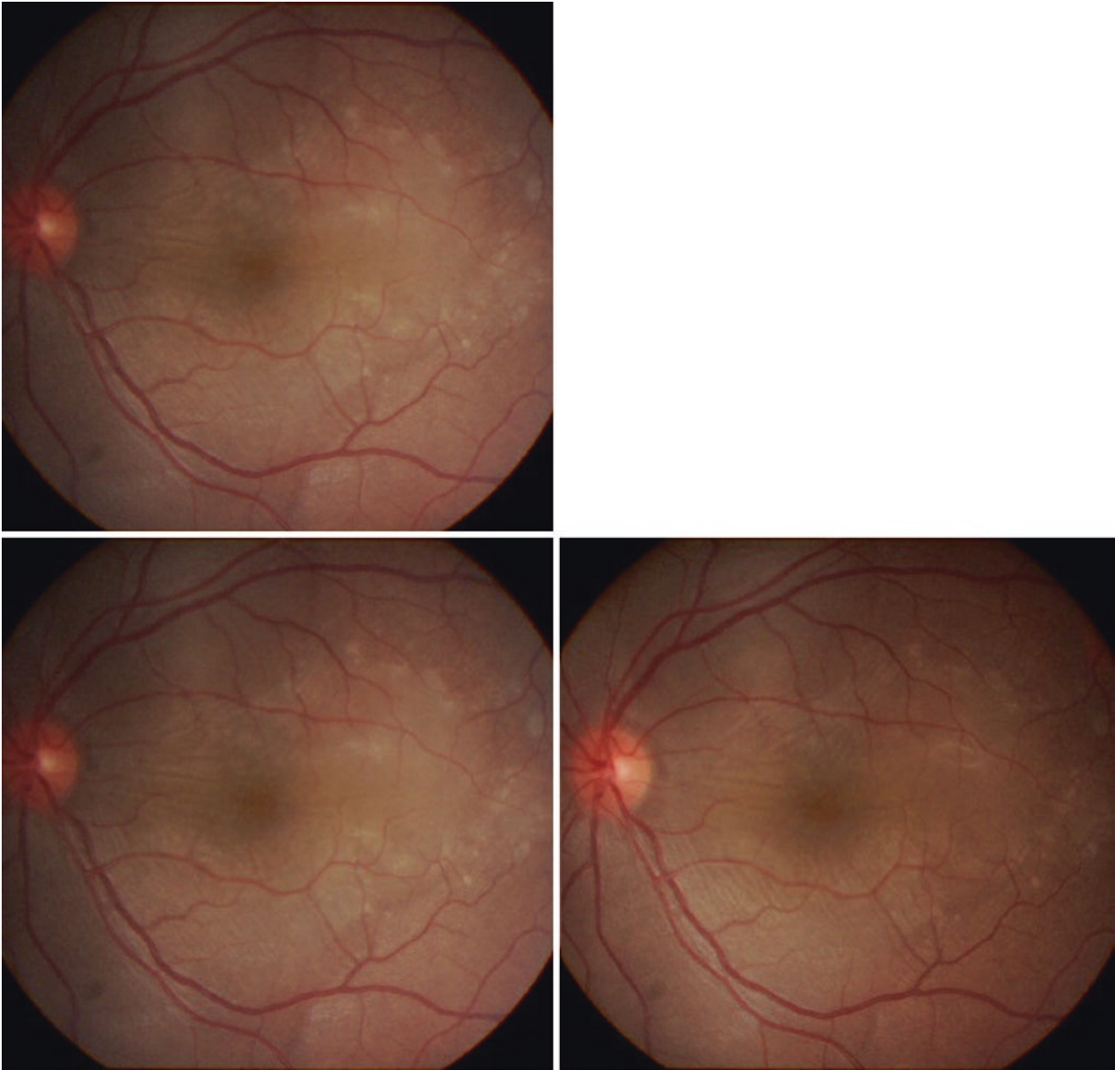


Fig. 3.5 Central serous chorioretinopathy
I. Retinal detachment in the posterior pole
II. One of the apices of detachment

III. Bottom of detachment
IV. The other one of apices of detachment
V. Retinal fold

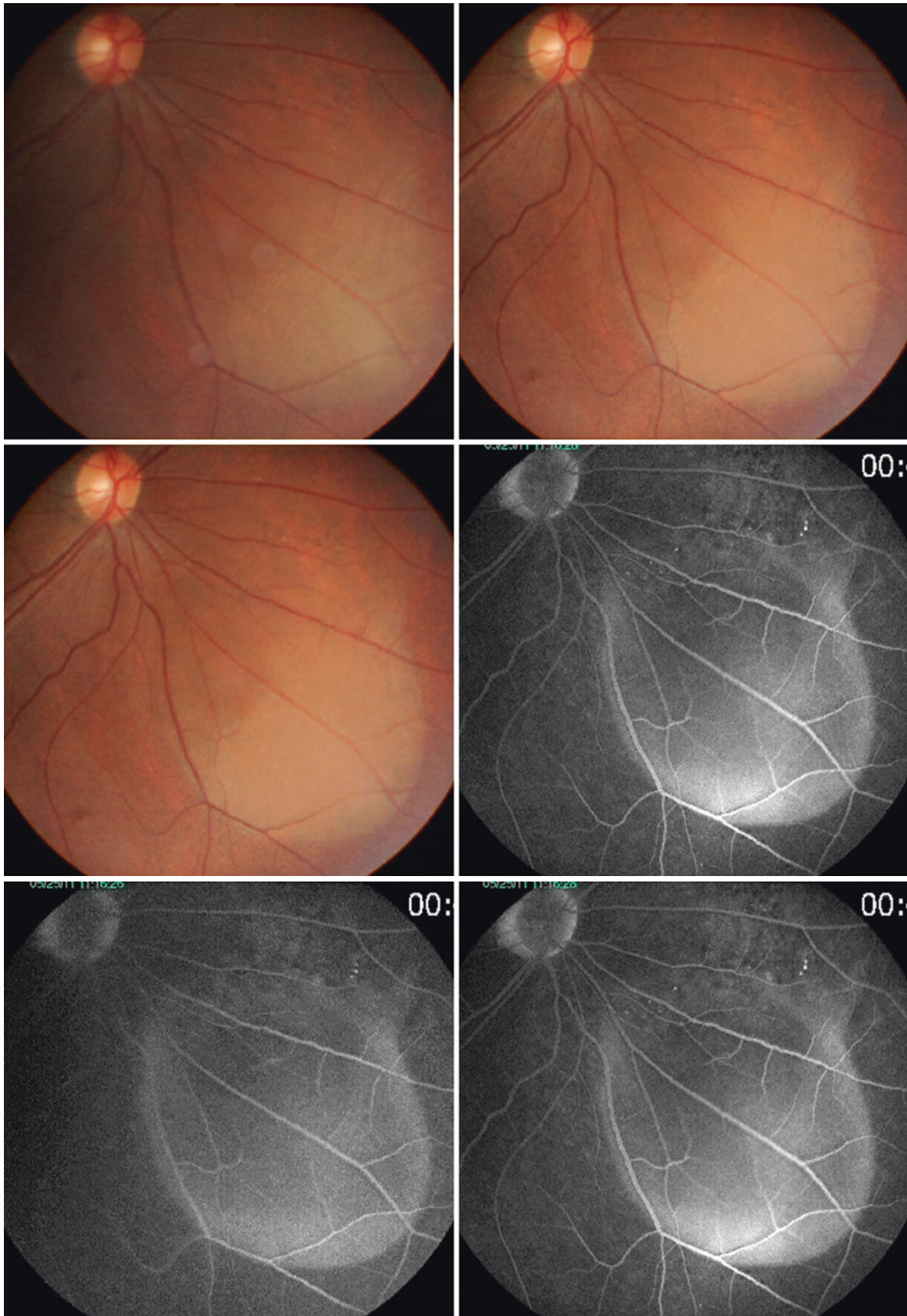


Fig. 3.6 Pouch-shaped retinal pigment epithelium detachment

I. The area of RPE detachment, which was larger in the color photograph than in the fluorescein angiography
 II. Apex of detachment

III. Sub-RPE fluid

IV. Blocked fluorescence by hemorrhage and intermediate hyperfluorescence

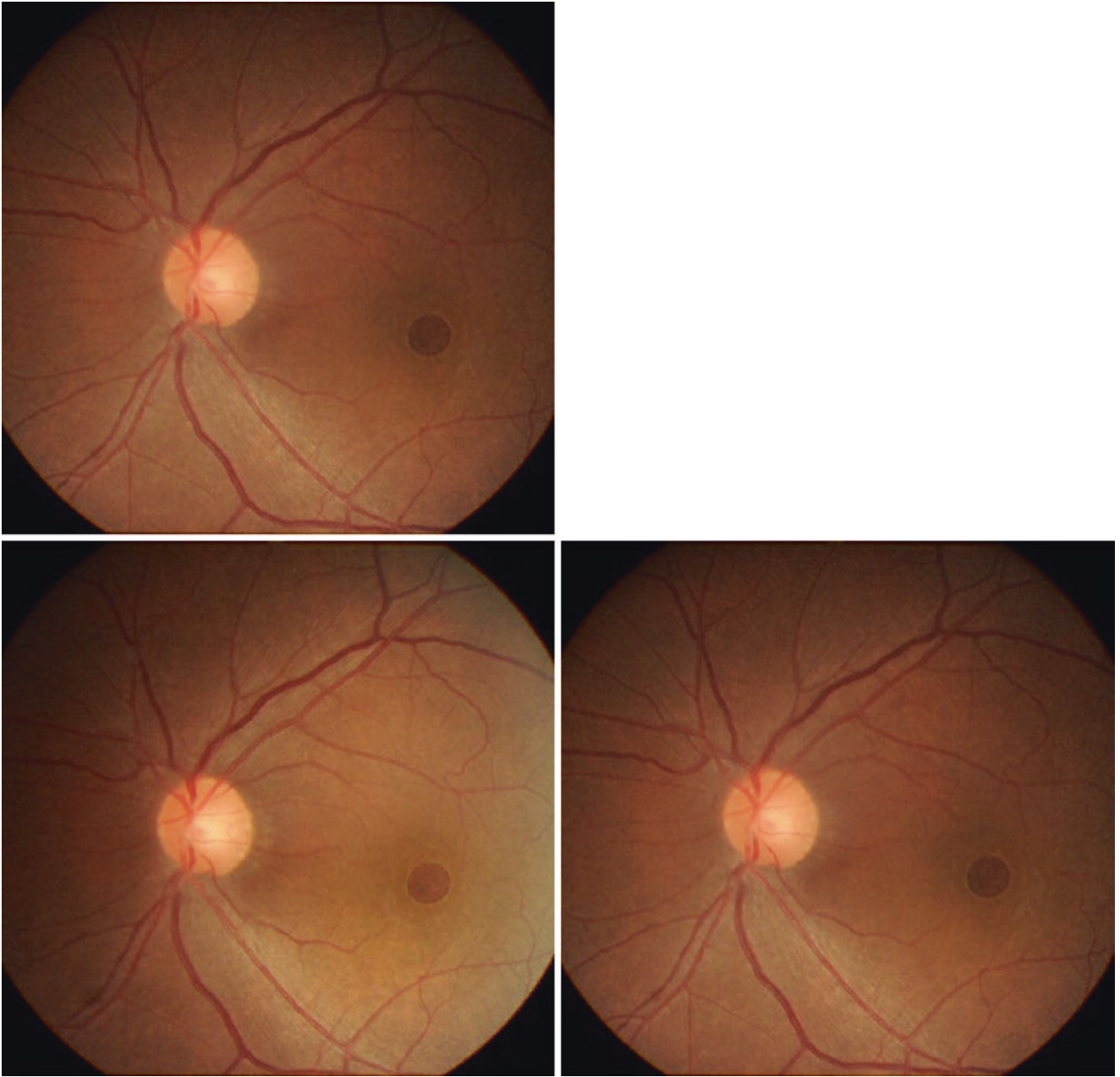


Fig. 3.7 Idiopathic macular hole

I. Full thickness macular hole

II. Shallow retinal detachment in the adjacent area

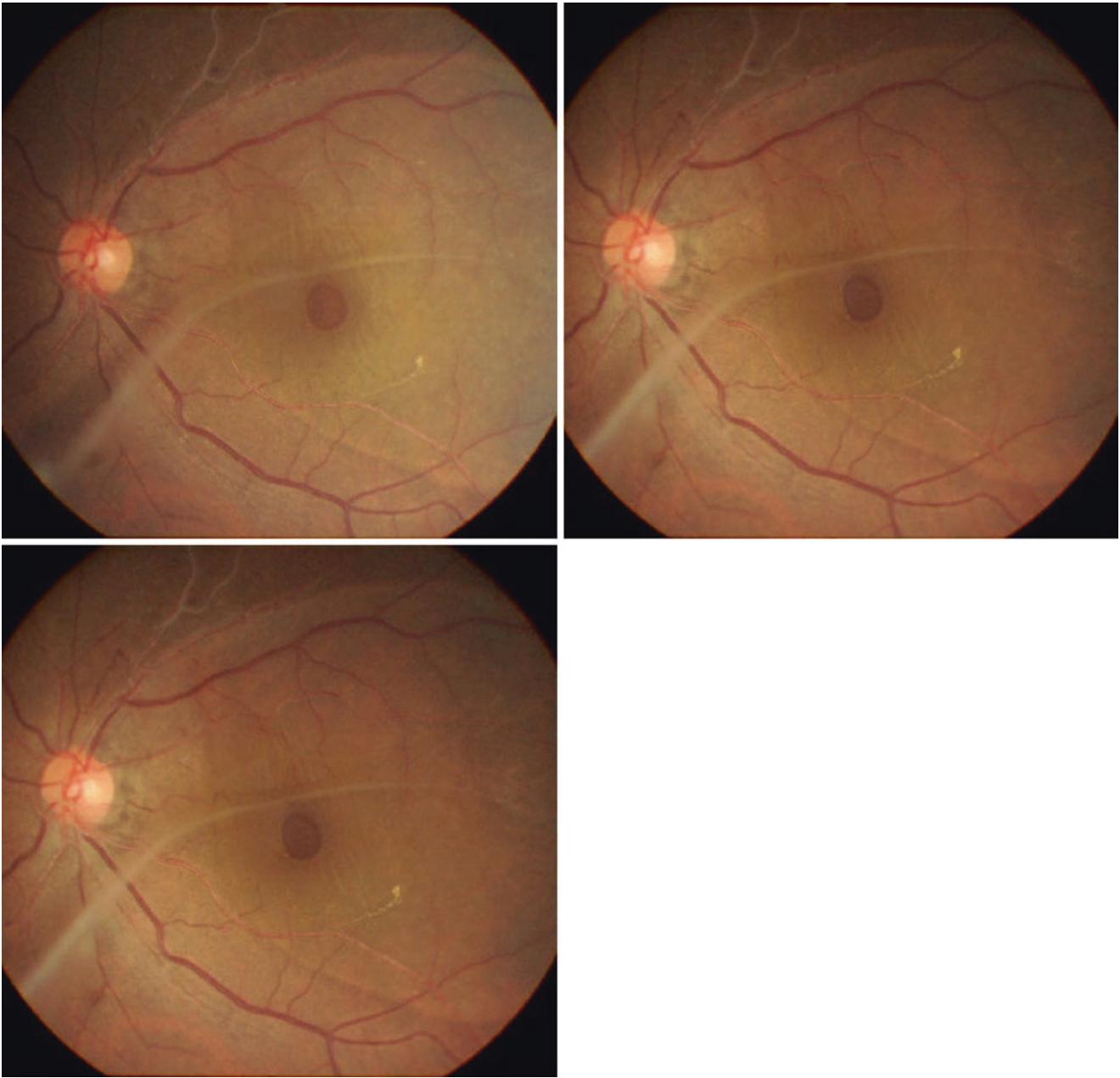


Fig. 3.8 Secondary macular hole

- I. Full thickness macular hole
- II. Proliferative membrane in the vitreous
- III. Retinal detachment
- IV. Ghost vessels

- V. Segmented sheath in the retinal arteries
- VI. Artery-Vein crossing of the superficial retinal artery and deep retinal vein
- VII. ILM fold

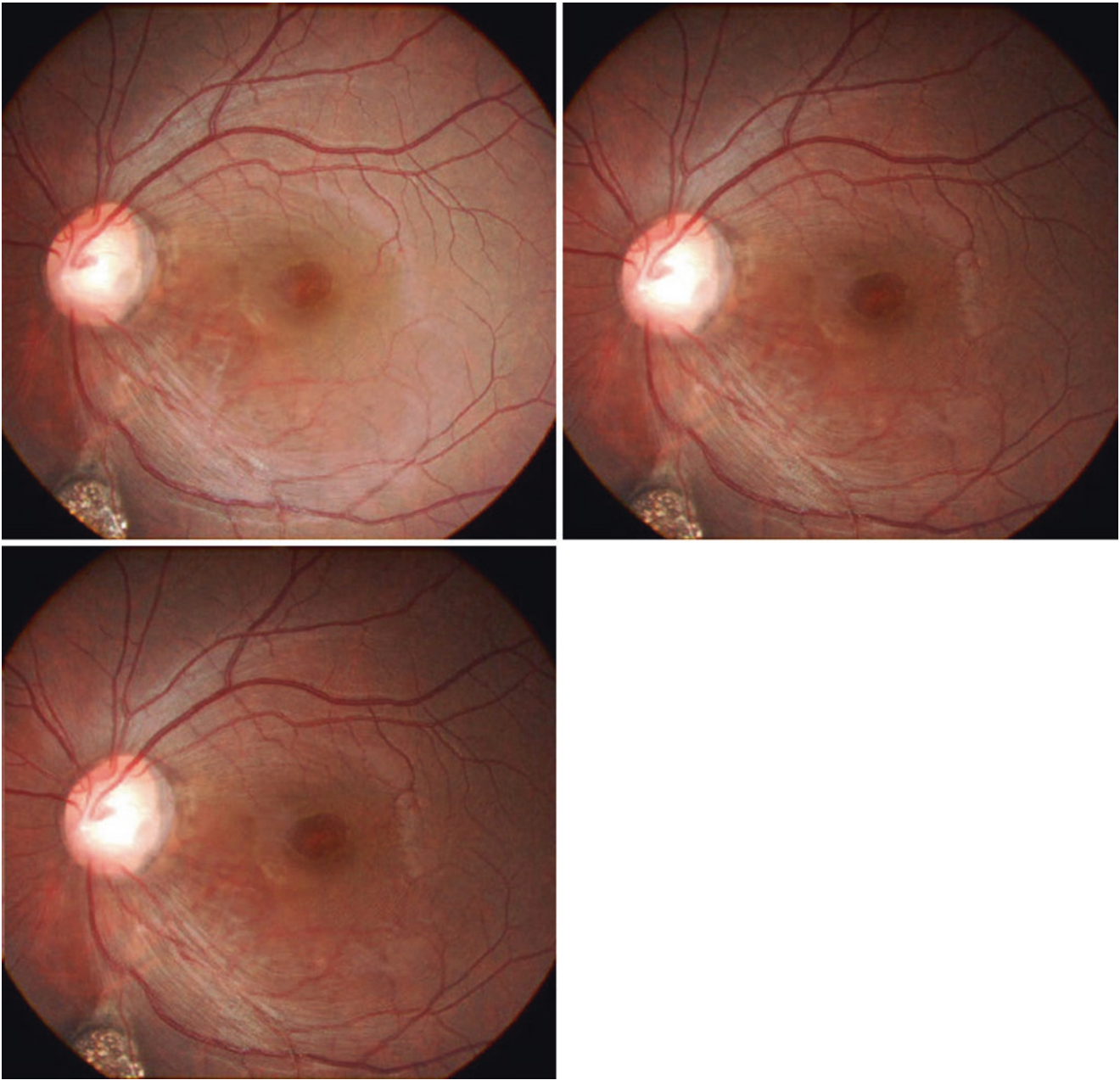


Fig. 3.9 Macular hole secondary to optic disc pit

I. Full thickness macular hole

II. Shallow retinal detachment in the adjacent area

III. Fold of posterior hyaloids and ILM, the retinal vessels were obscure

IV. Choroidal coloboma

V. Optic disc pit

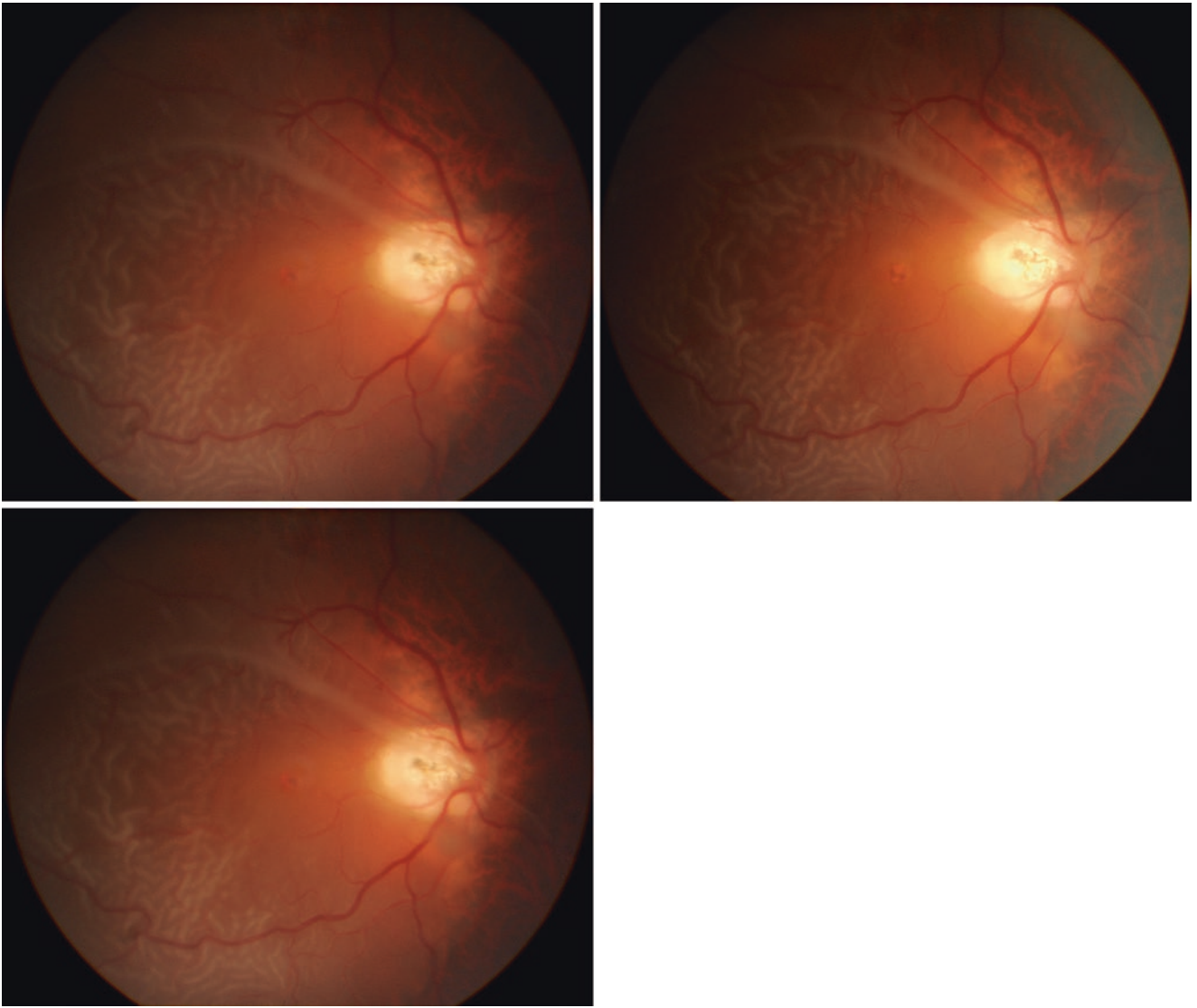


Fig. 3.10 Retinal detachment secondary to macular hole
I. Full thickness macular hole, approximately 1/4 PD in diameter
II. Retinal detachment and fold

