Retinal and Choroidal Inflammation

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Retinal and choroidal inflammation can be assessed by various investigations. For example, fundus examination can reveal retinal tortuous vessels, hard exudates, and hemorrhages; FFA can reflect the leakage status of retinal vessels; OCT and OCTA can show deep retinal vascular structures through signal reflection [1].

Retinal and choroidal inflammation usually accompany the vitreous opacities or anterior segment inflammation, which may reduce the quality of OCT images by artifacts or weak signals [2]. Macular edema is shown as lower signal among the retinal layers. Secondary macular epiretinal membrane may be seen on OCT as a layer of hyperreflective signal attached to the retinal surface (Figs. 11.1 and 11.2) [3–4].

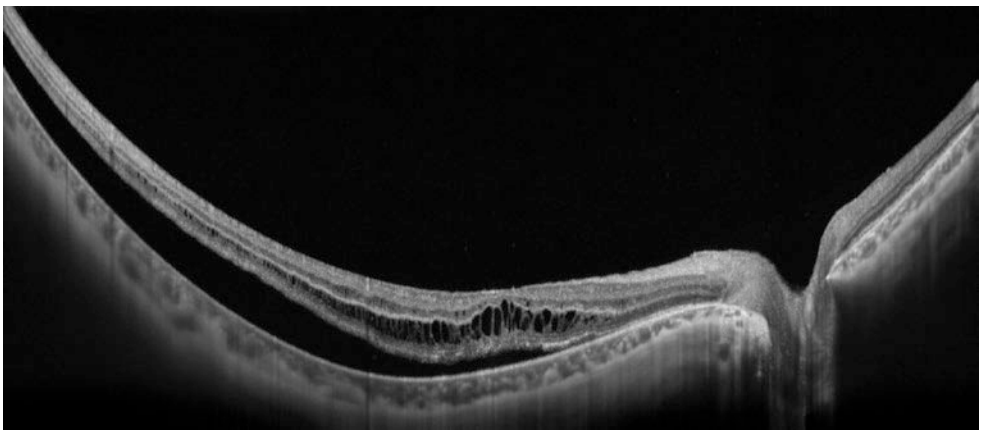

Fig. 11.1 SS-OCT single line B-scan. A 51-year-old male presented with blurred vision and ocular pain in his right eye for 10 months. SS-OCT (12 mm) shows retinal detachment and inner retinal cystic edema