III. Proliferation under the retina like a streak
IV. Peripapillary atrophy

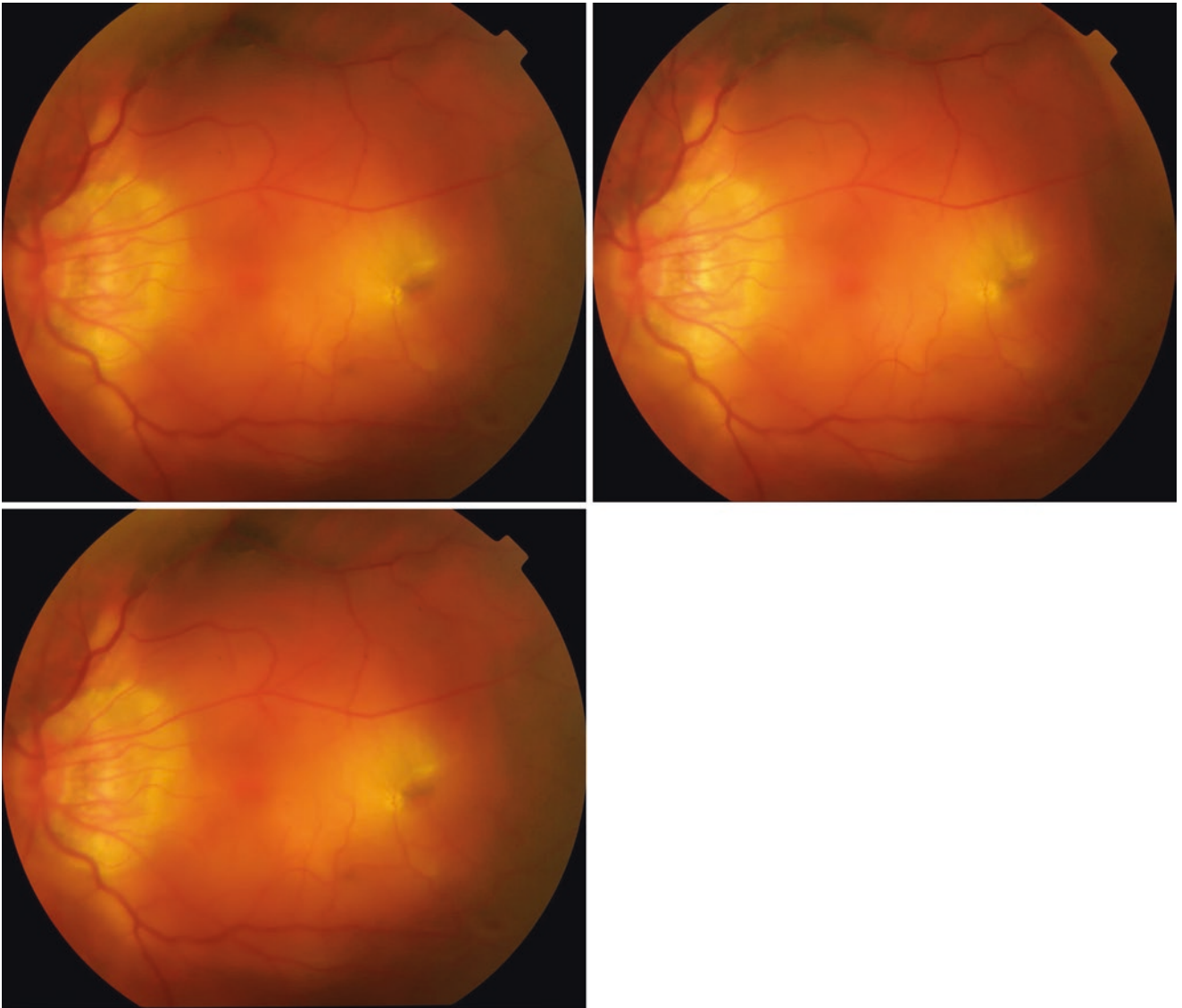


Fig. 3.11 Retinal detachment secondary to juxta foveal hole
I. Suspected para-macular hole, approximately 1/4 PD in diameter,
which was confirmed by OCT

II. Detached macula
III. Vitreous band
IV. Peripapillary atrophy

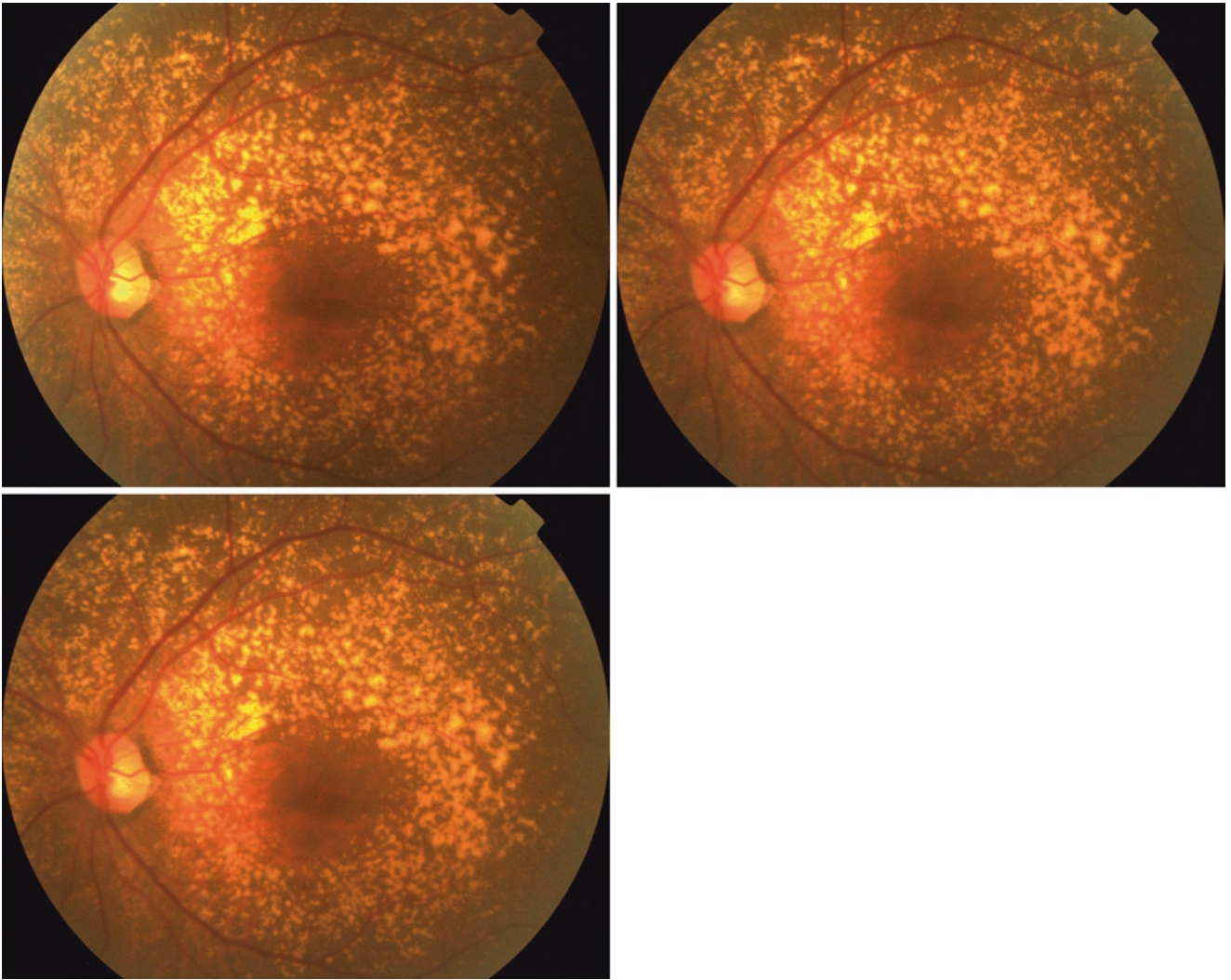


Fig. 3.12 Dry age-related macular degeneration
I. Diffused hard drusen

II. Confused soft drusen
III. Macula uninvolved

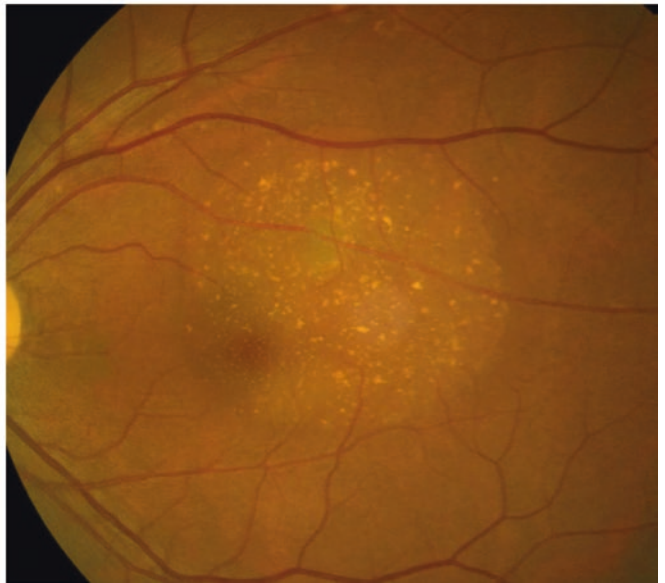
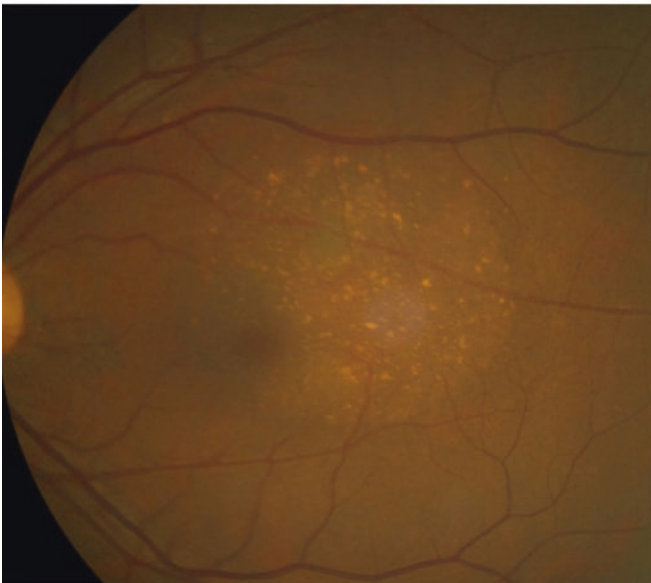
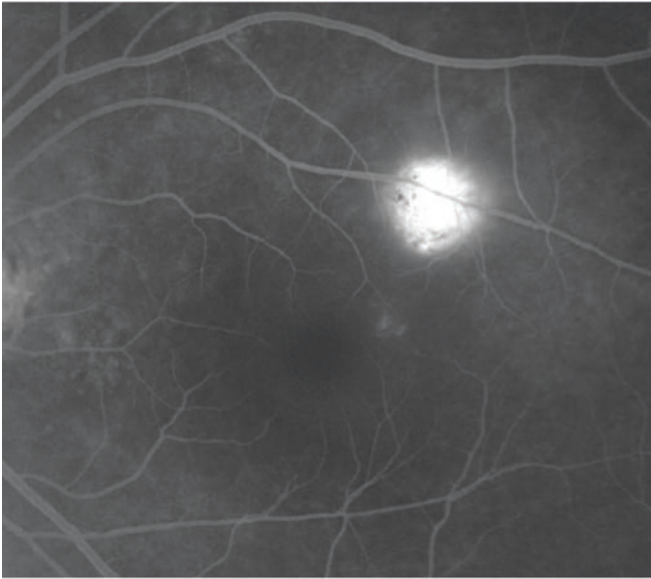


Fig. 3.13 Juxtafoveal choroidal neovascularization
I. Juxtafoveal choroidal neovascularization
II. RPE detachment and exudates

III. The area of sensory retinal detachment
IV. Macular edema

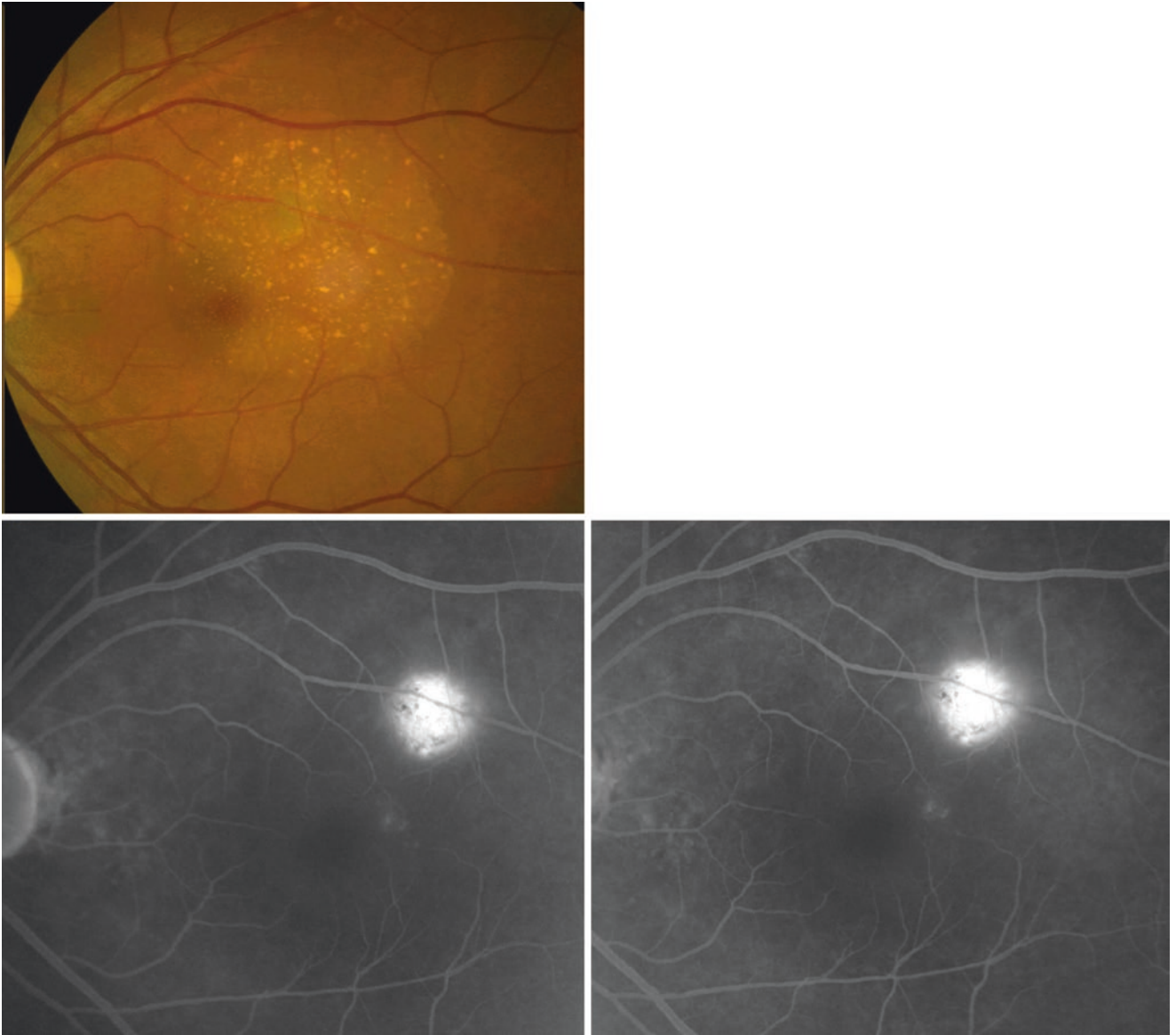


Fig. 3.13 (continued)

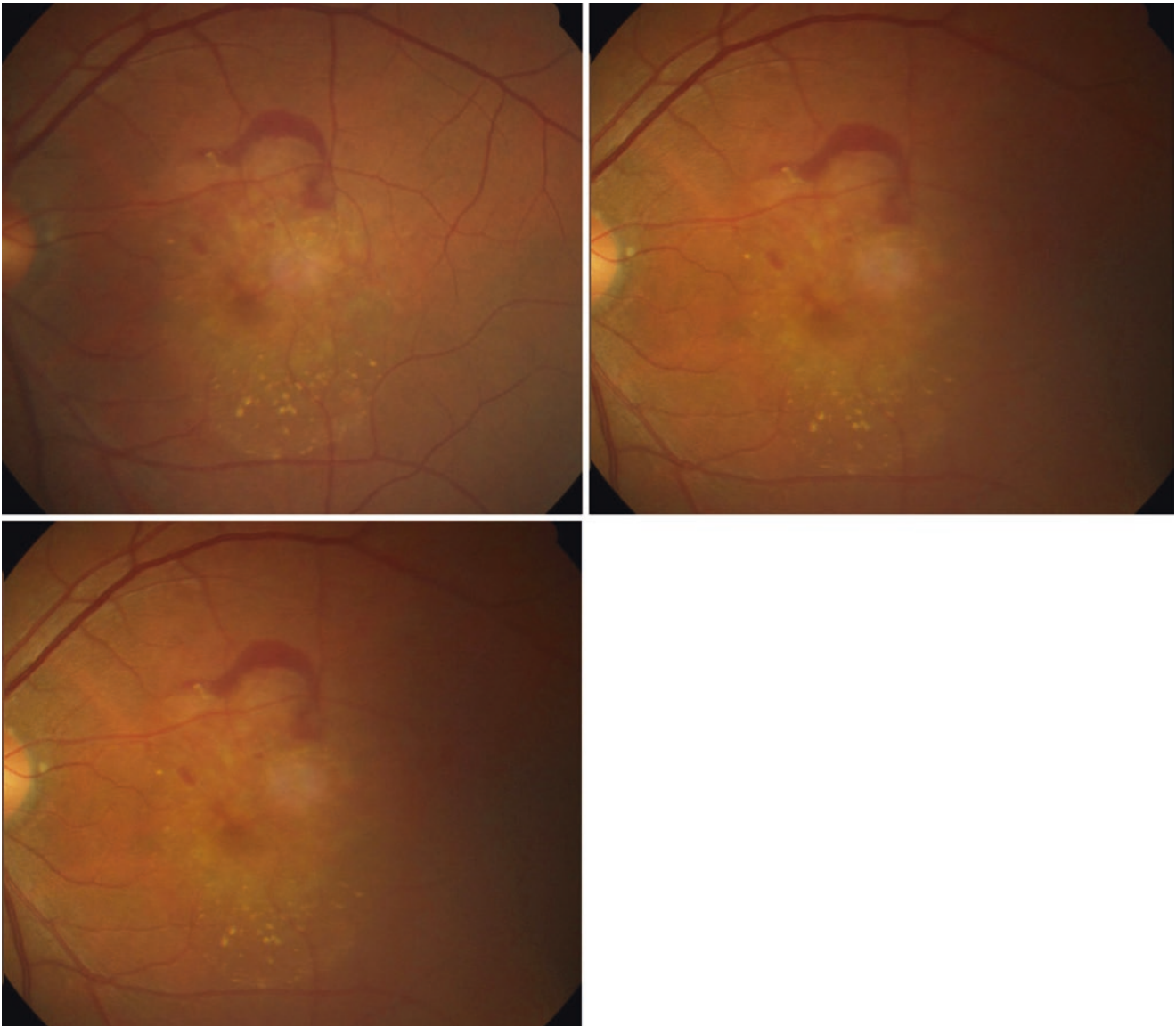


Fig. 3.14 Choroidal neovascularization
I. Subfoveal CNV
II. Superficial retinal exudates

III. Exudates in the inner retina
IV. Deep retinal hemorrhage
V. Suspected area of CNV

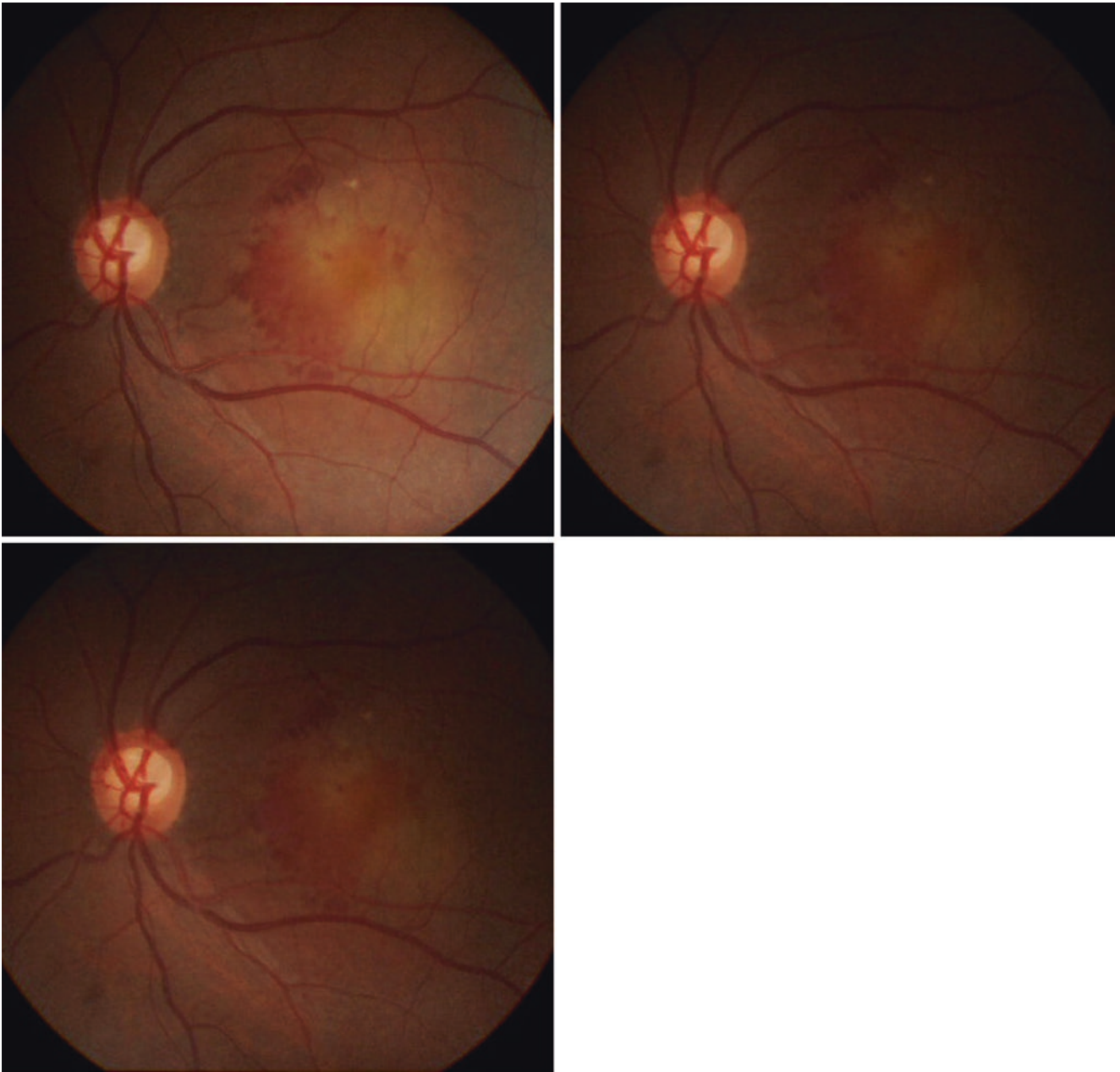


Fig. 3.15 Choroidal neovascularization
I. Subfoveal choroidal neovascularization

II. Intra-retinal hemorrhage around the lesion

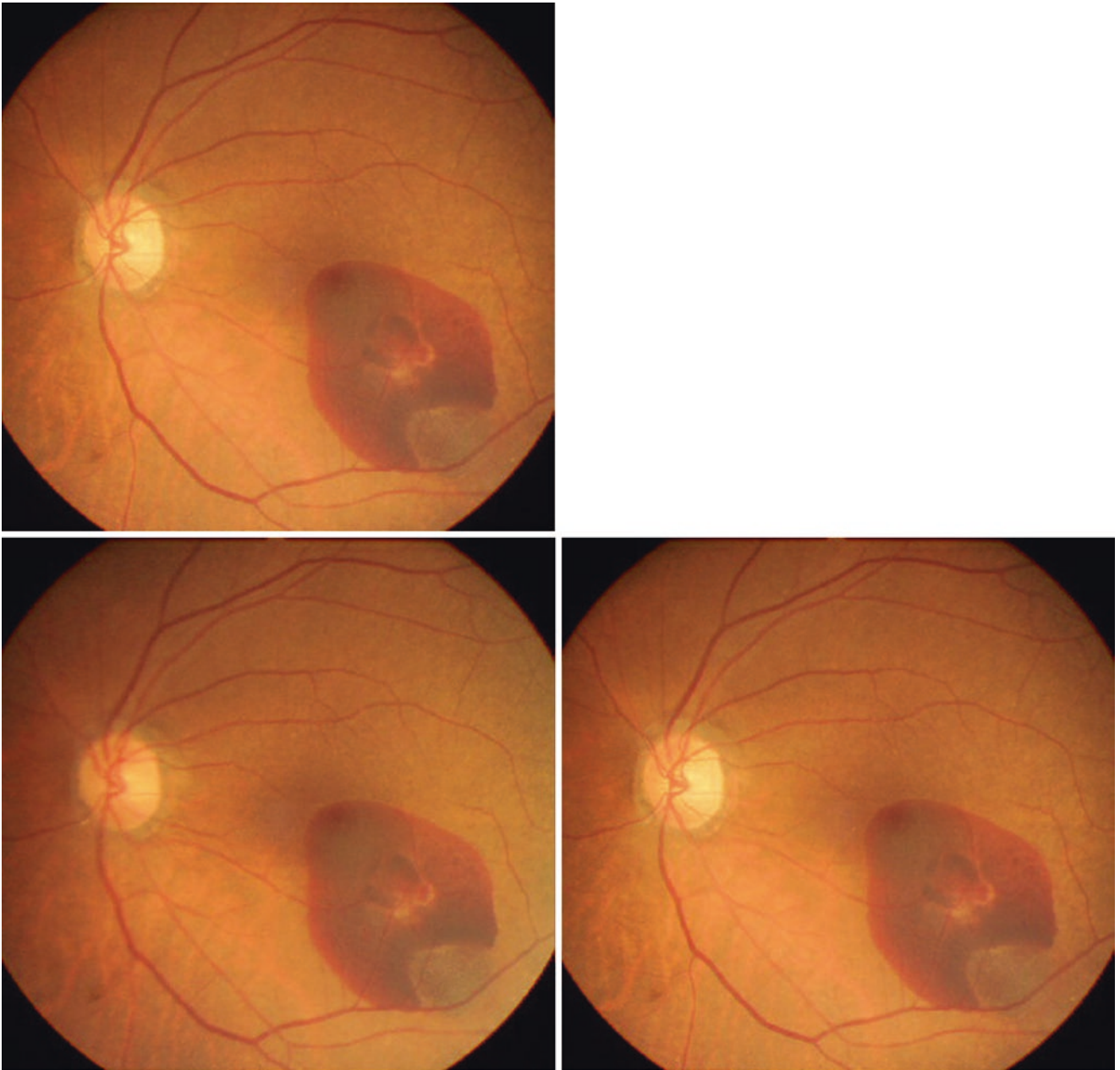


Fig. 3.16 Juxtafoveal CNV
I. Suspected area of CNV
II. Sensory retinal detachment

III. Sub-RPE hemorrhage
IV. Sub-retinal hemorrhage

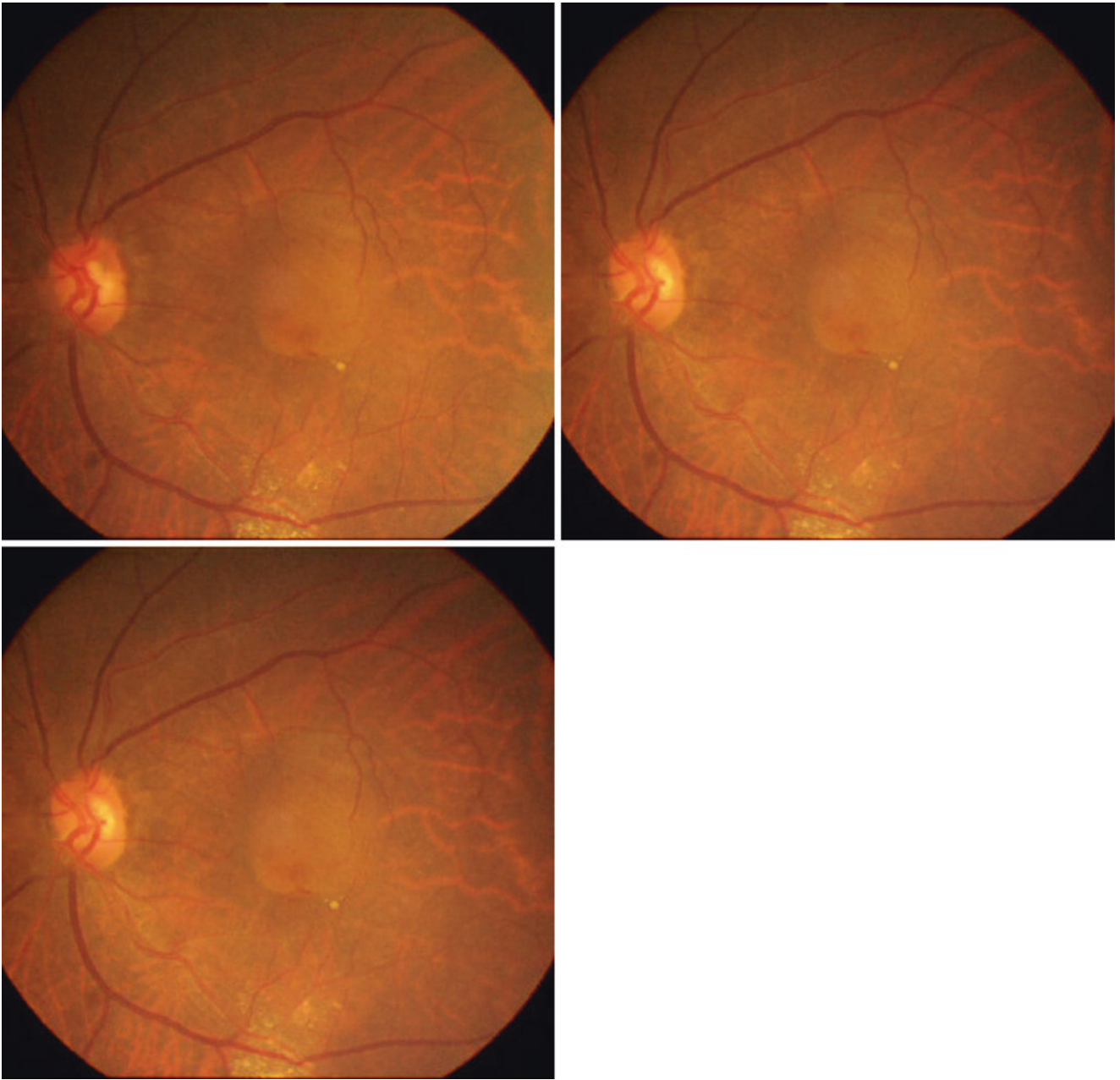


Fig. 3.17 Sub-macular choroidal neovascularization

I. Sub-macular choroidal neovascularization

II. Small thread-like hemorrhage

III. Dotted exudates

IV. Localized narrowing of retinal vessels and drusen

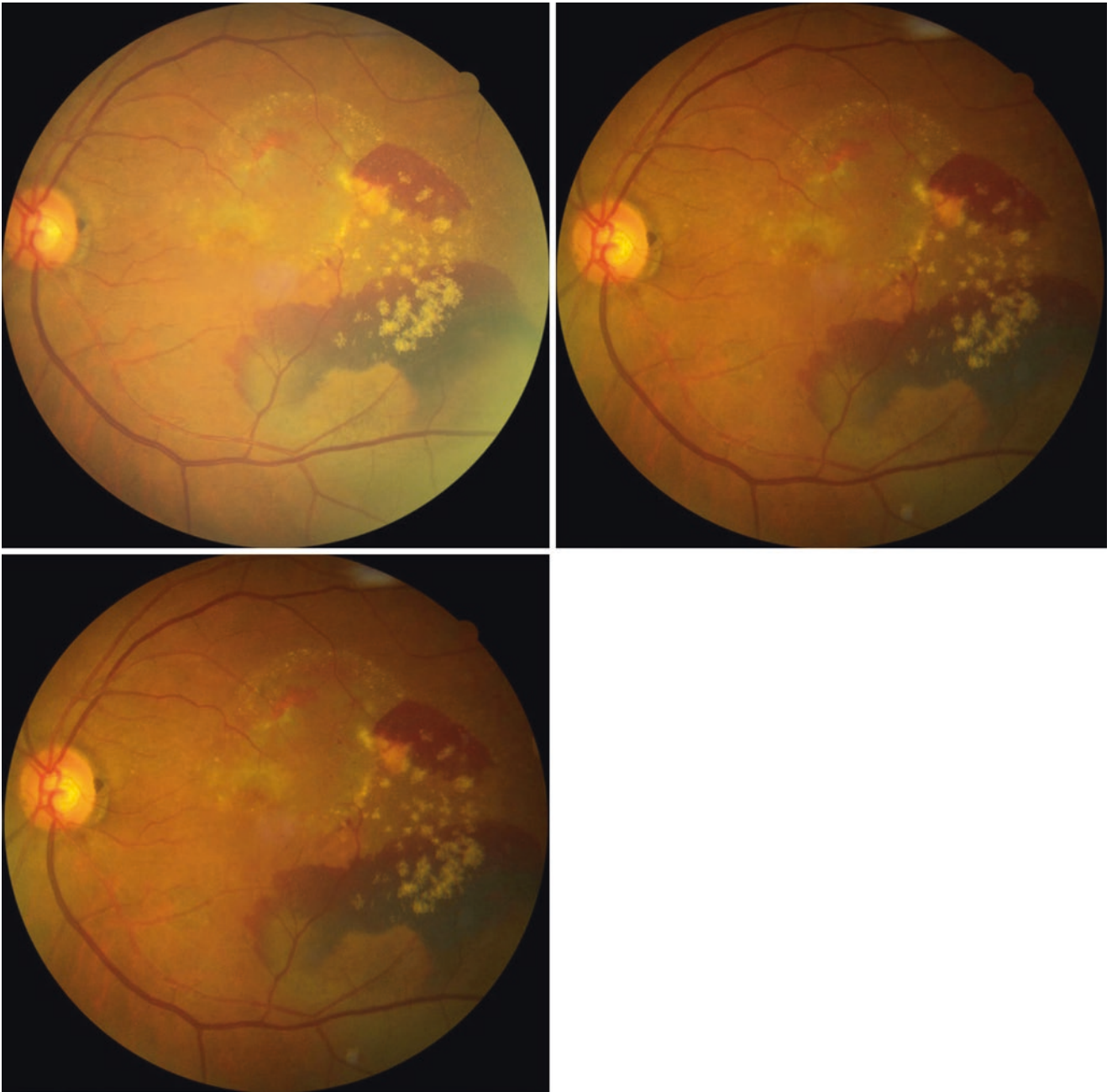


Fig. 3.18 Polypoidal choroidal vasculopathy
I. Multiple suspected polypoidal lesions
II. Deep retinal hemorrhage

III. Superficial retinal exudates
IV. Sub-retinal hemorrhage
V. Sub-RPE hemorrhage

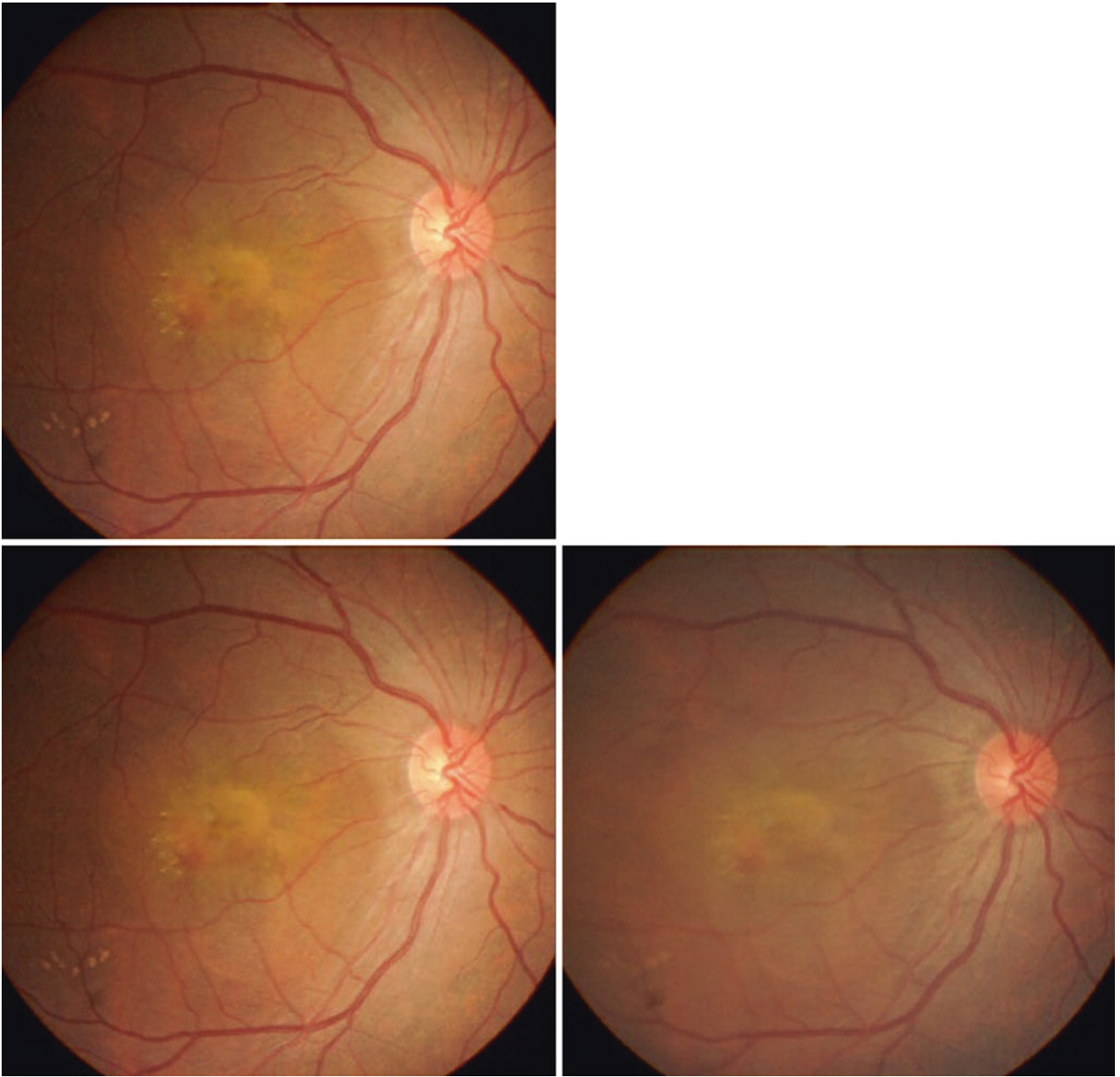


Fig. 3.19 Retinal angiomatous proliferation (RAP)

I. Intra-retinal neovascularization

II. Sub-retinal choroidal neovascularization

III. Deep retinal exudates

IV. White-dotted sub-retinal exudates

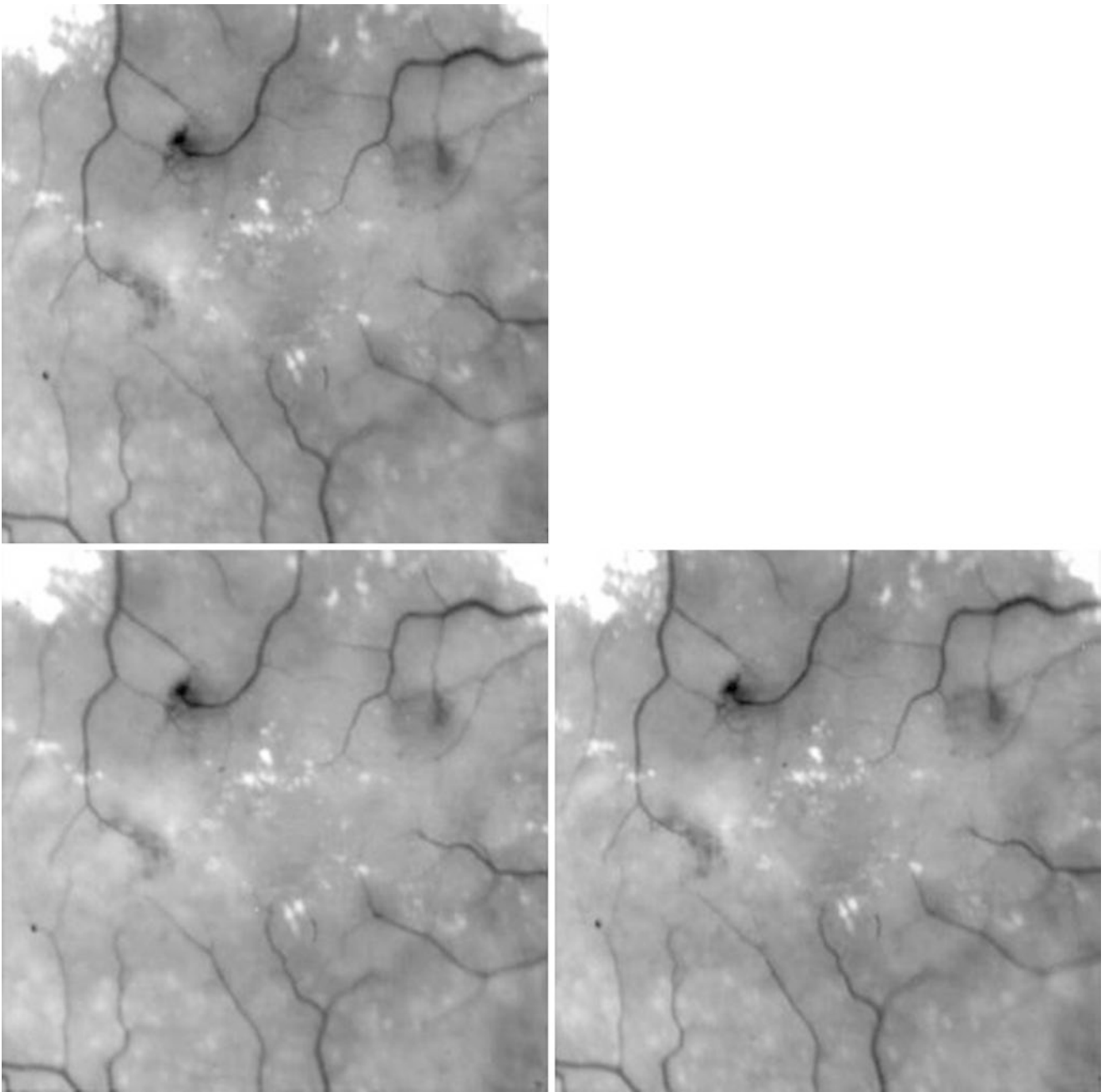


Fig. 3.20 Retinal angiomatous proliferation (RAP) on FFA
I. Intra-retinal neovascularization