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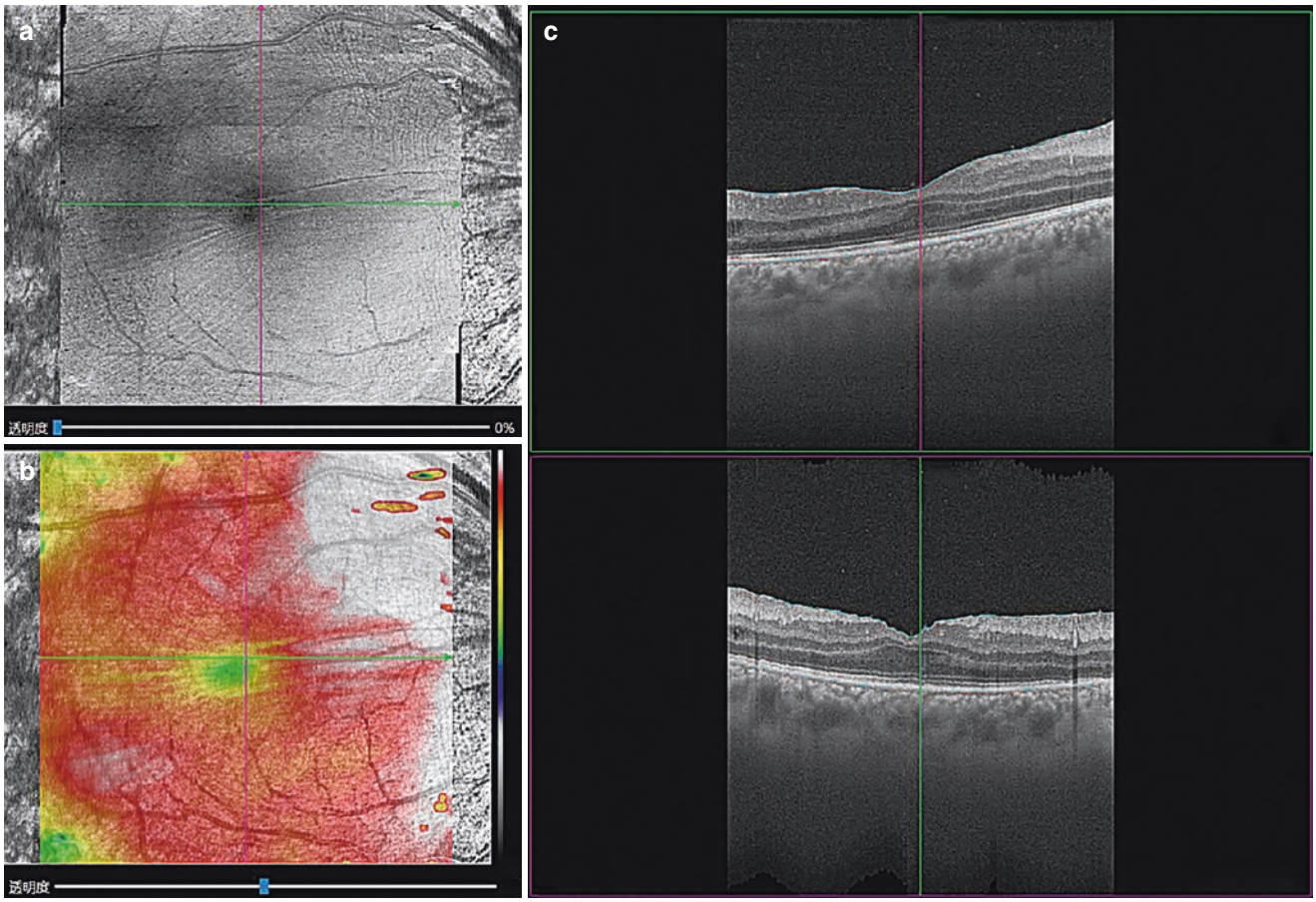

Fig. 11.2 SS-OCT image A 15-year-old female presented with red eye and decreased vision in both eyes for 1 week. She was treated with ocular glucocorticoid therapy. (**a**, **b**) SS-OCT en face image shows retinal folds and retinal thickening in macular; (**c**) B-scan shows small inner retinal cyst

Infectious Uveitis

According to the different pathogens and the stages of the disease, infectious uveitis has various manifestations in OCT. For example, the acute phase of herpetic uveitis (e.g., ARN) may present with severe retinal thickening, inner retinal hyperreflectivity, outer retinal disruptions, or subretinal fluid. In fungal uveitis, hyperreflective signals in the retina could be found under the RPE, which gradually protrude into the outer retina in OCT. In human immunodeficiency virus (HIV) infection, microvascular lesions such as cotton wool spots and hemorrhages are likely to occur due to the damage of small arteries, and hyperreflective lesions could be seen on OCT. When combined with opportunistic infection, cytomegalovirus infections can be seen as localized hyperreflective retinal masses on OCT (Figs. 11.3, 11.4, 11.5 and 11.6) [5].