II. Sub-retinal choroidal neovascularization
III. Elevated fovea and scattered exudates

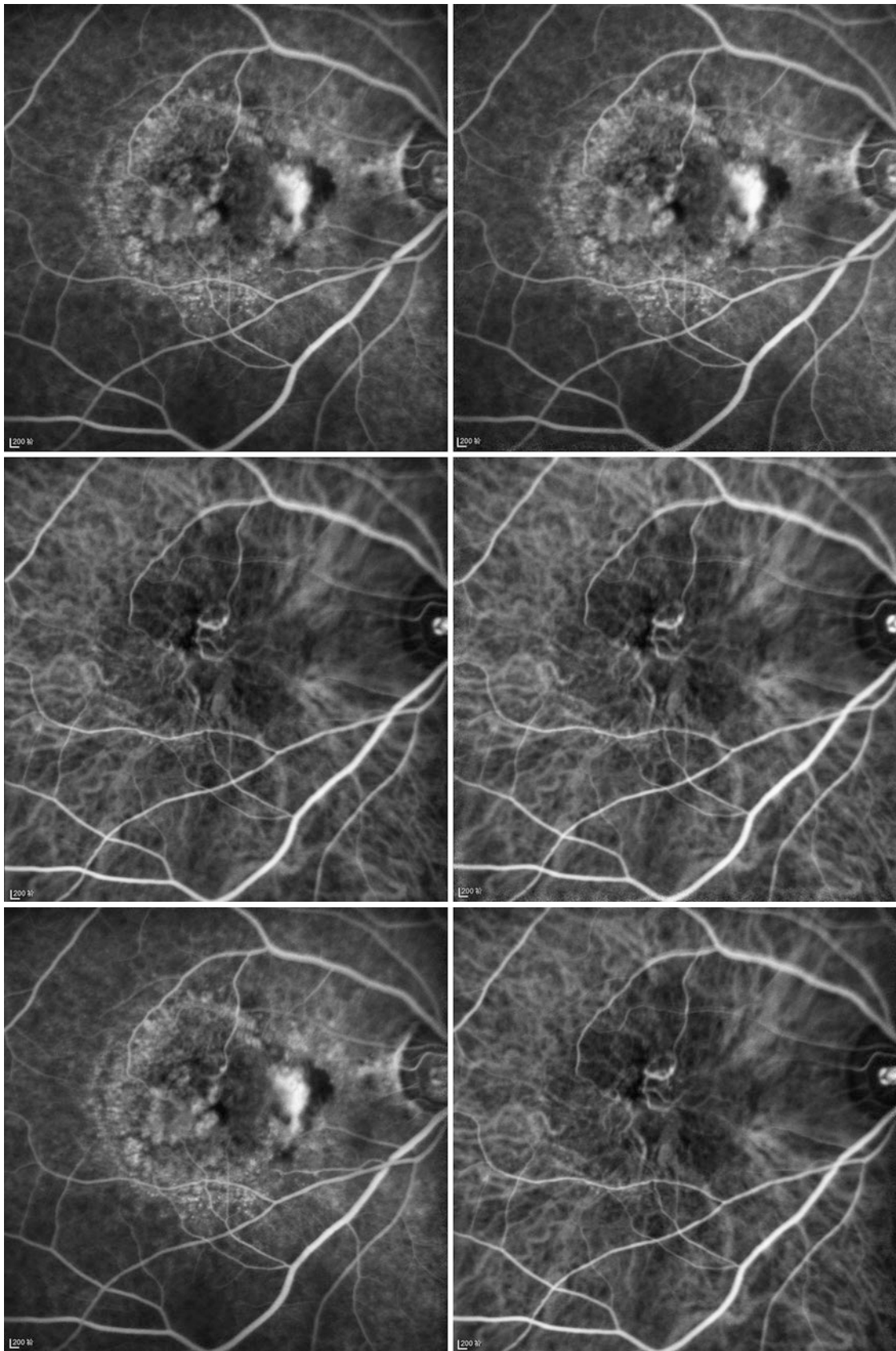


Fig. 3.21 Retinal angiomatous proliferation (RAP) on FFA
I. Intra-retinal neovascularization on early FFA

II. Intra-retinal neovascularization on mid-stage FFA
III. Fluorescent leakage of neovascular of neovascular

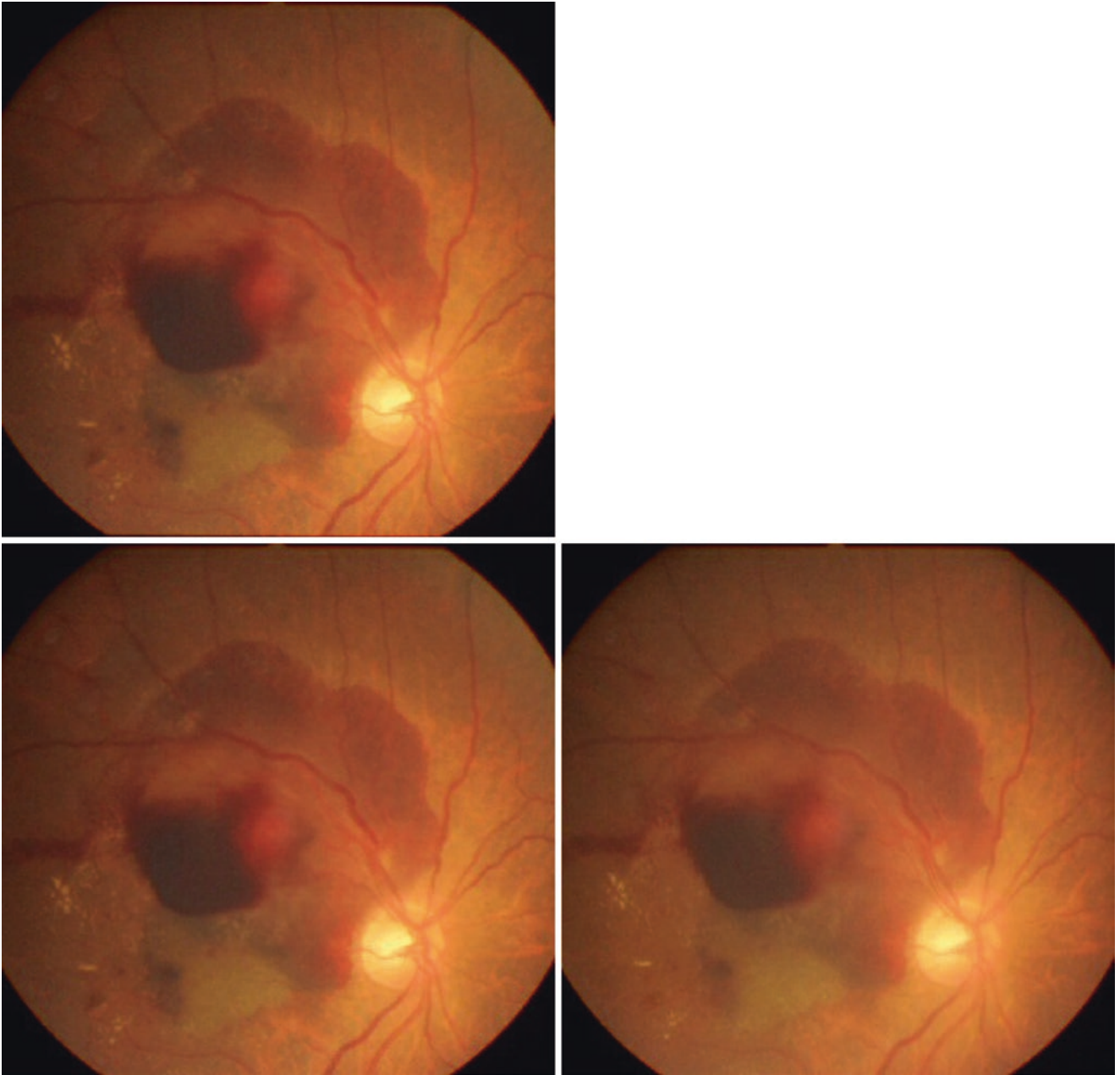


Fig. 3.22 Polypoidal choroidal vasculopathy

- I. Orange elevation
- II. Vitreous hemorrhage

- III. Sub-retinal hemorrhage
- IV. Chronic sub-RPE hemorrhage
- V. Intermediate retinal exudates

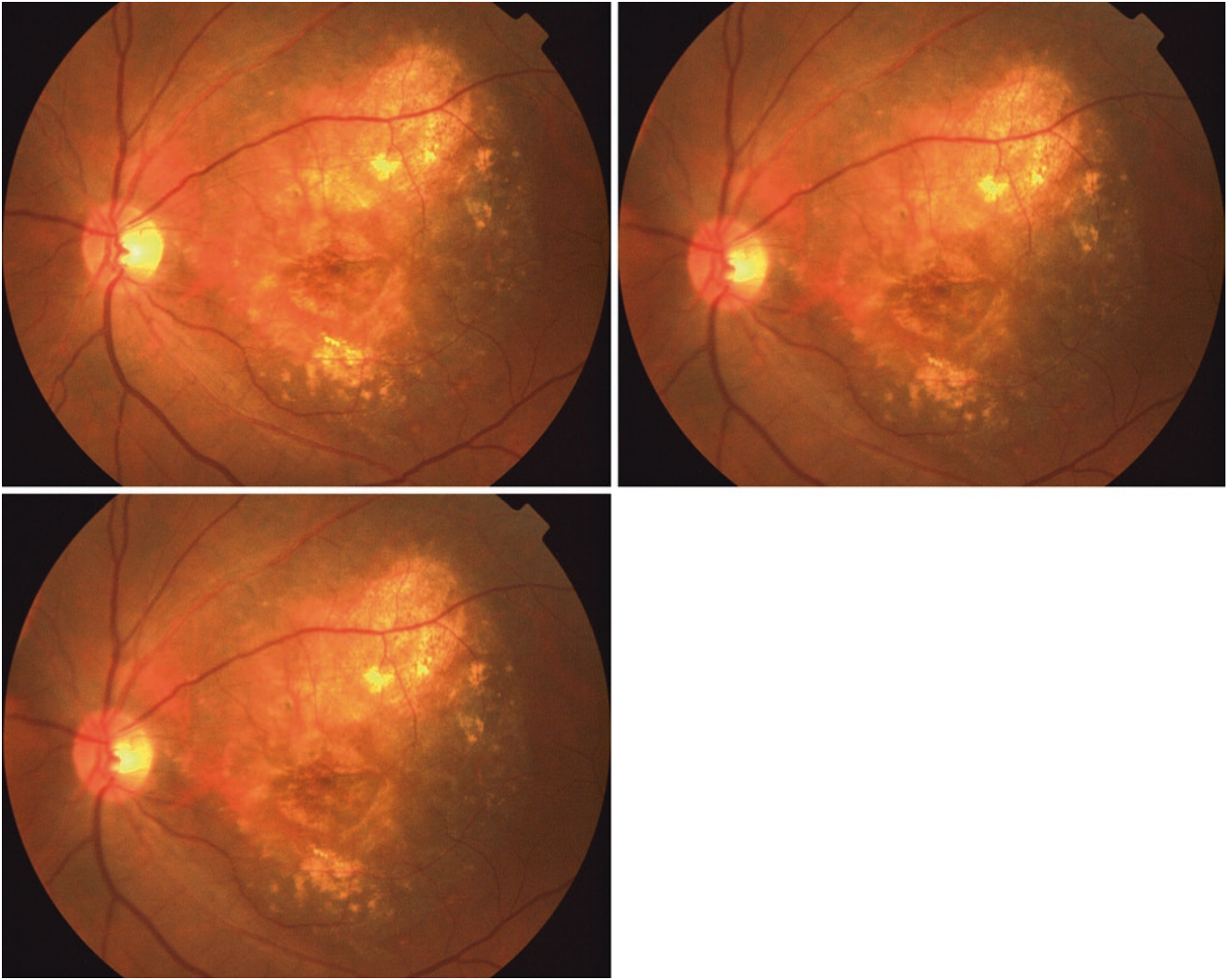


Fig. 3.23 RPE tear
I. Folded RPE in triangle shape

II. Exposed sclera
III. Intra-retinal exudates

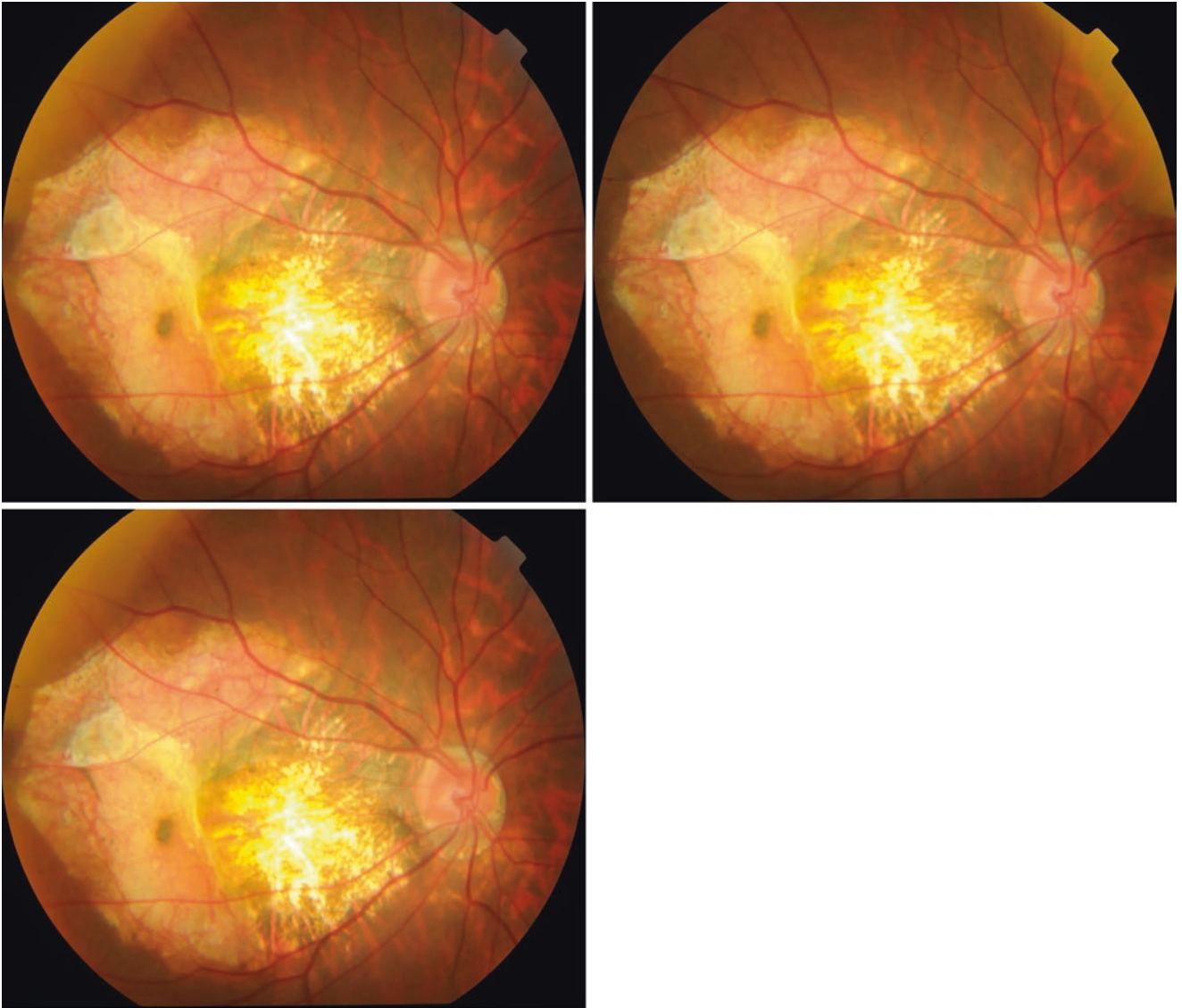


Fig. 3.24 RPE tear
I. Folded RPE or RPE tear
II. Exposed sclera

III. Normal RPE area
IV. Geographic atrophy of macula

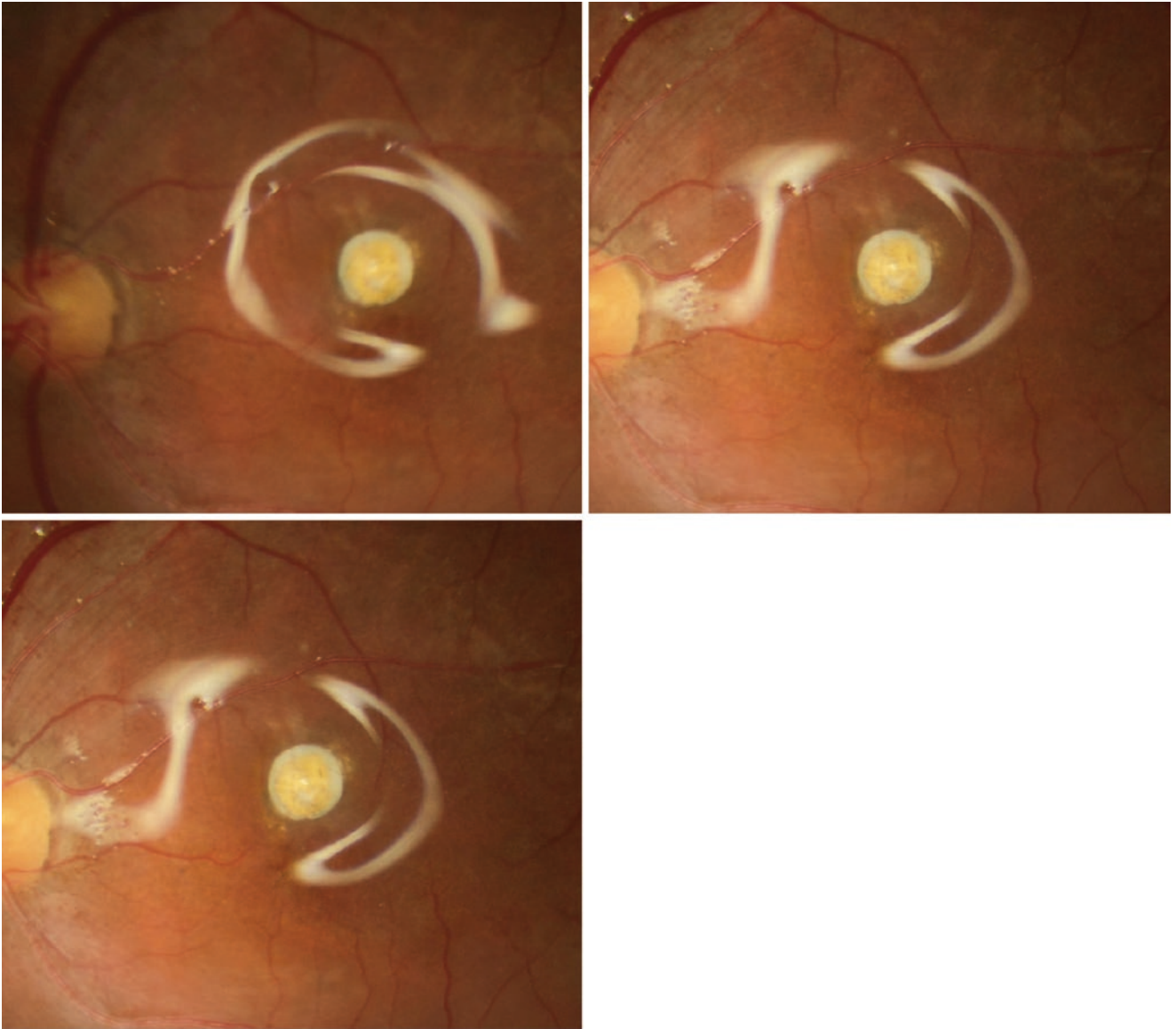


Fig. 3.25 Silicone oil tamponade of PCV
I. Chronic choroidal lesion
II. The retinal artery went over the retinal vein