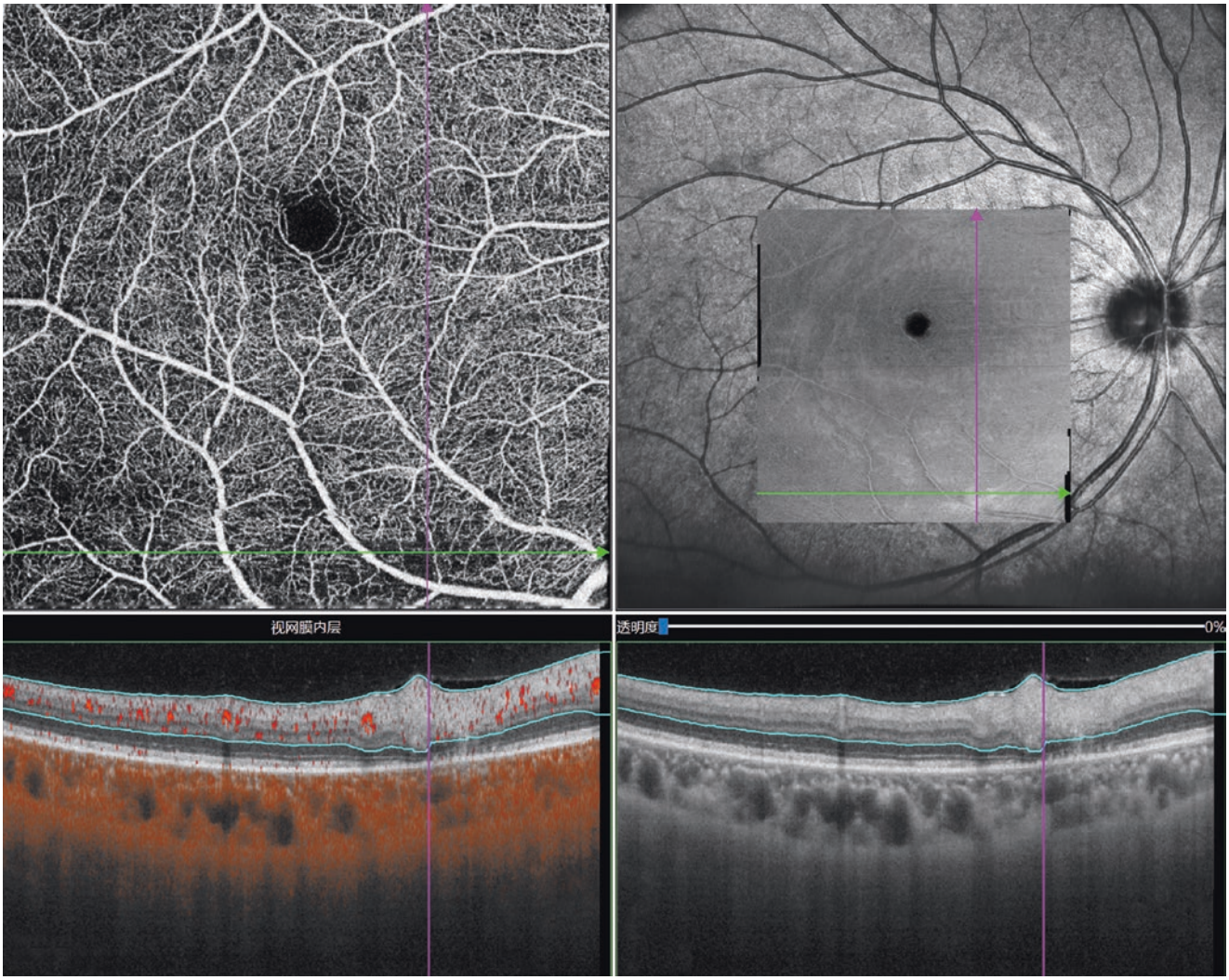

Fig. 11.3 SS-OCTA image. A 28-year-old male had blurred vision in his right eye for 2 weeks. He had previously been diagnosed with HIV infection. OCT shows cotton wool spot in the inferior temporal retina with a superficial retinal hyperreflective lesion; OCTA (6 mm × 6 mm) shows a slightly reduced capillary blood flow density in the inner retinal layer, and the arch ring was still intact