III. The reflex of silicone oil
IV. Deep retinal hemorrhage

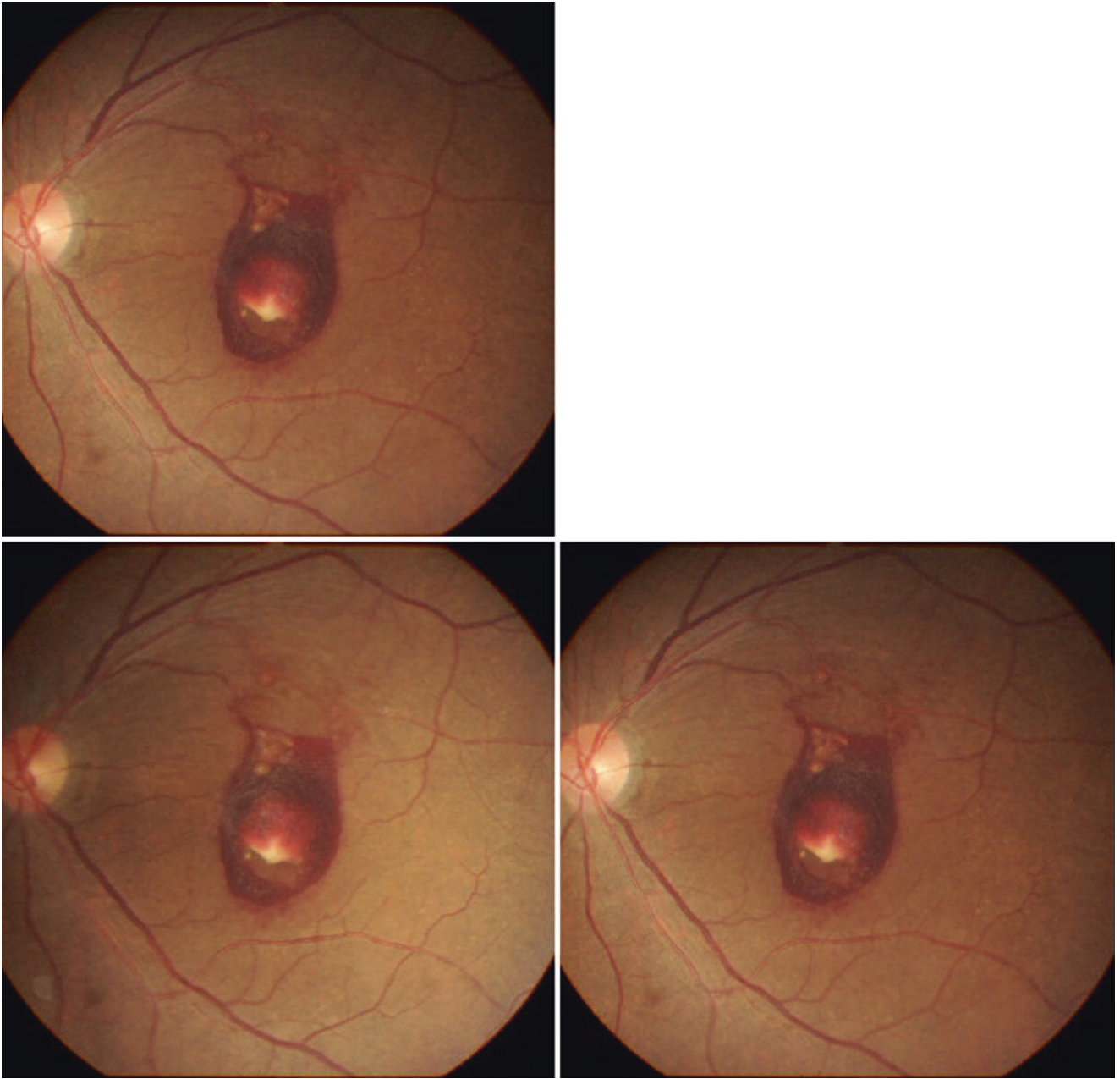


Fig. 3.26 Macular hemorrhage
I. Thickened posterior hyaloid and strong reflex

II. Yellow-white epiretinal hemorrhage
III. Sub-retinal hemorrhage

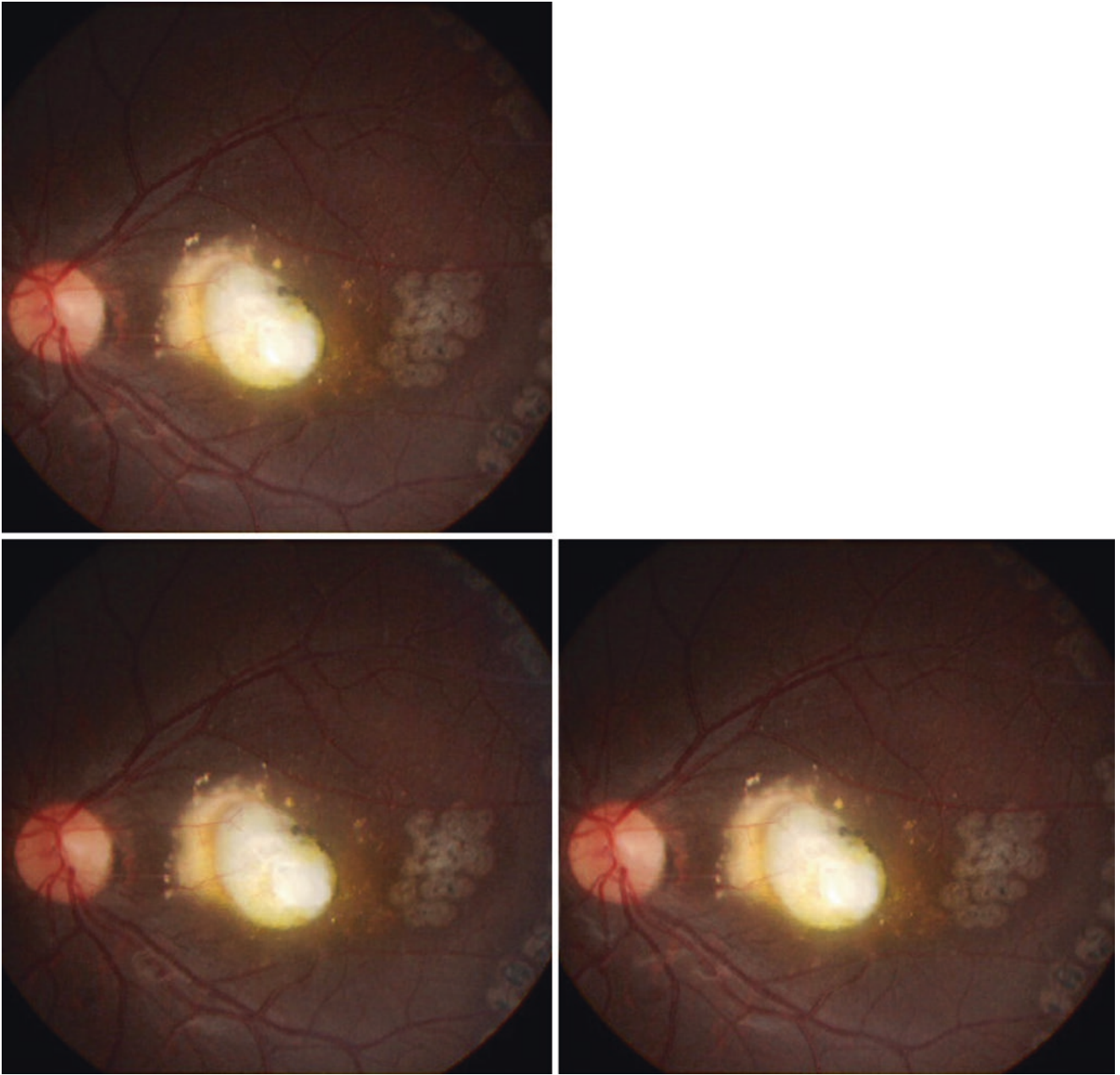


Fig. 3.27 Sub-macular choroidal neovascularization membrane

I. Apex of elevation and appeared white

II. The second layer of exudates

III. The third layer of exudates and appeared yellow-white

IV. Pigment proliferation and small sub-retinal membrane

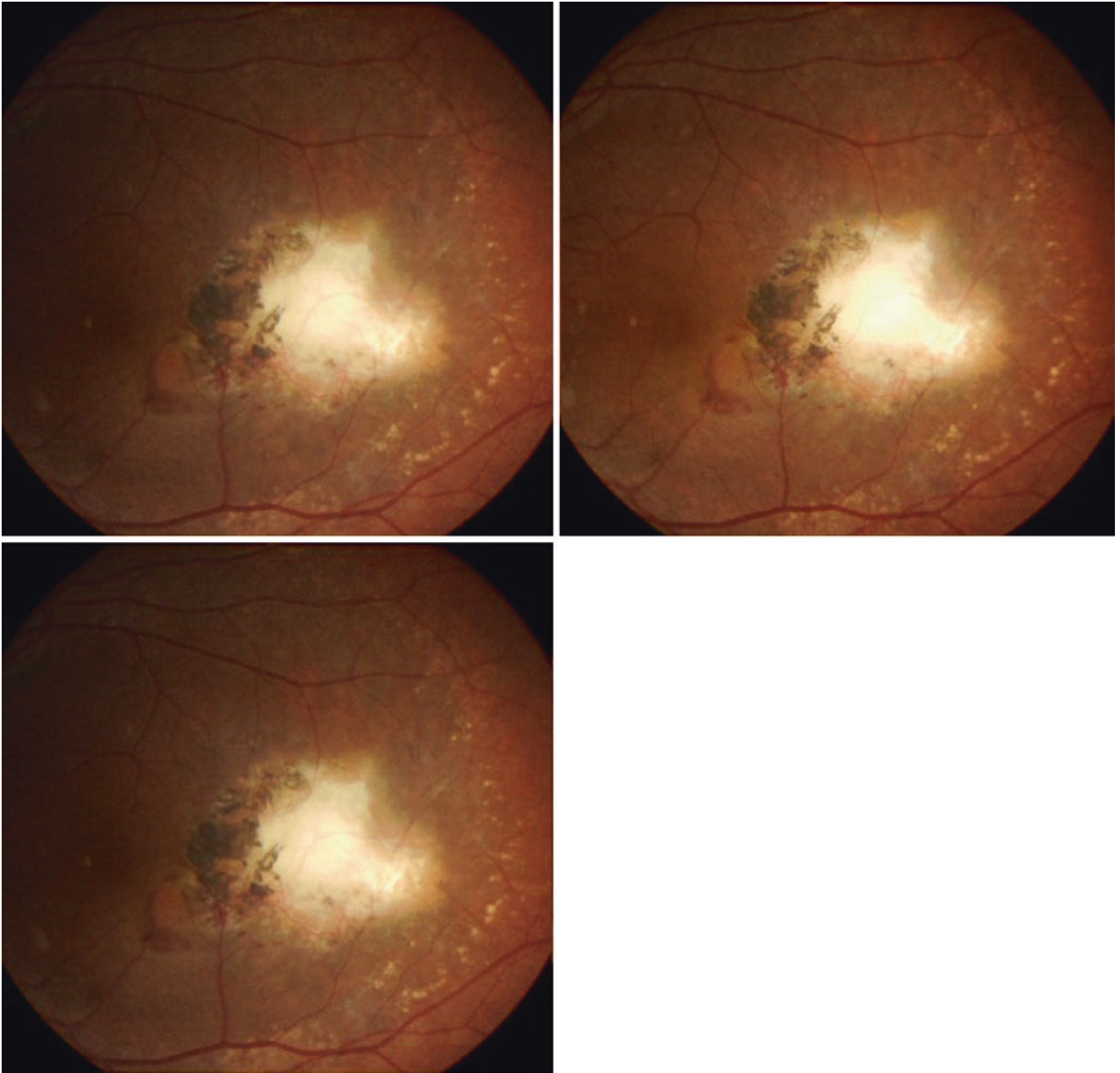


Fig. 3.28 Juxtafoveal sub-retinal mixed choroidal neovascularization

I. Sub-retinal grey scar

II. Sub-retinal choroidal neovascularization

III. Sub-retinal pigment proliferation

IV. Sub-retinal hemorrhage

V. Epiretinal hemorrhage

VI. Scattered sub-retinal dotted exudates

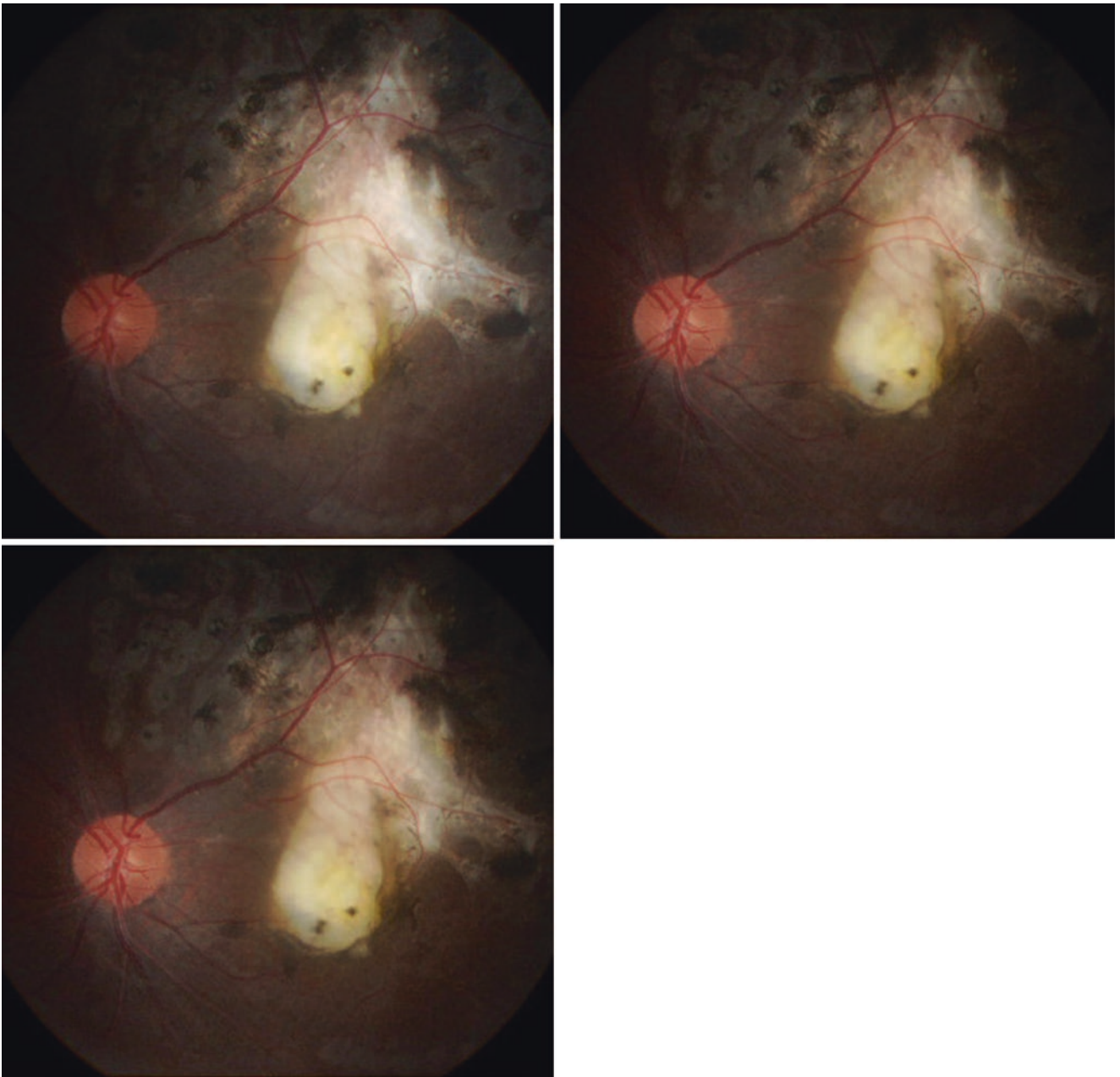


Fig. 3.29 Sub-macular fibrous membrane

I. Apex of sub-retinal membrane

II. Suspending retinal vessels

III. Pigment proliferation

IV. Retinal artery sheath

V. The retinal artery went over the membrane

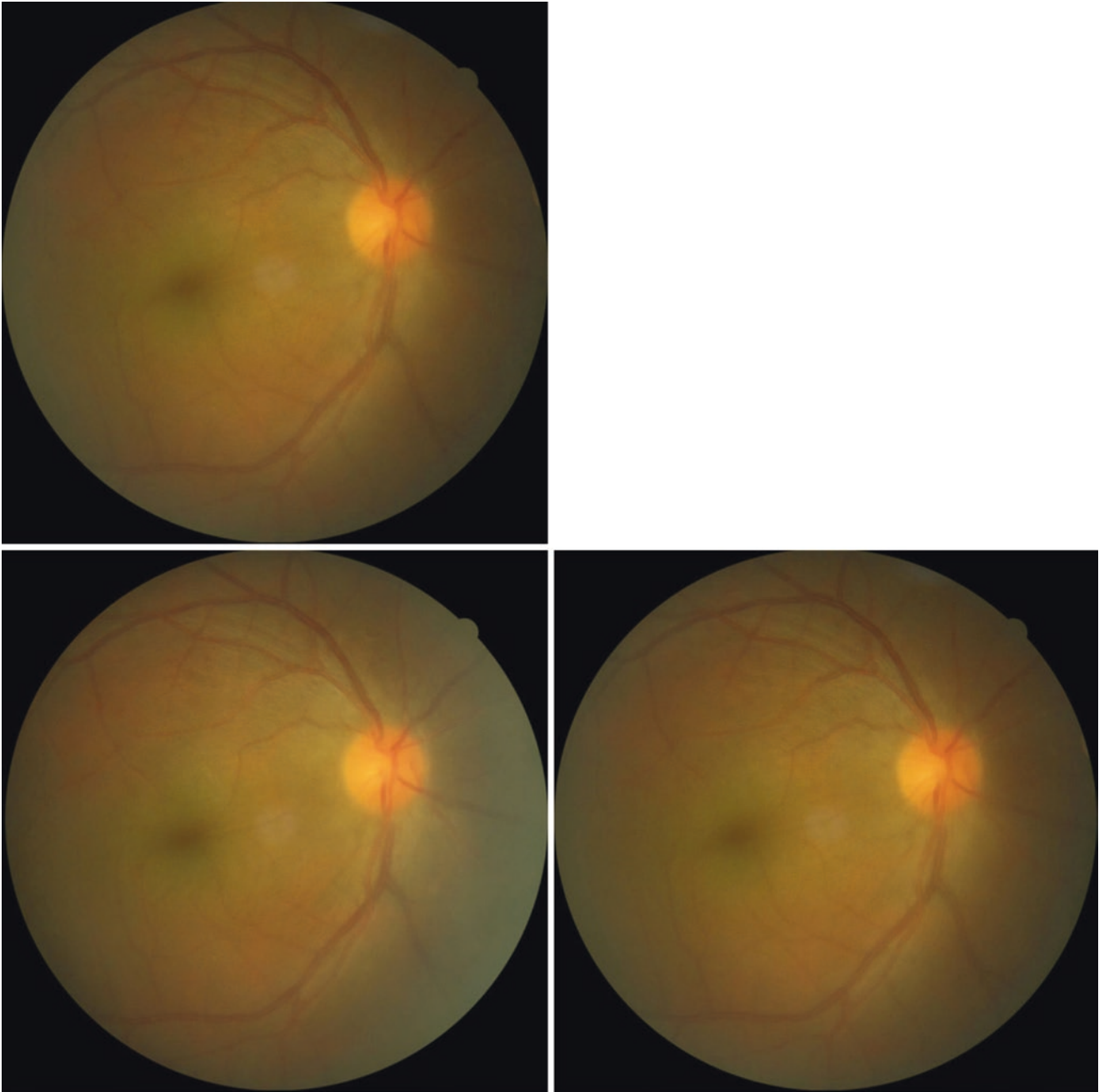


Fig. 3.30 Macular edema
I. Elevation of macula and loss of central reflex

II. The retinal artery went beneath the retinal vein

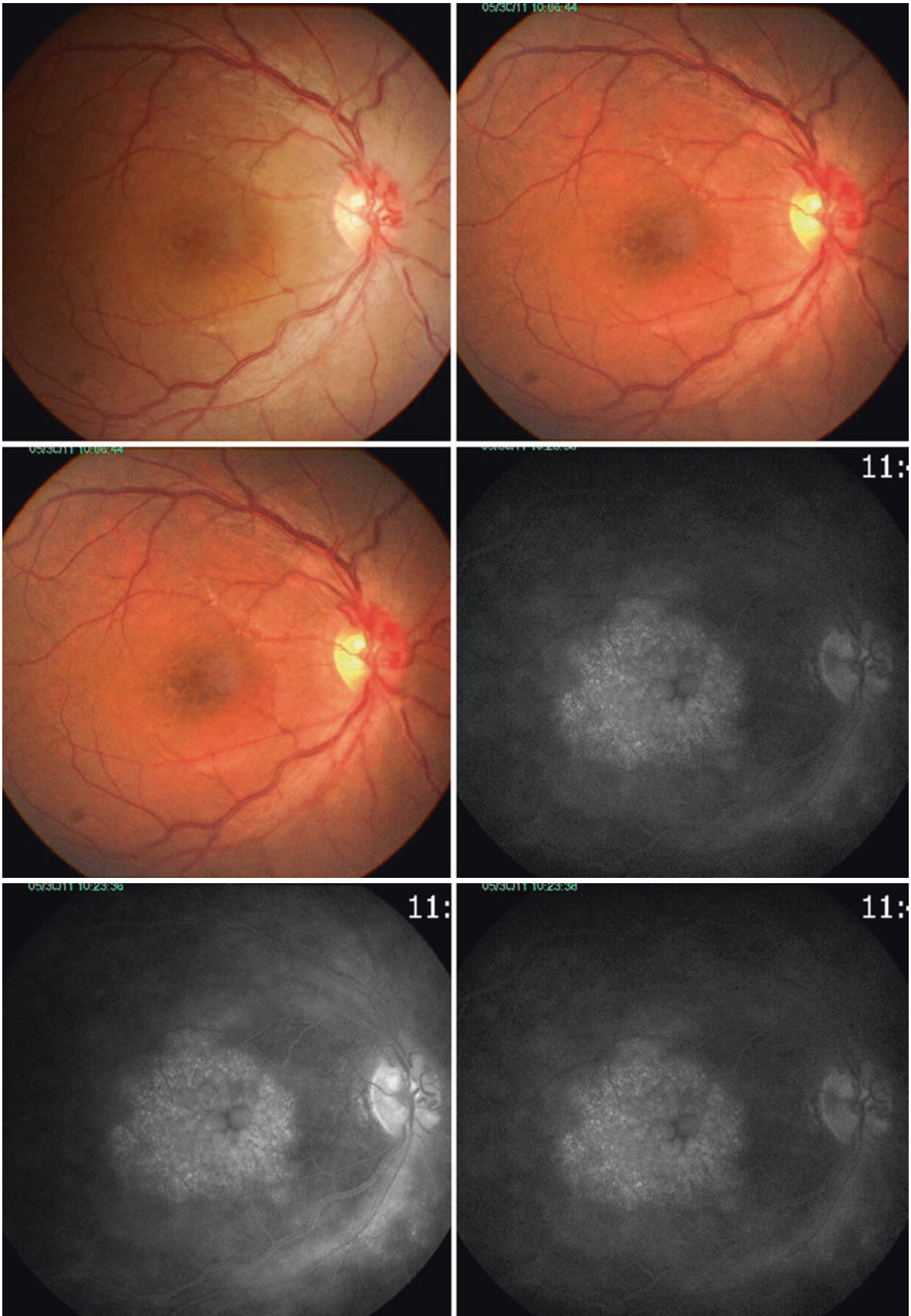


Fig. 3.31 Macular edema
I. Cystoid macular edema

II. Distorted veins on the optic disc
III. Vitreous opacities

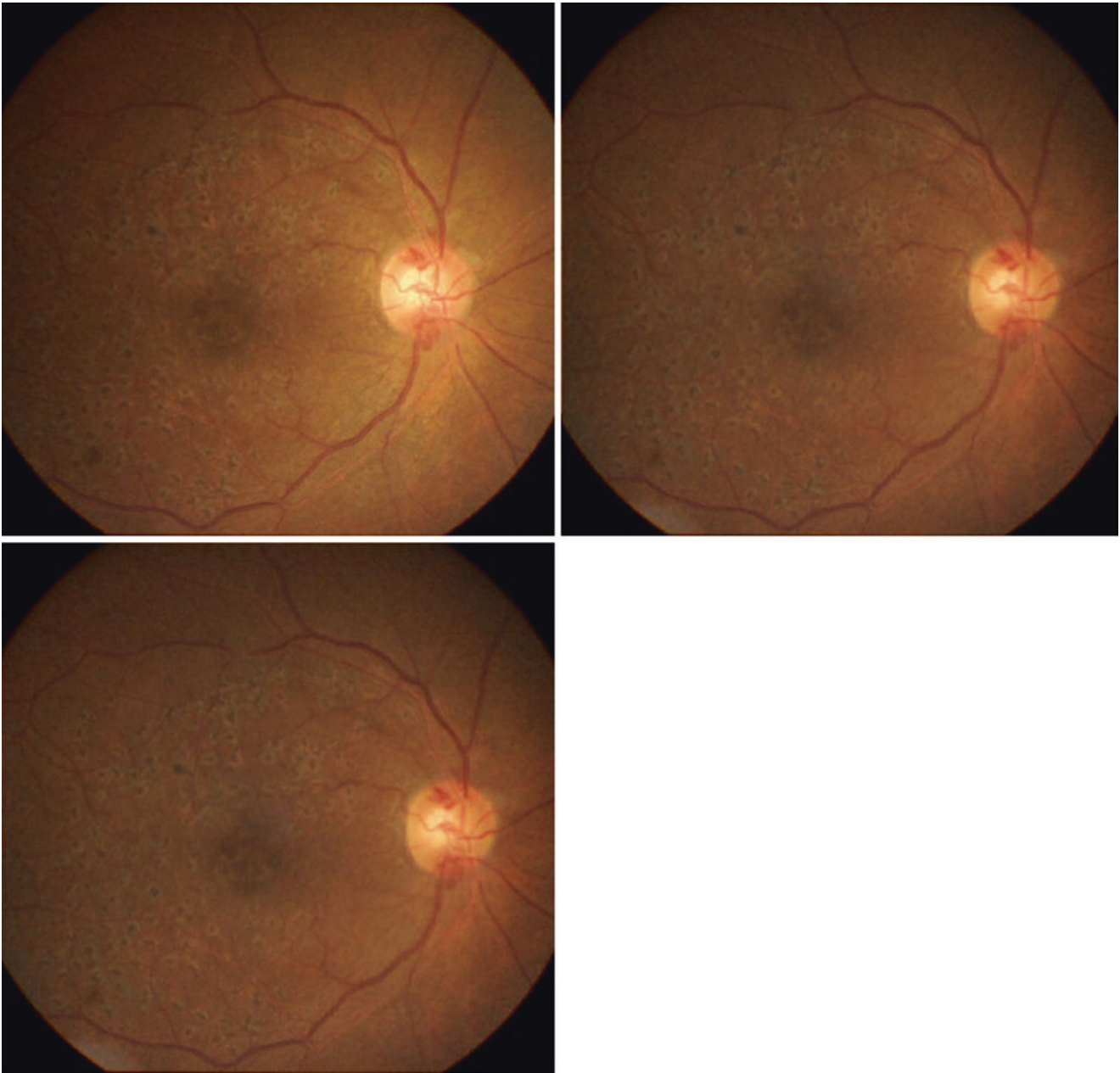