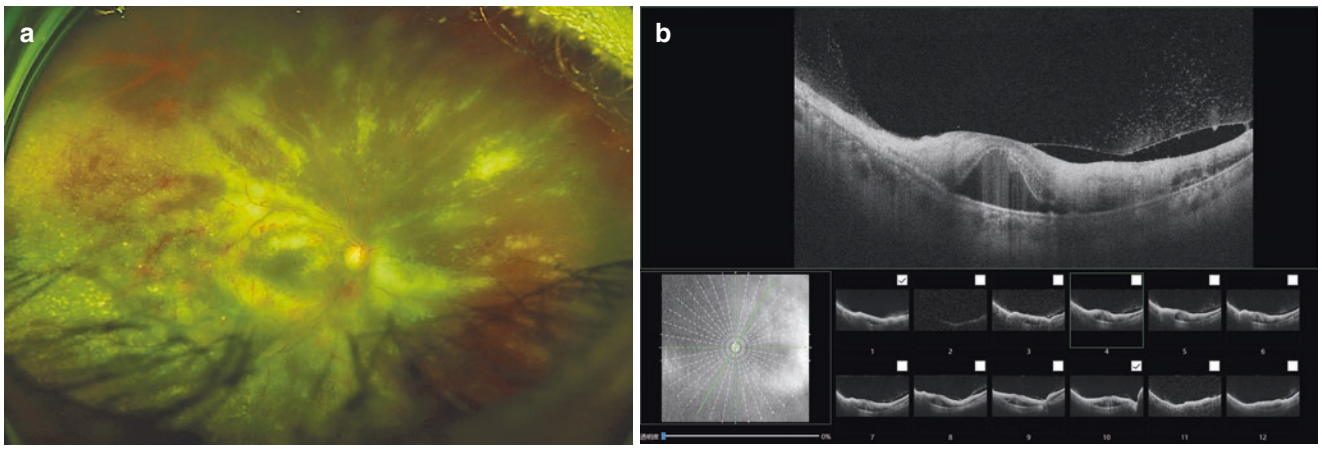

Fig. 11.4 Ultra-wide-field fundus photography and SS-OCTA image 24-year-old male, HIV-infected. (**a**) ultra-wide-field fundus photograph shows large yellow-white exudate lesion near the vascular arch, with scattered large exudates and hemorrhages in the peripheral retina; (**b**) SS-OCT (9 mm × 9 mm) shows vitreous opacity with partial posterior vitreous detachment, macular thickening, inner retina structural disorganization with homogenized moderate reflection; subretinal fluid in fovea. (Youan Hospital of Capital Medical University provided this medical record)

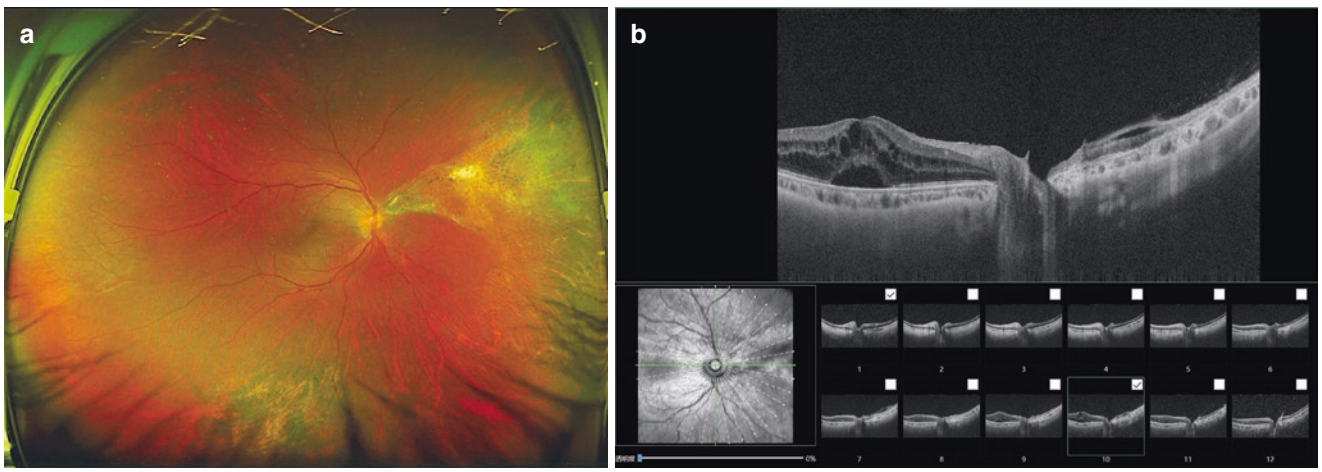

Fig. 11.5 Ultra-wide-field fundus photography and wide-field SS-OCT. A 29-year-old male, with cytomegalovirus (CMV) infection. (**a**) ultra-wide-field fundus photograph shows yellow-white retinal necrotic lesion nasal to the optic disc with surrounding hemorrhagic spots, resembling "tomato cheese"; (**b**) SS-OCT (9 mm) shows retinal thickening and multiple cystic edema with subretinal fluid in the macula; retinal thinning and structural disorganization nasal to the optic disc. (Youan Hospital of Capital Medical University provided this image)

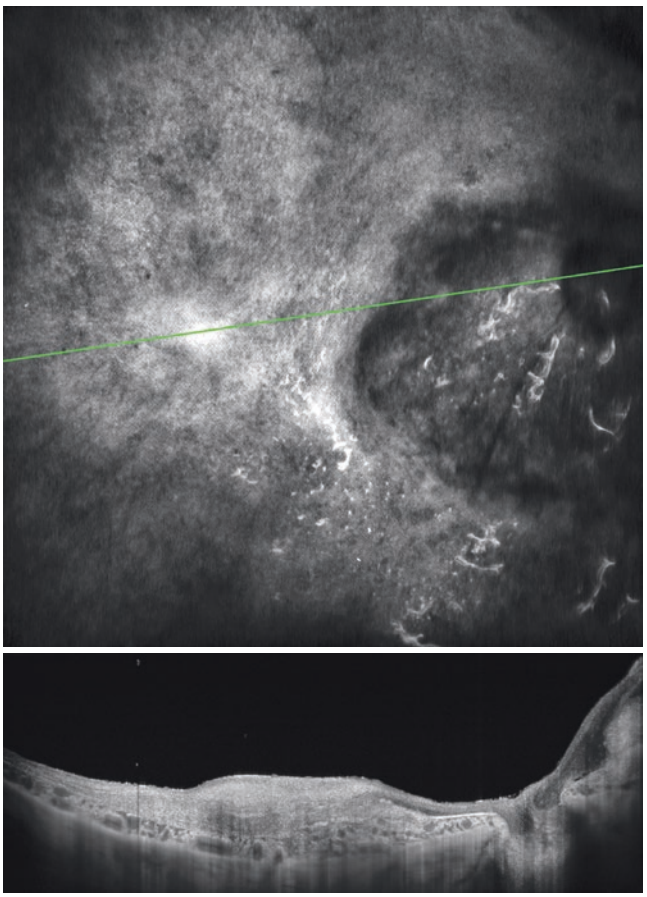

Fig. 11.6 Wide-field SS-OCT single line scan A 31-year-old female had decreased vision in her right eye for 1 month. SS-OCT shows retinal thickening in the temporal retina, superficial retinal hyperreflective masses, and retinal disorganization. These were the lesions of *Toxoplasma gondii* infection

Retinal Vasculitis

Retinal vasculitis is a large spectrum of inflammatory diseases characterized by inflammation of the retinal vessels, especially retinal veins. It usually manifests as retinal vasculitis or perivasculitis. Any retinal vein from the optic disc to the peripheral retina can be involved, either segmentally or throughout. If the inflammation persists, secondary changes such as vascular hyaline degeneration, luminal stenosis or occlusion, thrombosis, necrosis, or even rupture of the blood vessel wall will occur, which can lead to retinal hemorrhage, edema, exudation, telangiectasia, microaneurysm, retinal neovascularization, and even retinal detachment. According to the origin of inflammation, the retinal vasculitis can be broadly classified into: (1) infectious or secondary to uveitis; (2) systemic inflammatory disorders, such as systemic lupus erythematosus; and (3) unknown causes, such as Eales disease. The major investigation for retinal vasculitis is FFA, which is usually characterized by vascular wall staining and leakage. OCT usually reveals retinal edema and structural disorganization, while OCTA can clearly show retinal vascular density changes and nonperfusion areas (Figs. 11.7, 11.8, 11.9, 11.10, and 11.11).