Fig. 3.32 Macular edema after grid laser treatment

I. The macula was flat

II. Laser spot

III. The end of retinal vein was dilated

IV. Neovascularization of the optic disc

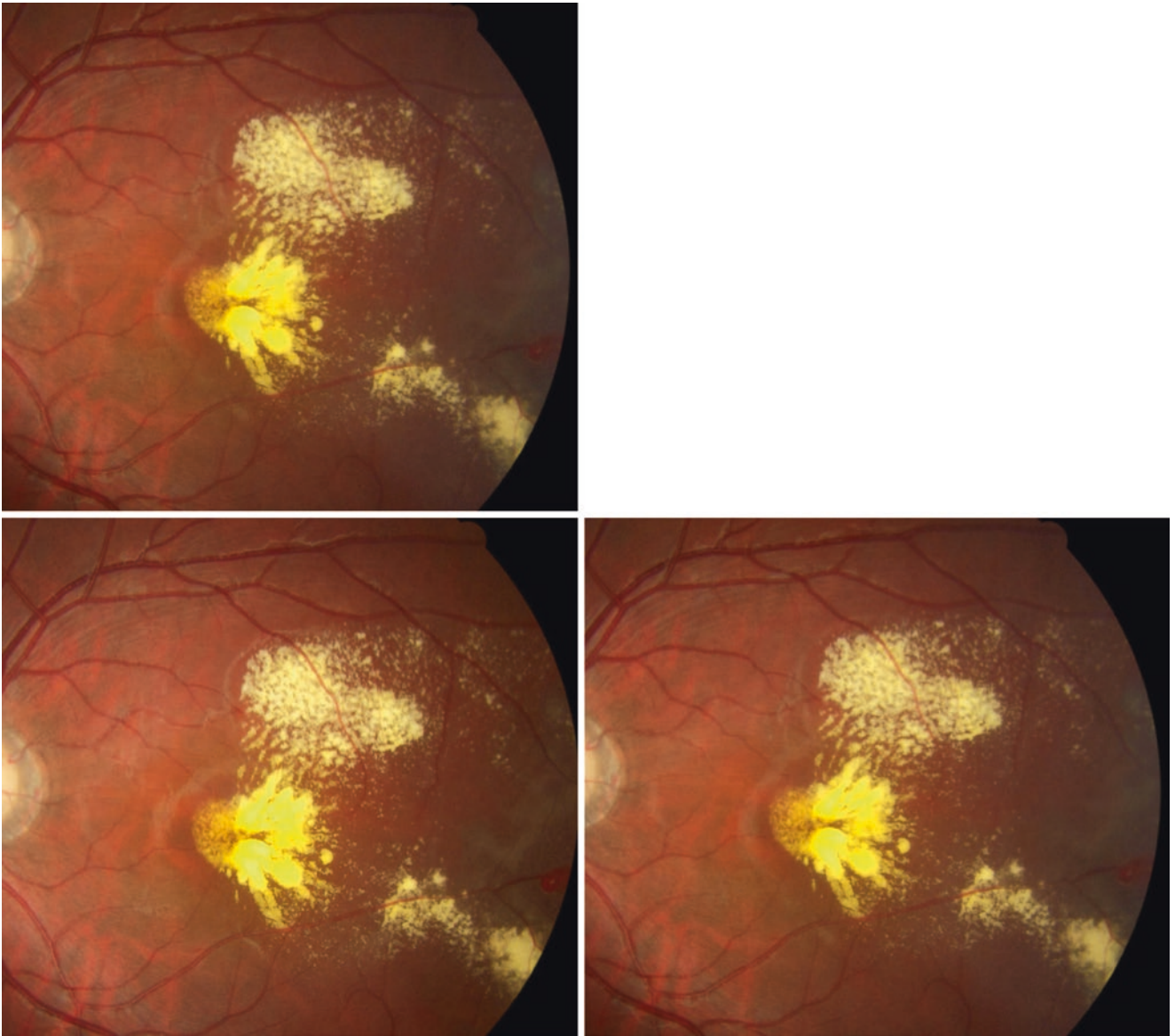


Fig. 3.33 Macular radial hard exudates
I. The end of retinal artery was dilated
II. Superficial retinal exudates

III. Deep retinal exudates
IV. Grey-whitish exudates

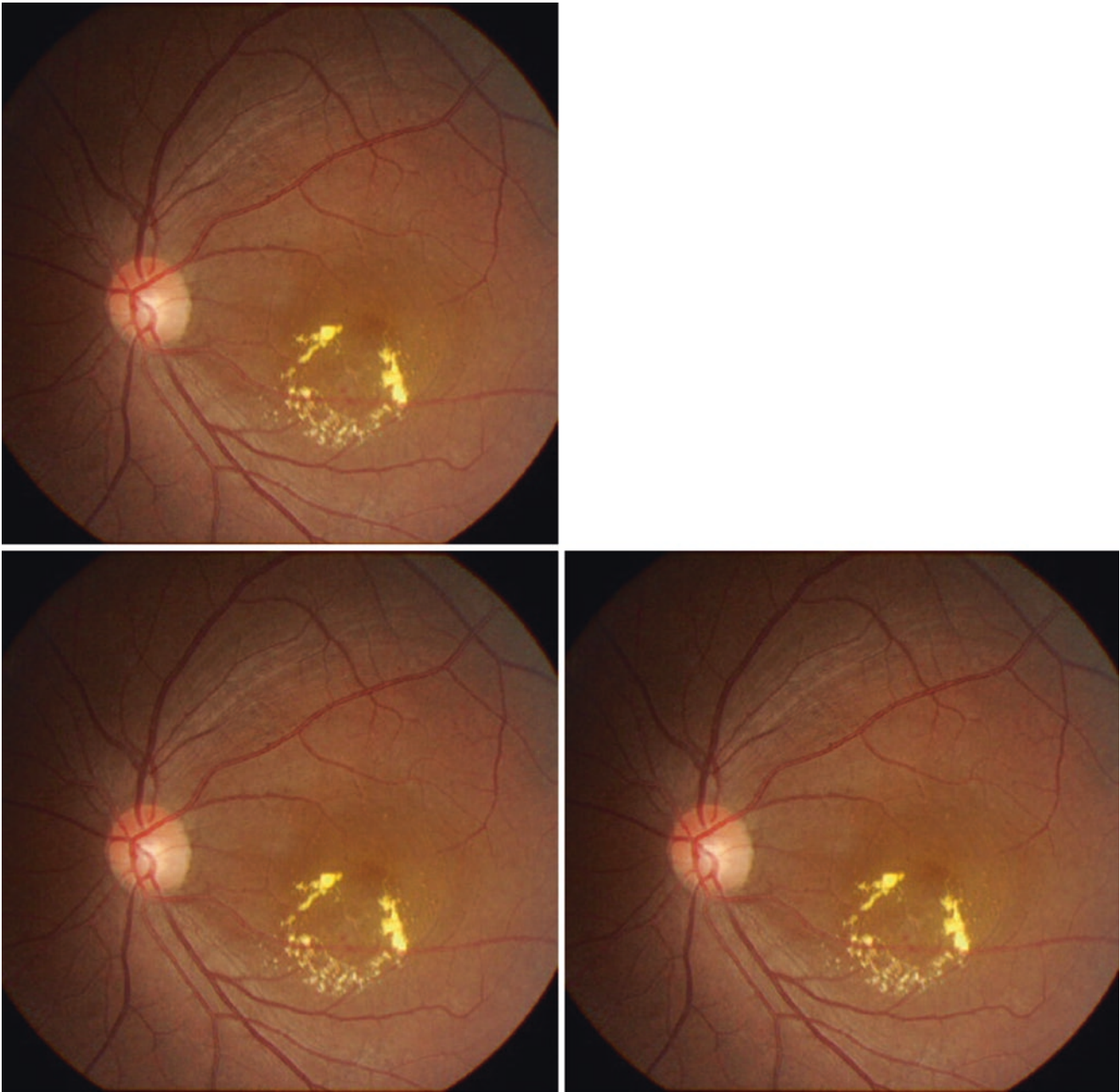


Fig. 3.34 Juxtafoveal telangiectasis
I. The area of macular edema

II. The deep retinal vessels were dilated
III. Intermediate retinal exudates

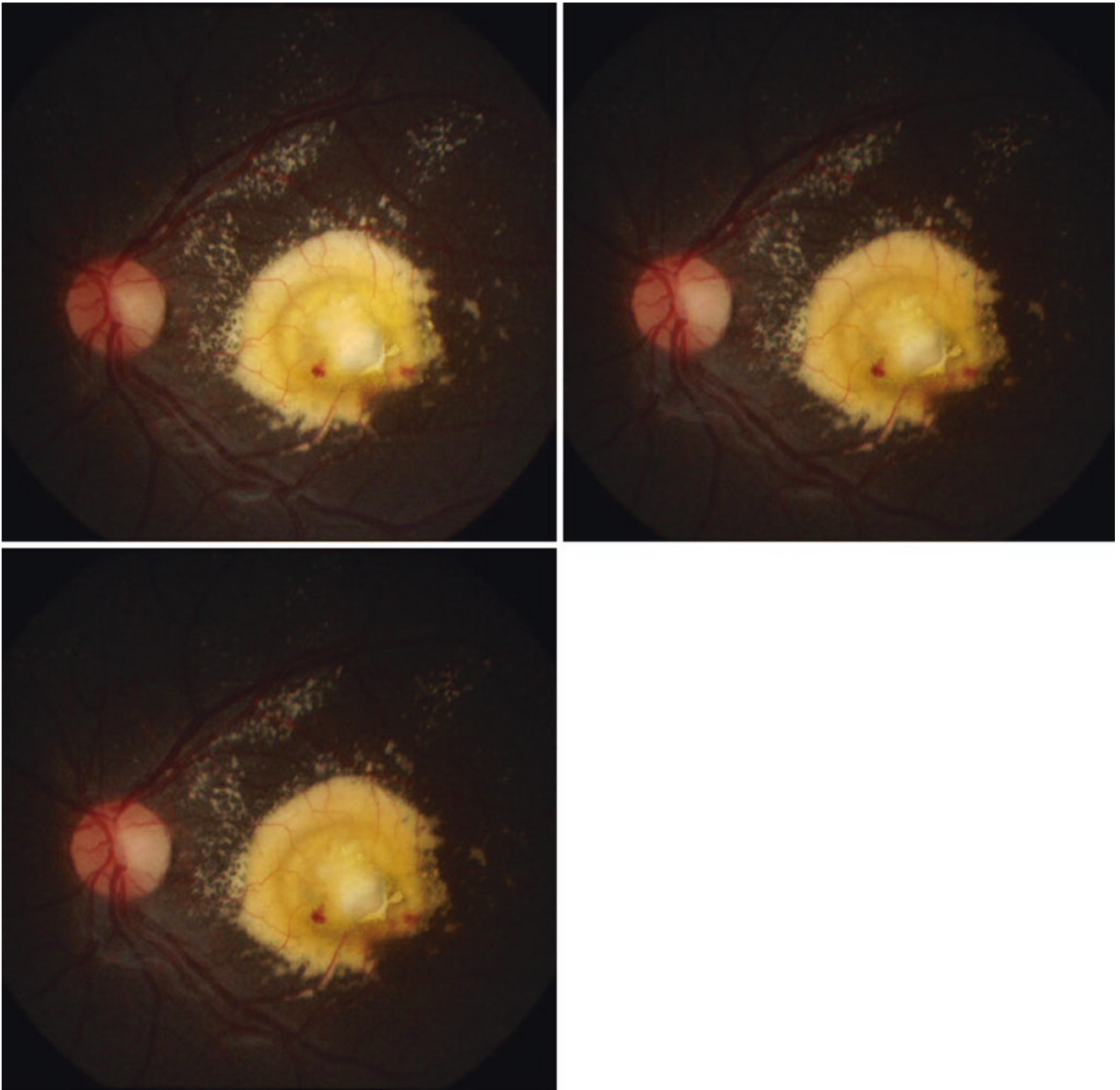


Fig. 3.35 Adult Coats' disease

- I. The lesion was elevated like three layers of cake
- II. The blood supply of the retinal artery was insufficient than the other retinal branches

III. Suspected abnormal vessels

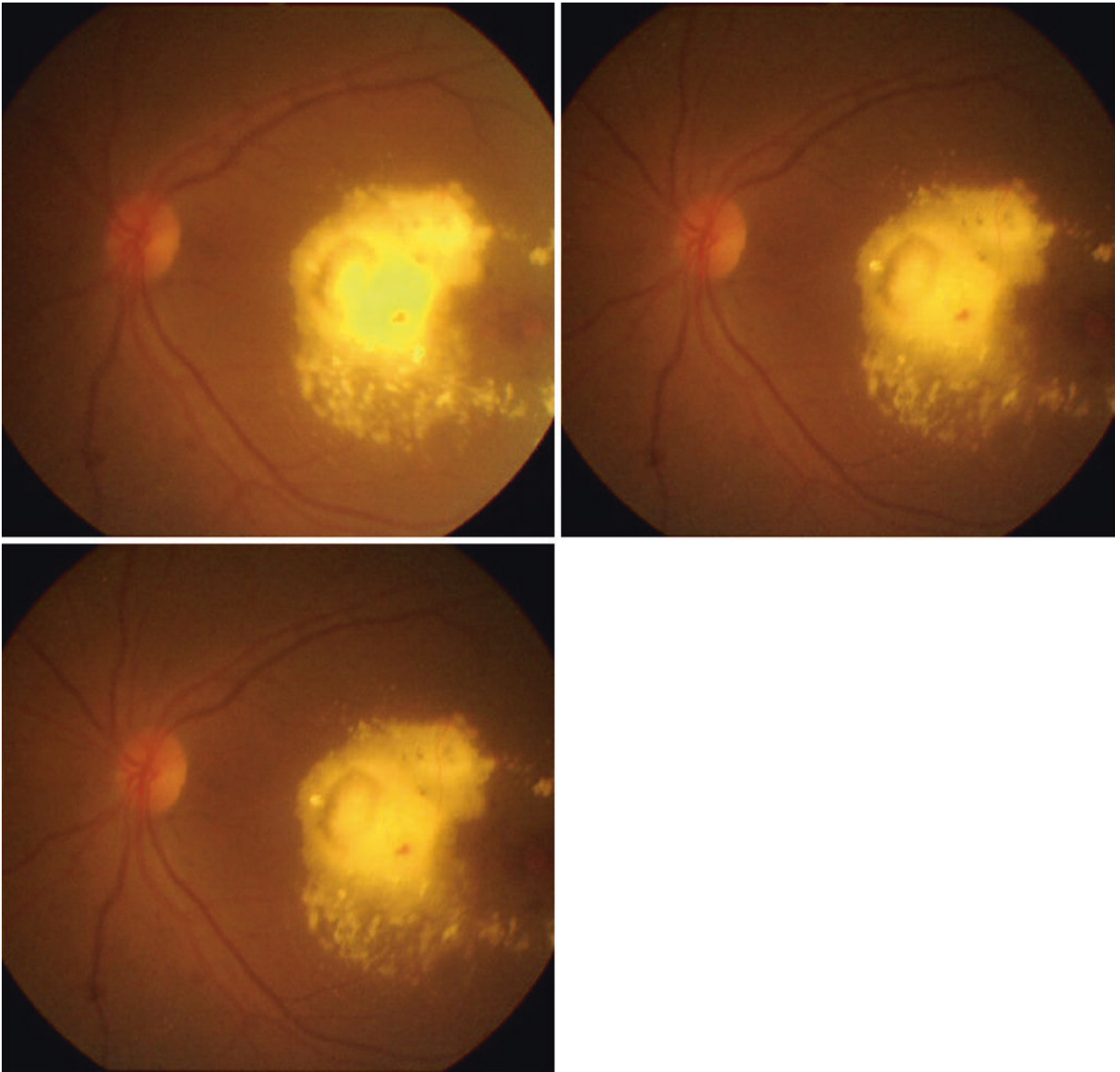


Fig. 3.36 Adult Coats' disease of the macula
I. Superficial retinal exudates

II. Large amount of yellow-white exudates and crystal in the deep retina
III. Deep retinal hemorrhage

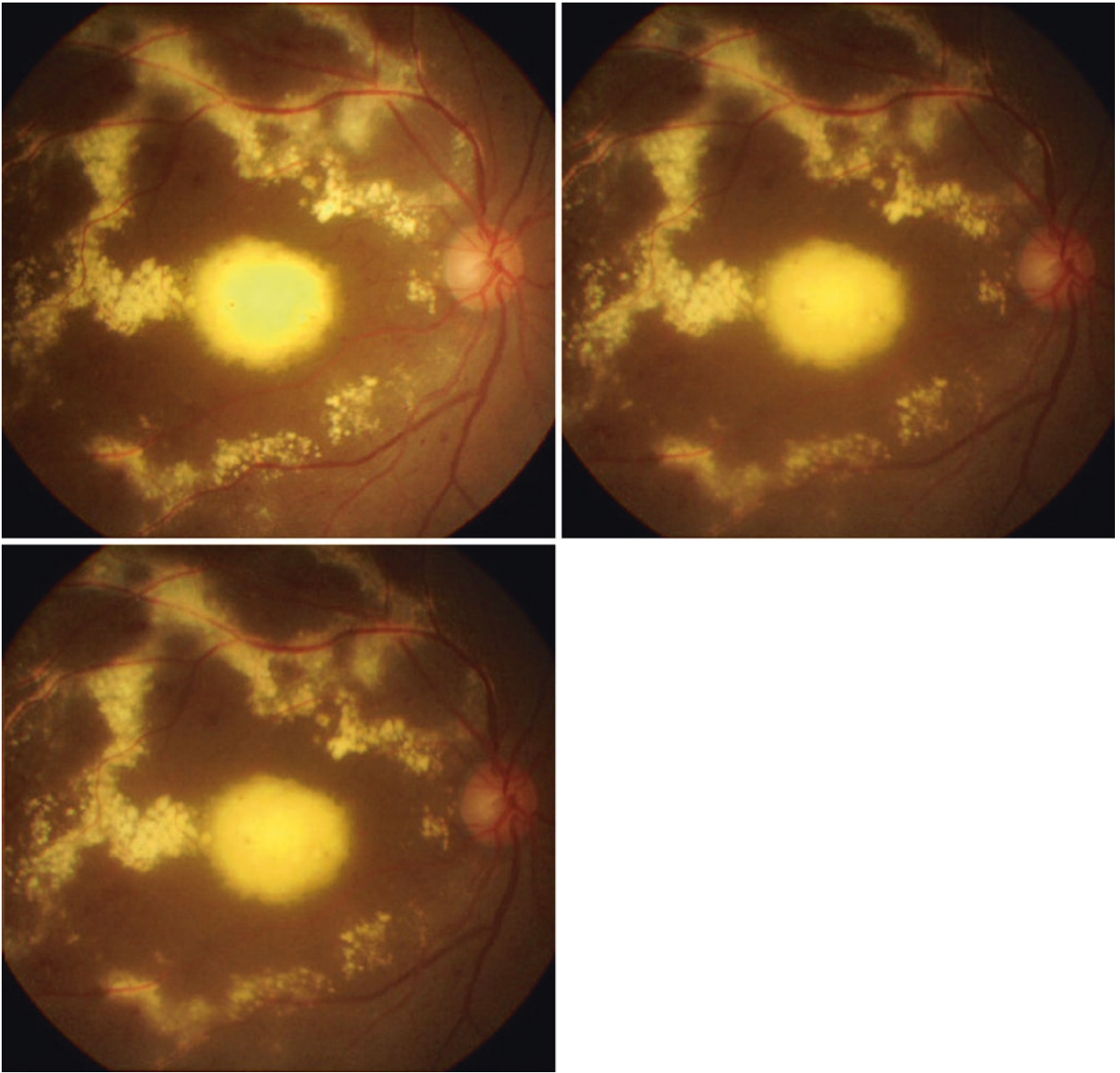


Fig. 3.37 Coats' disease

- I. Yellow-whitish sub-retinal exudates
- II. Exudates of the retinal artery

- III. Irregular diameter and exudates of retinal veins
- IV. The end of the retinal vessels was dilated

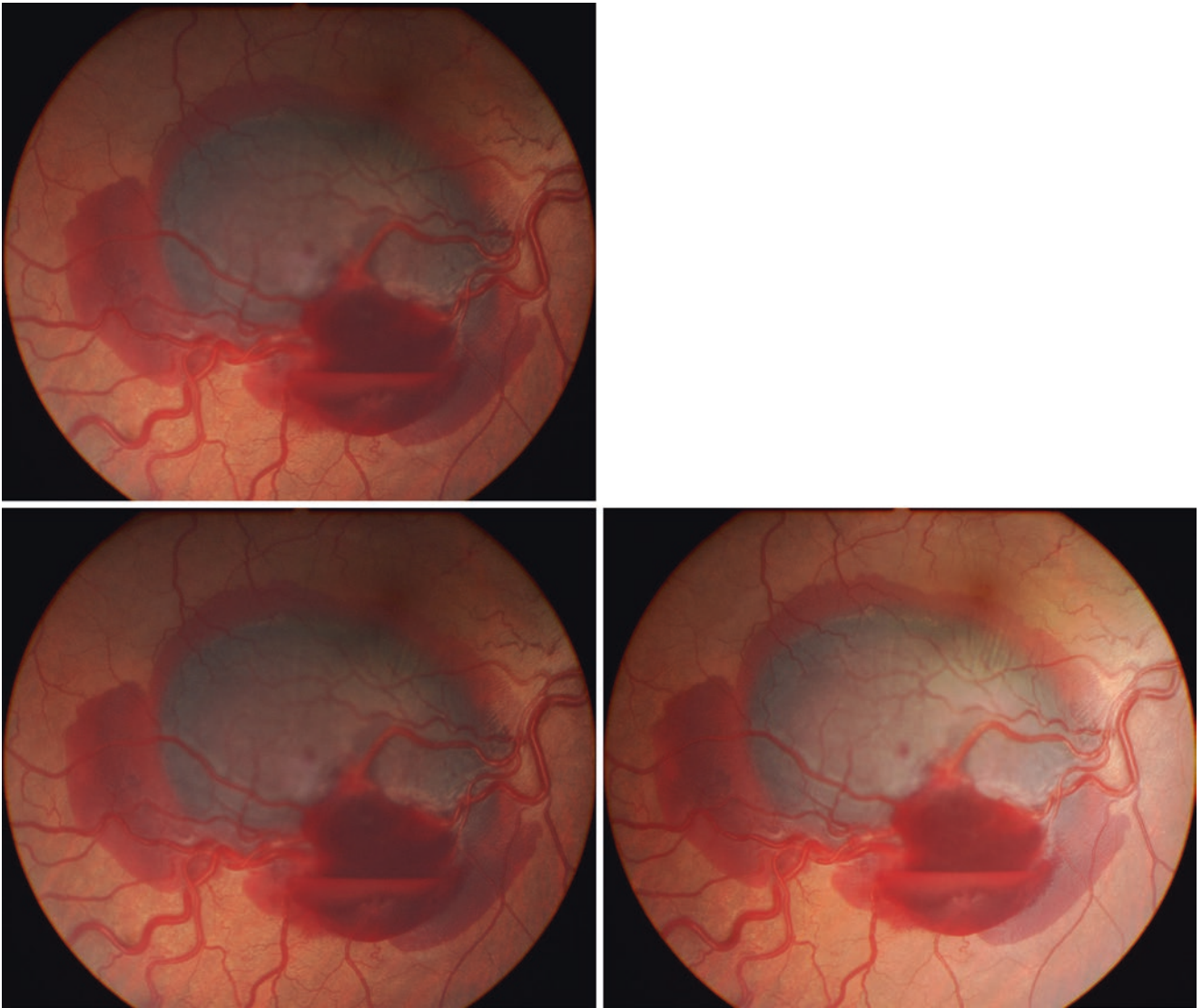


Fig. 3.38 Macular hemorrhage in various layers
I. Pre-retinal hemorrhage looked alike a boat
II. Sub-retinal hemorrhage