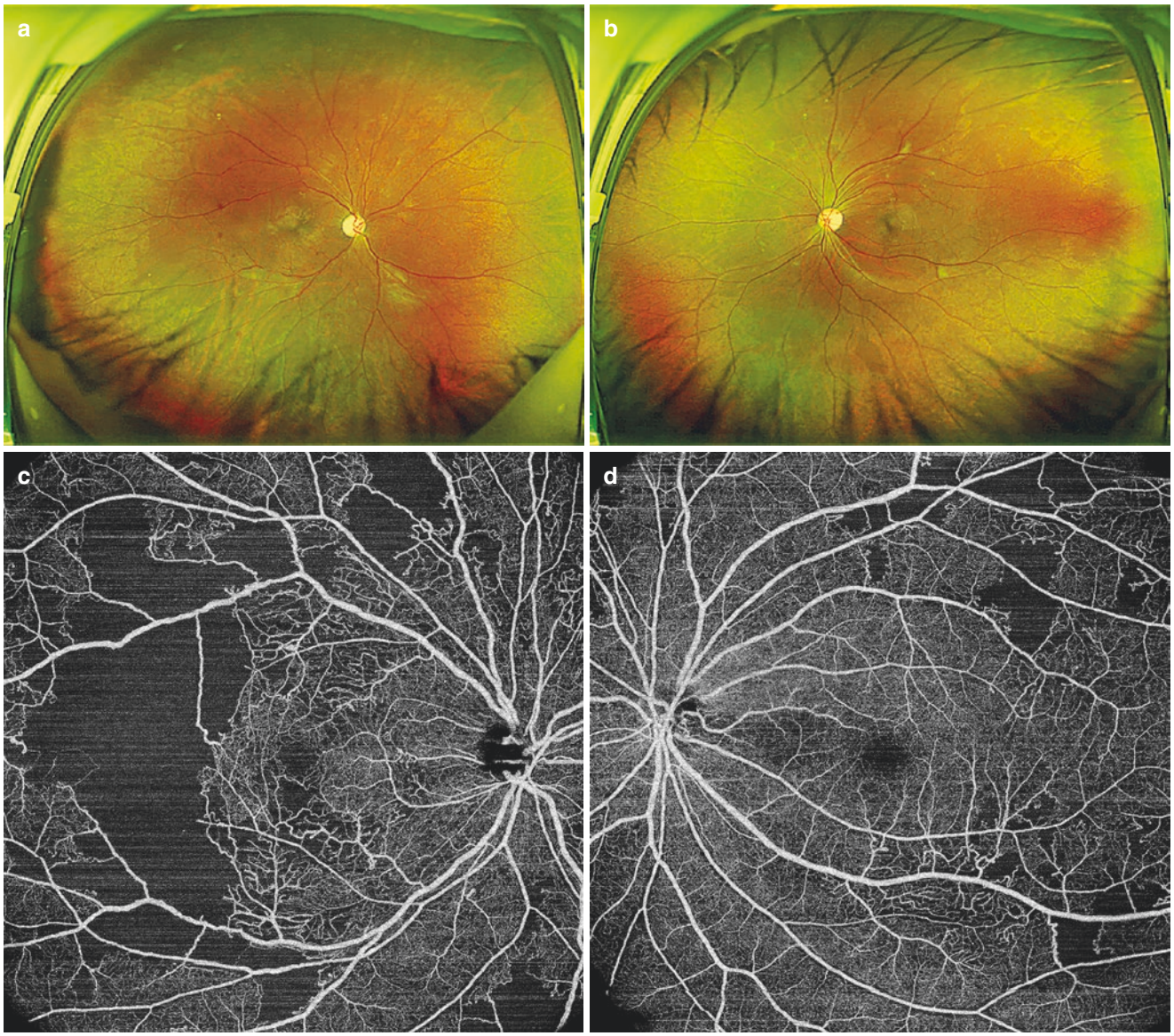

Fig. 11.7 Ultra-wide-field fundus photography and wide-field SS-OCTA images A 20-year-old female was diagnosed with systemic lupus erythematosus. **a** (right eye), **b** (left eye): ultra-wide-field fundus photography shows scattered hemorrhages and cotton-wool spots in both eyes, with grayish white appearance of peripheral retina; **c** (right eye), **d** (left eye): OCTA (12 mm × 12 mm, retina layer) shows nonperfusion areas in the posterior pole of both eyes, with distorted and dilated retinal vessels; **e** (right eye), **f** (left eye): OCTA montage image (retina layer) presented with larger area of the fundus, the nasal nonperfusion area of the left eye is also noted

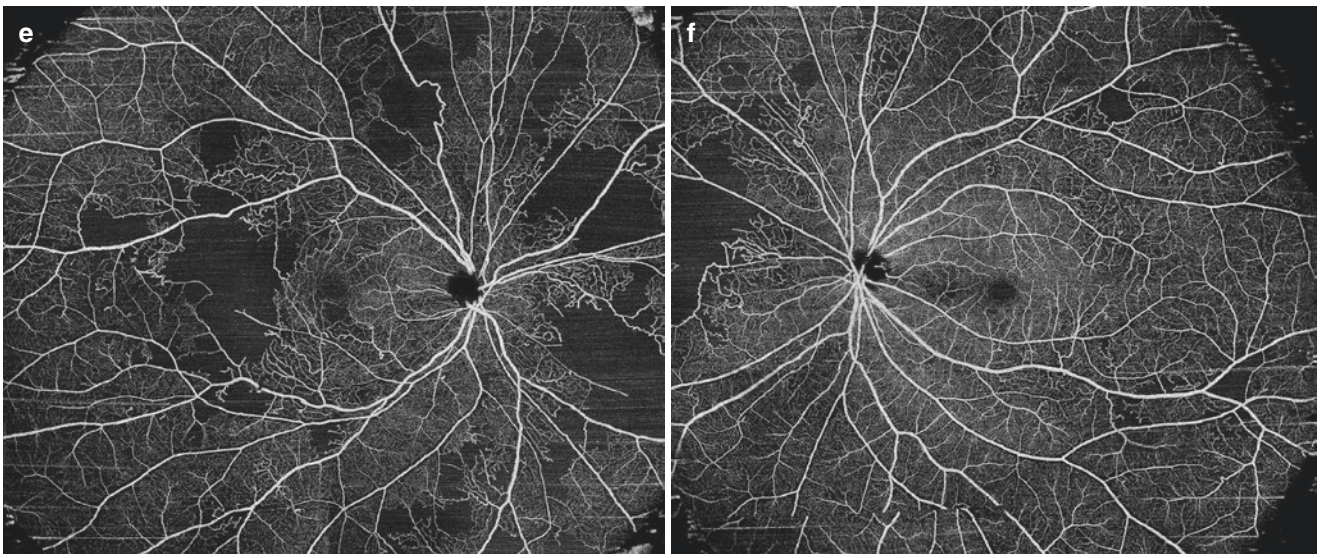

Fig. 11.7 (continued)