III. Sub-RPE hemorrhage
IV. The retinal vessels were distorted and dilated

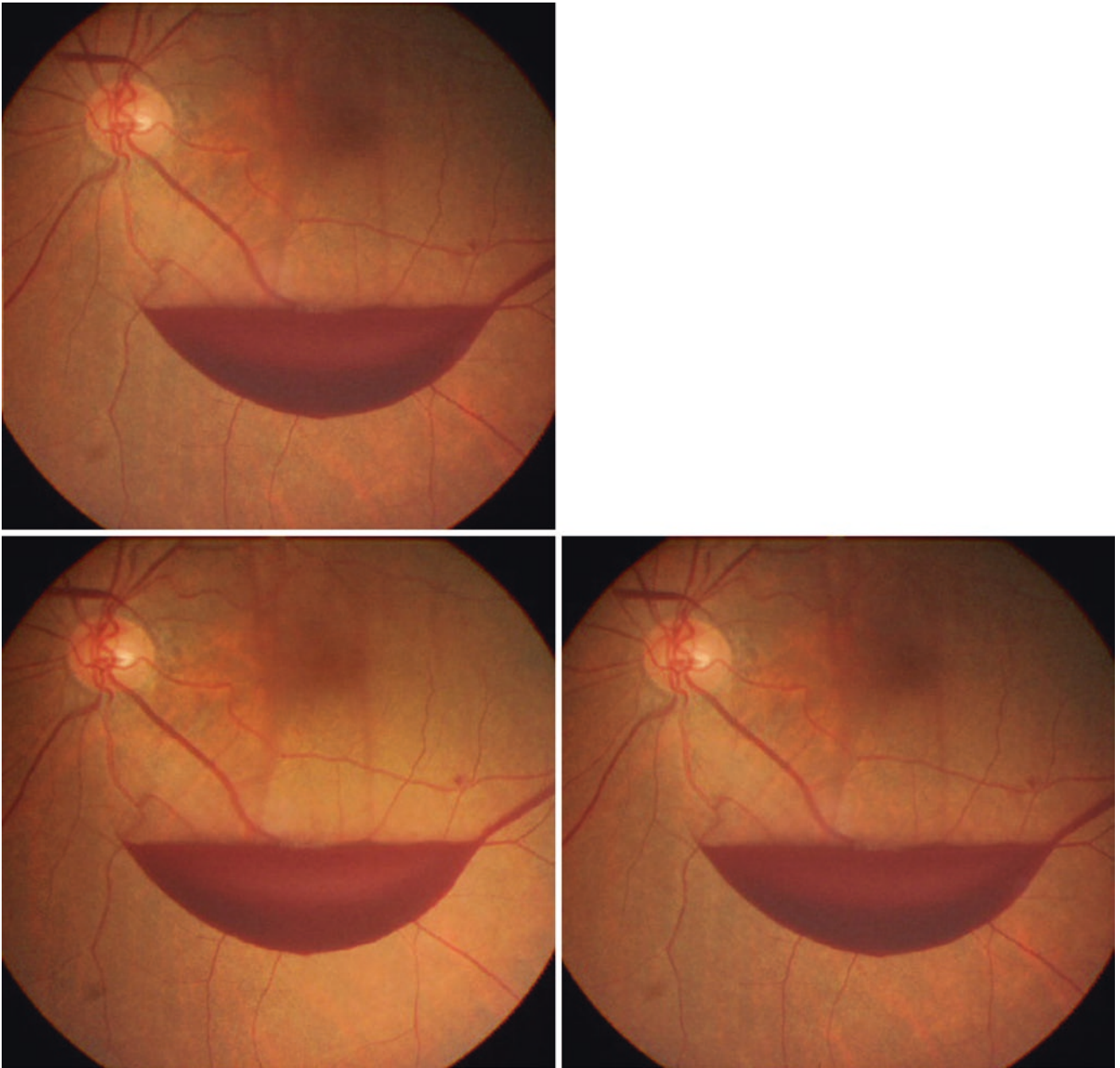


Fig. 3.39 Boat-like epiretinal hemorrhage
I. The serum
II. The platelets

III. The white blood cells
IV. The deoxygenated hemoglobin
V. The oxygenated red blood cells

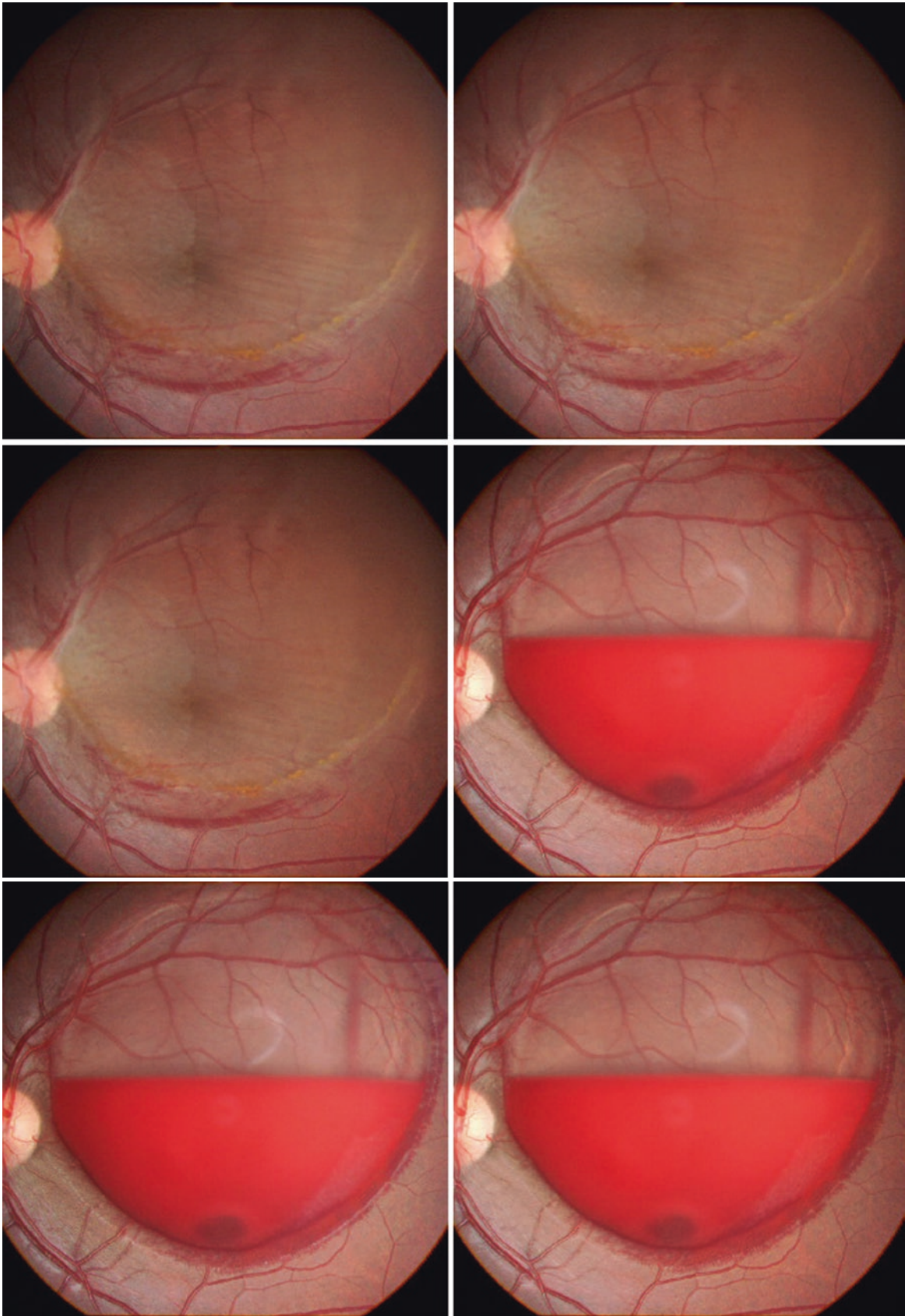


Fig. 3.40 Epiretinal hemorrhage

I. The boat-like epiretinal hemorrhage like a dome
II. After dissection of posterior limiting membrane by Nd:YAG laser,
the hemorrhage was disseminated and absorbed

III. Thickened posterior hyaloids and folds

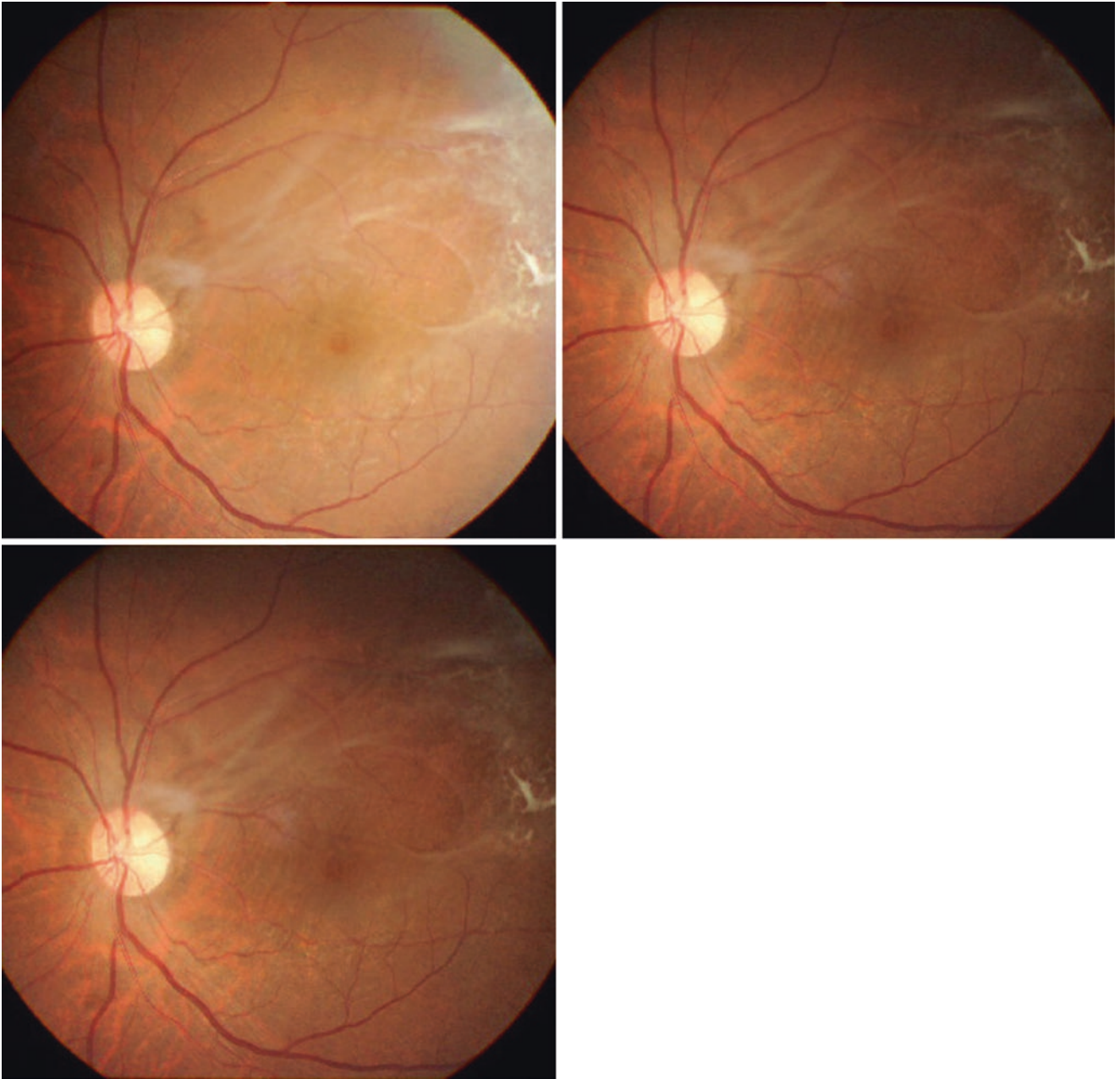


Fig. 3.41 Proliferative vitreoretinopathy
I. Proliferative membrane and streaks
II. Pseudo hole

III. Atherosclerosis of superior temporal retinal artery



Fig. 3.42 Epi-retinal membrane

- I. Epi-retinal membrane extended from the optic disc to the periphery
- II. Distorted retinal veins

- III. Irregular diameter of the retinal vein

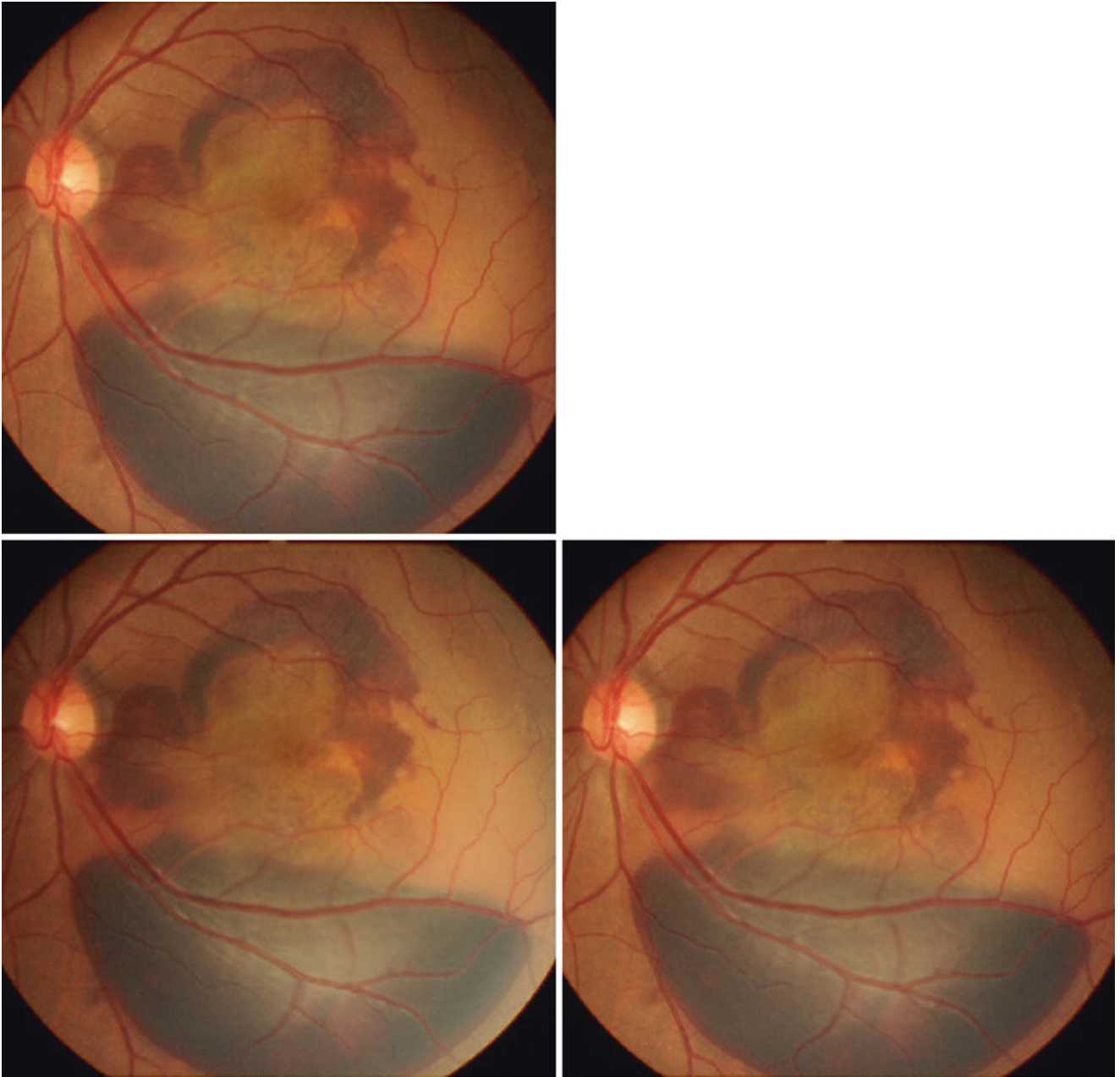


Fig. 3.43 Retinal hemorrhage of different retinal layers
I. Sub-RPE hemorrhage, the lesion was highly elevated
II. Deep retinal hemorrhage

III. Sub-retinal hemorrhage
IV. The central reflex was lost

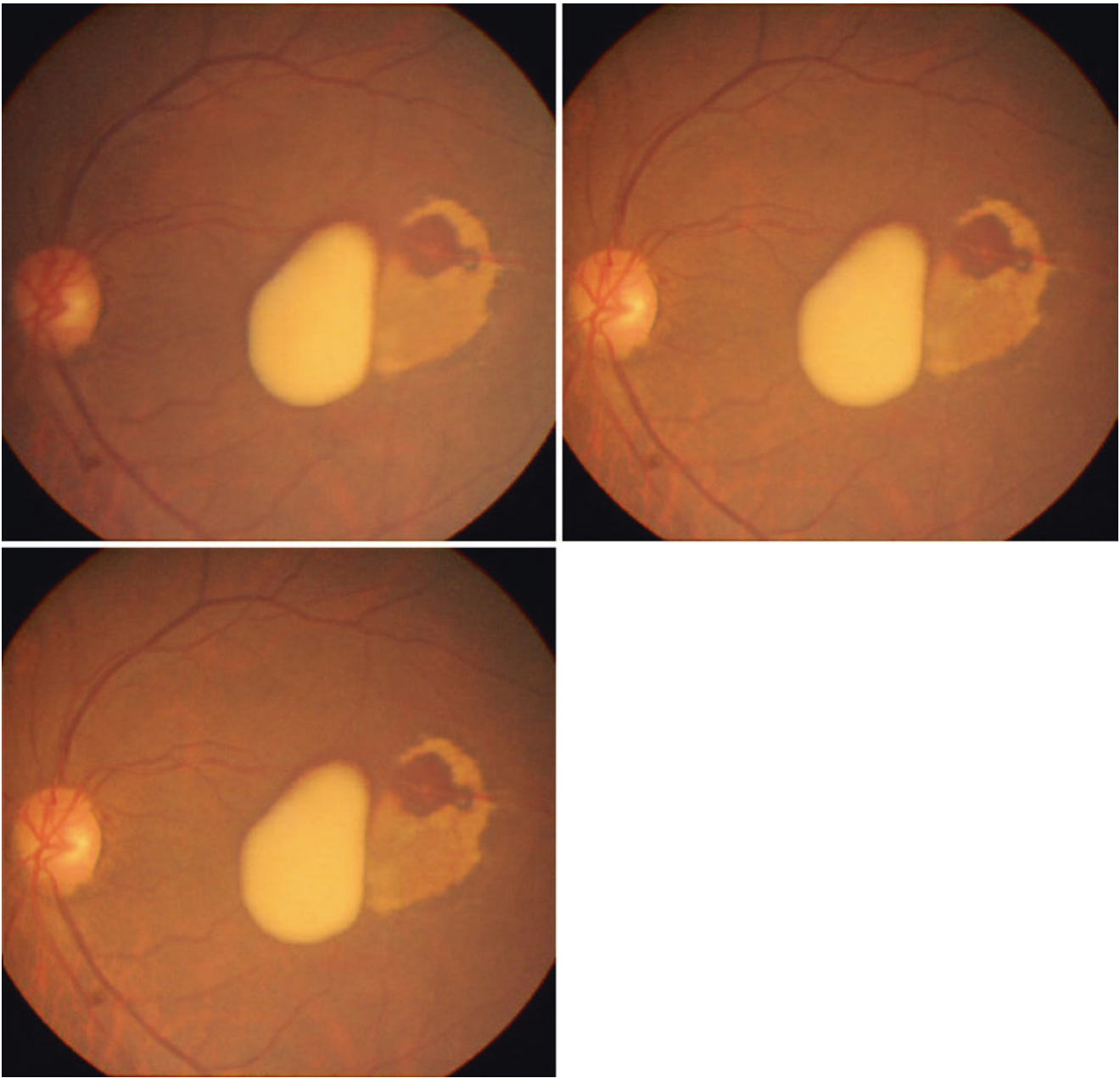


Fig. 3.44 Chronic retinal hemorrhage
I. Pre-retinal hemorrhage
II. Suspected location of retinal macroaneurysm

III. Intermediate retinal hemorrhage
IV. Artery-Vein nicking (Salus Sign)

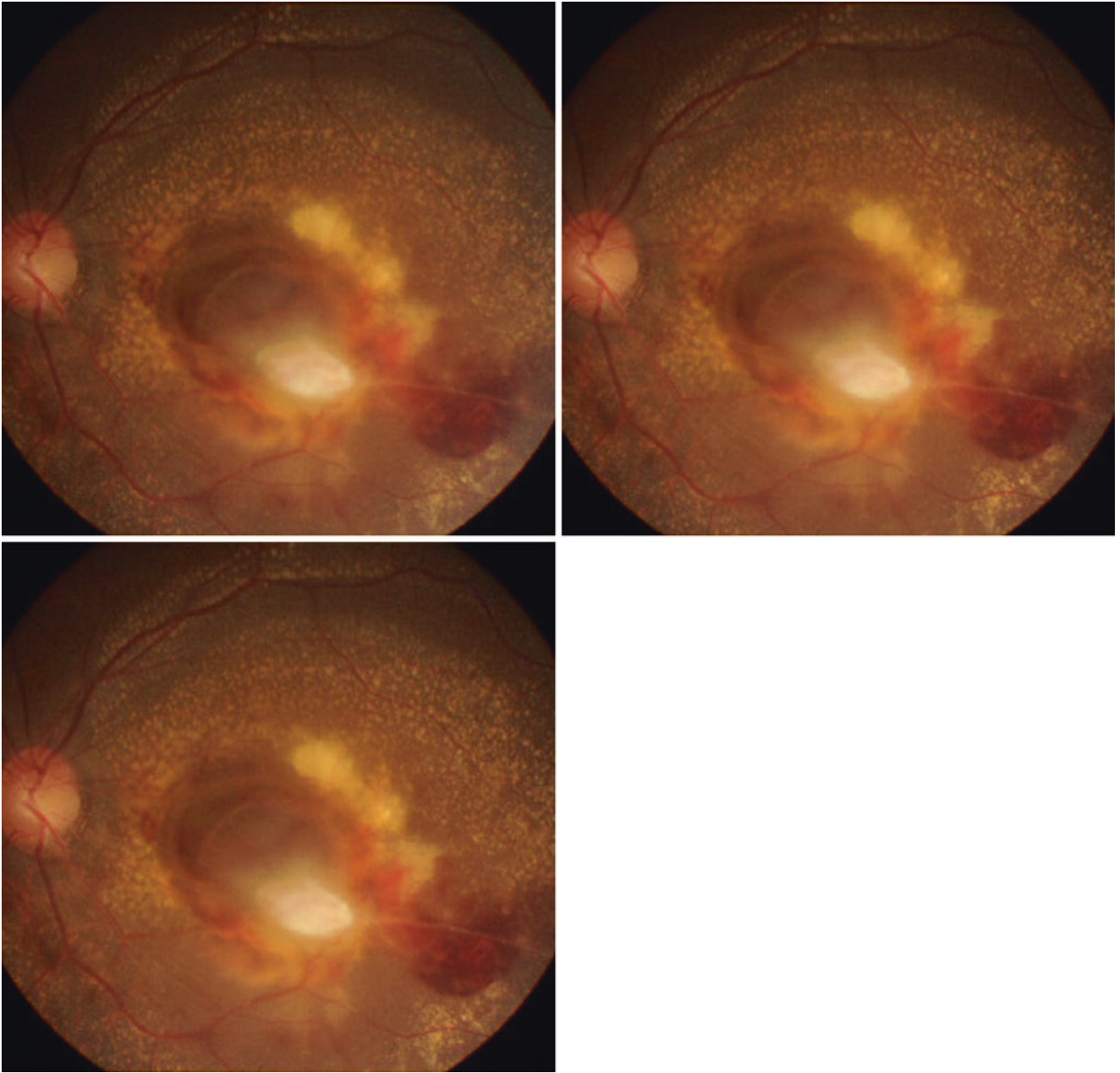


Fig. 3.45 Sub-retinal parasitic infection

I. Suspected scolex of the parasite

II. Intra-retinal reaction of multiple retinal layers

III. Exudative retinal detachment

IV. Dotted yellow-whitish exudates

V. Ghost vessel of retinal vein

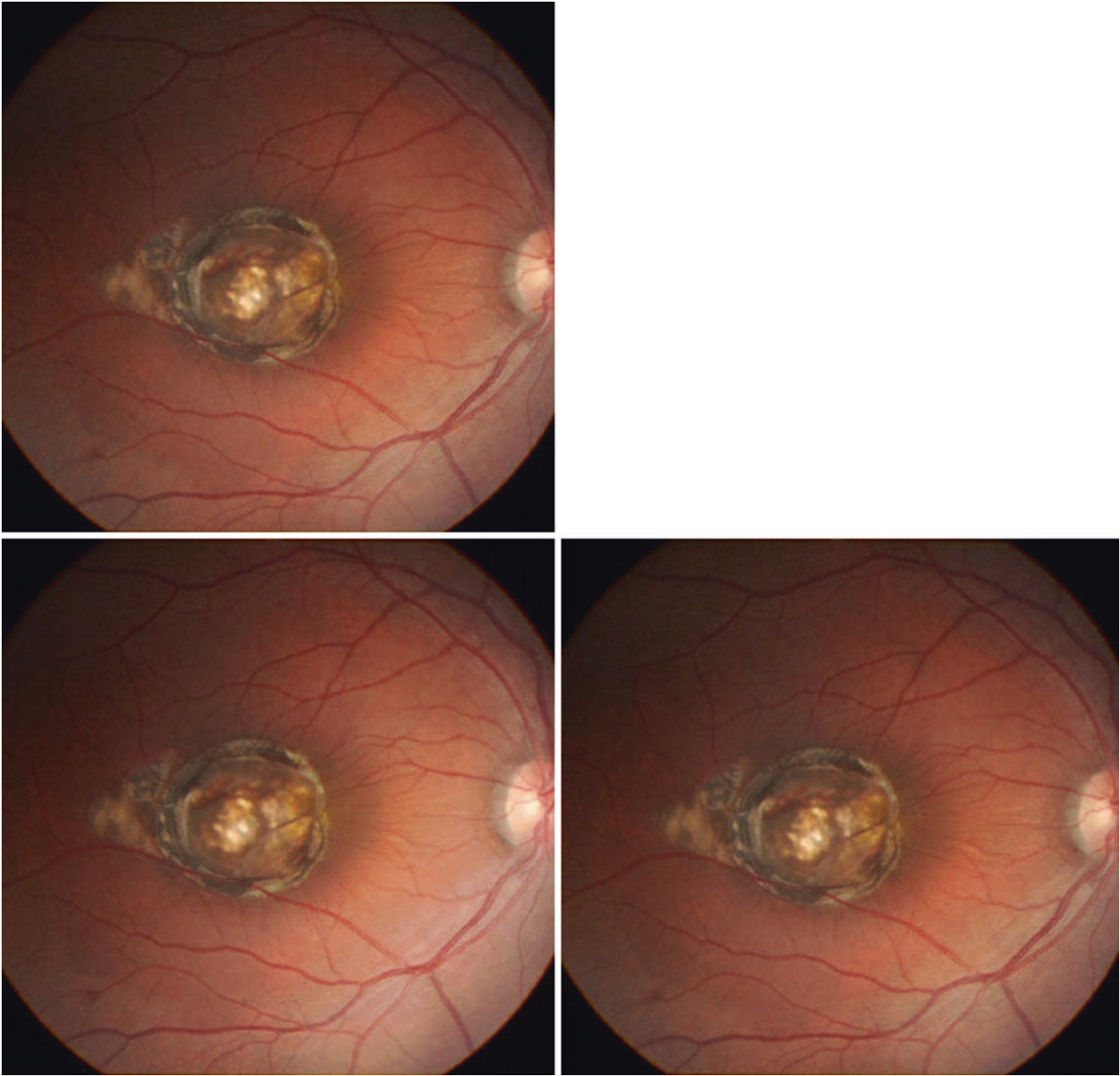


Fig. 3.46 Coloboma of macula

- I. Exposed sclera
- II. Choroidal vessels

- III. Boundary of coloboma
- IV. Impending retinal vessels

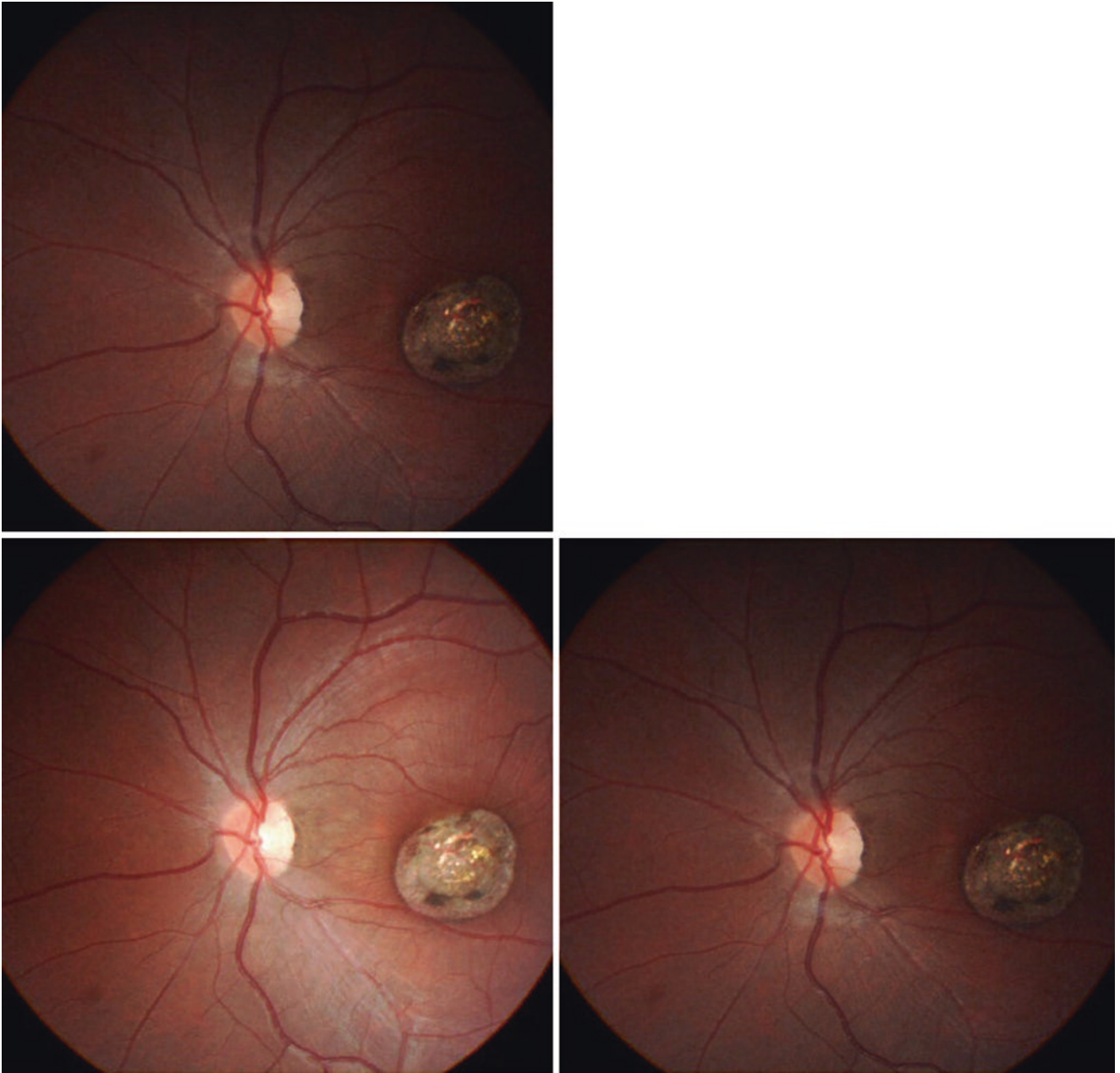


Fig. 3.47 Congenital coloboma of macula
I. The sclera was exposed in the area of coloboma
II. Suspending choroidal vessels

III. Mottled epithelial pigment proliferation
IV. The boundary of coloboma

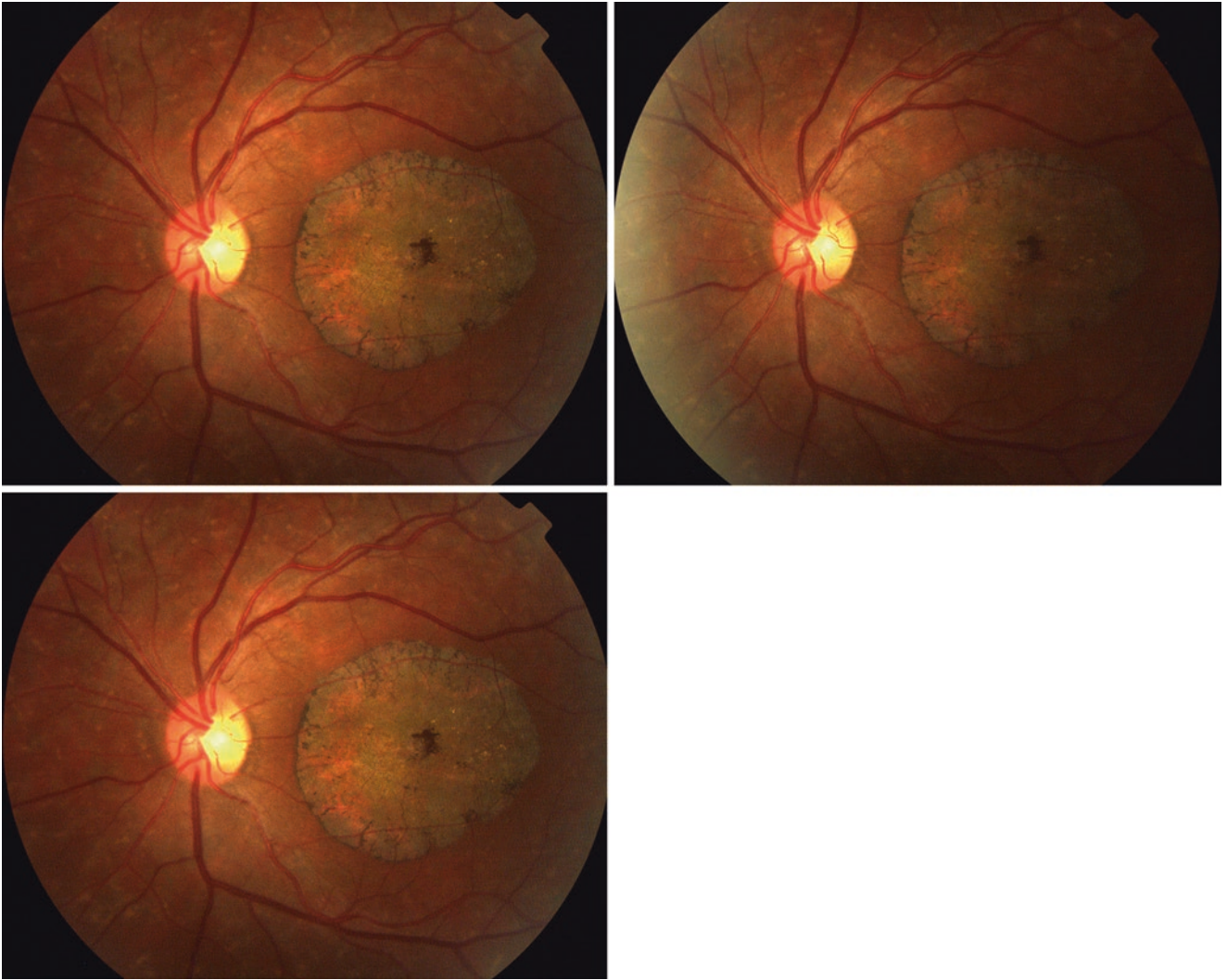


Fig. 3.48 Stargardt disease

- I. Bull-eye shaped lesion, irregular with pigmentation
- II. Retinal and choroidal atrophy like a basin

III. Retinal vessels that passed through the lesion went attenuated

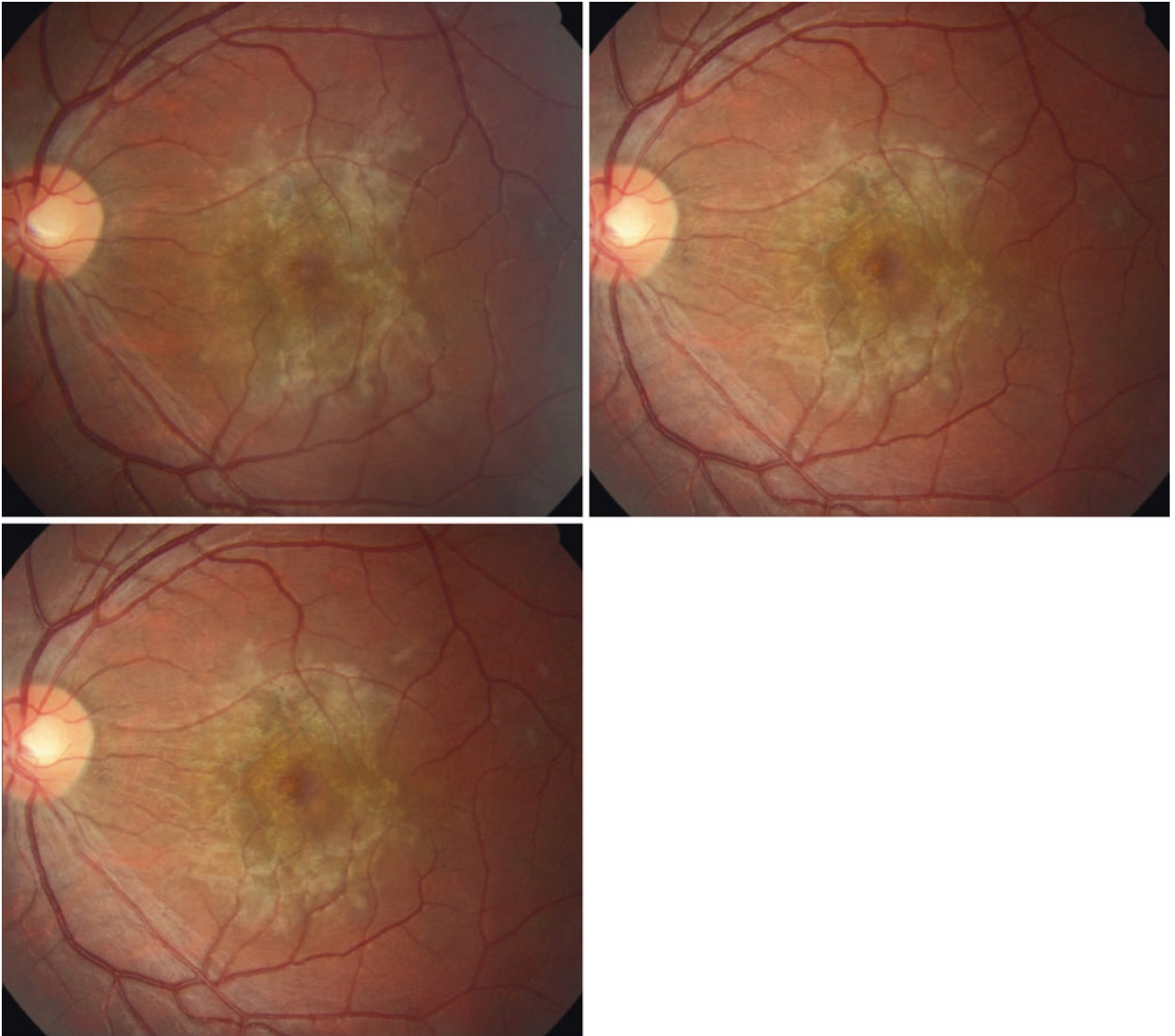


Fig. 3.49 Macular atrophy
I. Fovea

II. Irregular diameter of retinal veins
III. RPE atrophy

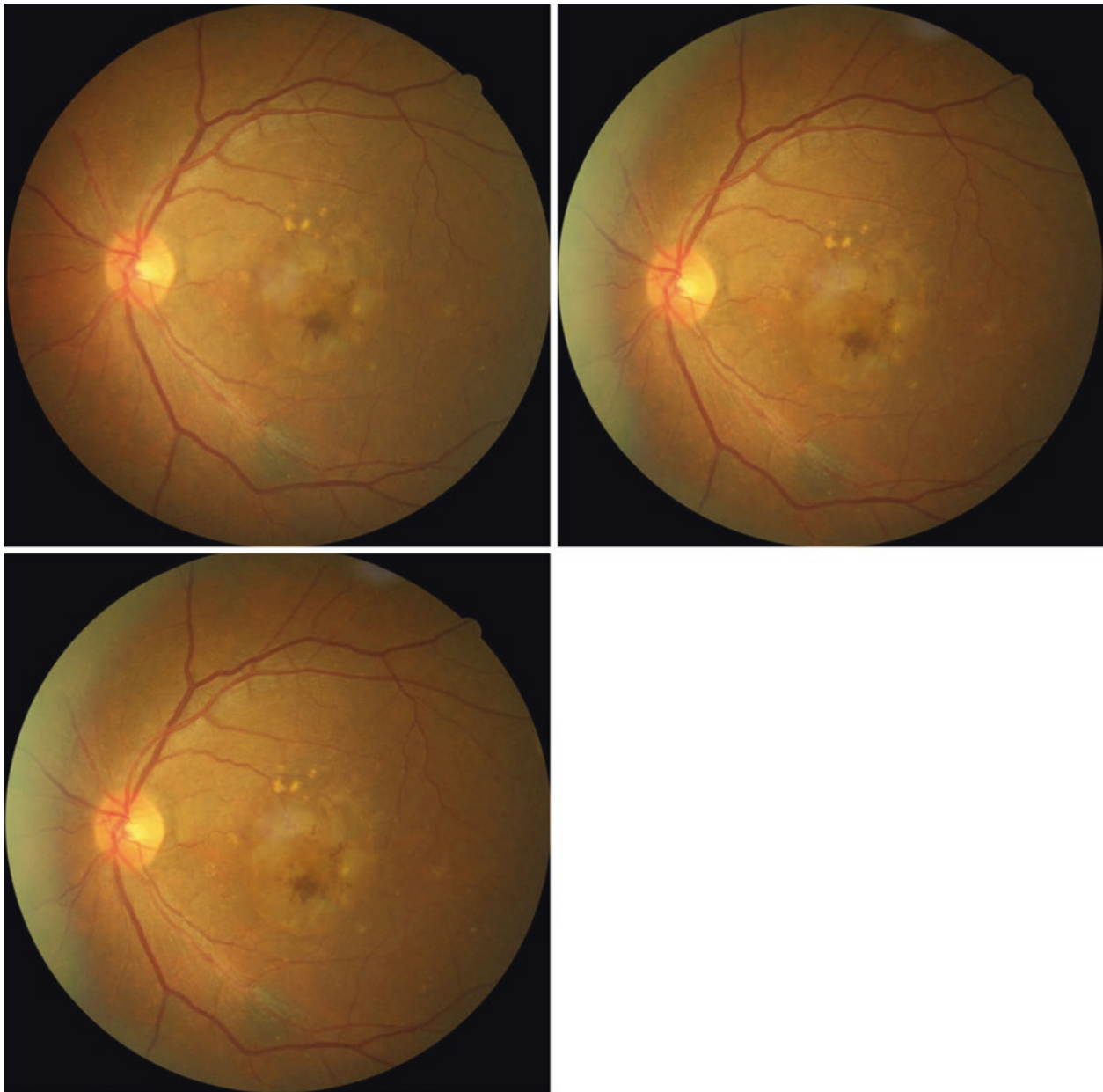


Fig. 3.50 Macular atrophy after branch retinal artery occlusion
I. Macular atrophy and thinning

II. Thinning of inferior temporal retinal artery
III. Pigment proliferation

References

1. Tyler ME. Stereo fundus photography: principles and techniques. In: Saine PJ, Tyler ME, editors. *Ophthalmic photography: retinal photography, angiography, and electronic imaging*. 2nd ed. Boston: Butterworth-Heinemann; 2002. p. 118–35.
2. Rudnisky CJ, Tennant MT, de Leon AR. Benefits of stereopsis when identifying clinically significant macular edema via teleophthalmology. *Can J Ophthalmol*. 2006;41:727–32.
3. Gass JDM. *Stereo atlas of macular diseases: diagnosis and treatment*. 4th ed. St. Louis: Mosby; 1997.
4. Hubbard LD, Danis RP, Neider MW. Brightness, contrast, and color balance of digital versus film retinal images in the age-related eye disease study 2. *Invest Ophthalmol Vis Sci*. 2008;49:3269–82.
5. The Age-Related Eye Disease Study Research Group. The age-related eye disease study system for classifying age-related macular degeneration from stereoscopic color fundus photography: the age-related eye disease study report number 6. *Arch Ophthalmol*. 2001;31:167–75.
6. Rudnisky CJ, Hinz BJ, Tennat MTS, et al. High-resolution stereoscopic digital fundus photography versus contact lens biomicroscopy for the detection of clinically significant macular edema. *Am J Ophthalmol*. 2002;109:267–74.
7. Slakter JS, Yannuzzi LA, Schneider U, et al. retinal choroidal anastomoses and occult choroidal neovascularization in age-related macular degeneration. *Ophthalmology*. 2000;107:742–53.
8. Agarwal A. *Gass atlas of macular diseases*. 5th ed. Edinburgh: Elsevier; 2012. p. 1–16.