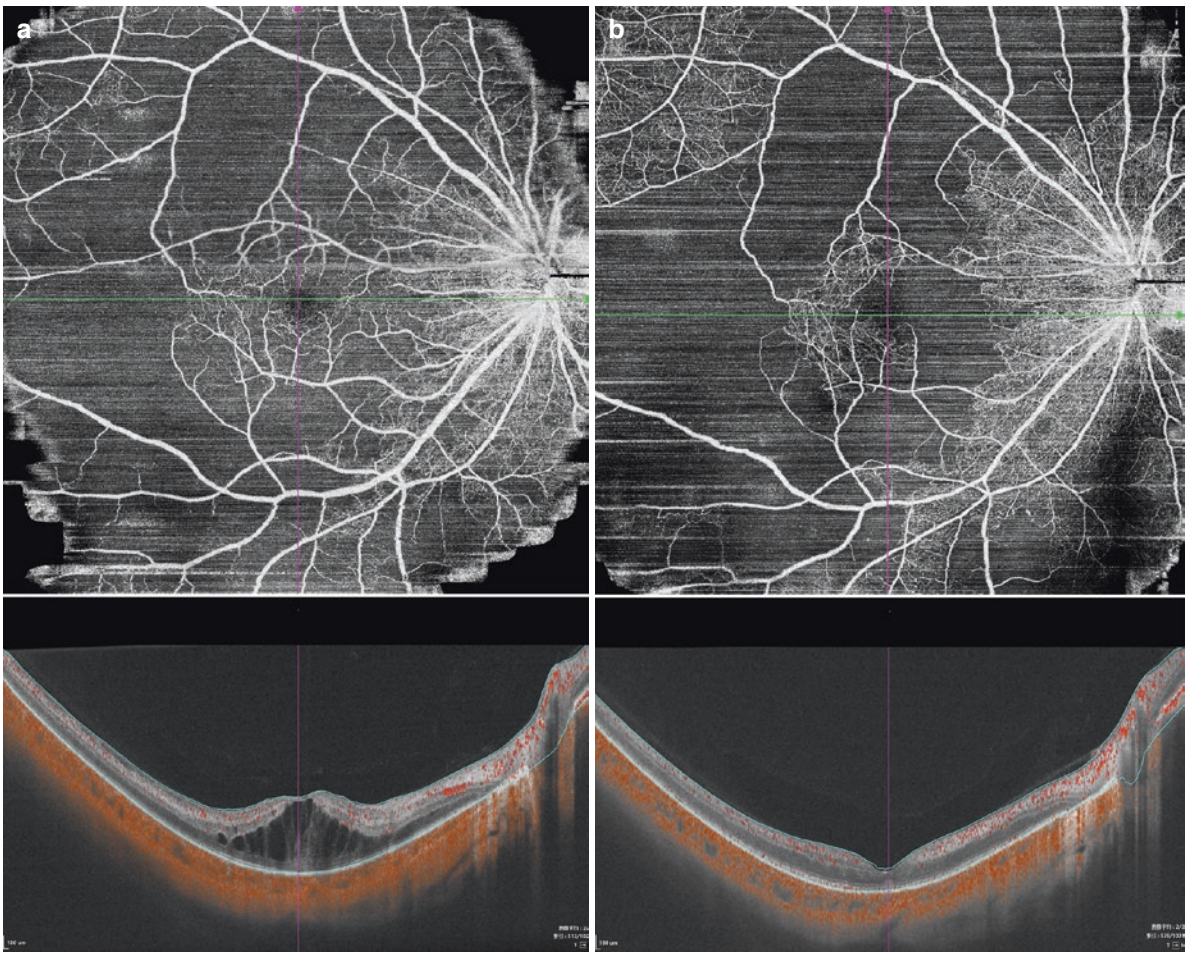

Fig. 11.8 Wide-field SS-OCTA image. A 26-year-old female with systemic lupus erythematosus retinopathy on the right eye. (**a**) OCTA (12 mm x 12 mm, retina layer) shows extensive nonperfusion area throughout the posterior pole, macular cystoid edema (CME) is also noted in the B-scan image. (**b**) OCTA (12 mm × 12 mm, retina layer) of the right eye after 1.5 months of active treatment showed that the retinal perfusion was restored, but the nonperfusion area was still extensive. Corresponding B-scan images showed that CME had vanished while the foveal ellipsoid zone had disappeared

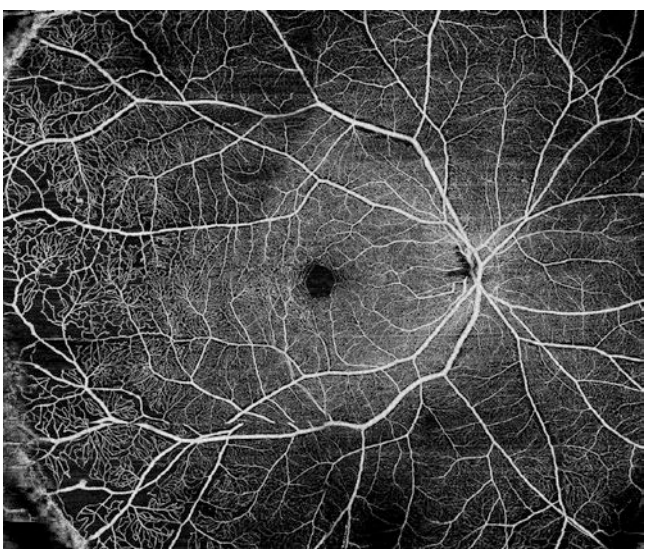

Fig. 11.9 Wide-field SS-OCTA image. A 33-year-old female was diagnosed with retinal vasculitis in the right eye. The visual acuity was 0.8. OCTA montage (inner retina) showed dilation and dense decrease of the retinal capillaries in the right eye.

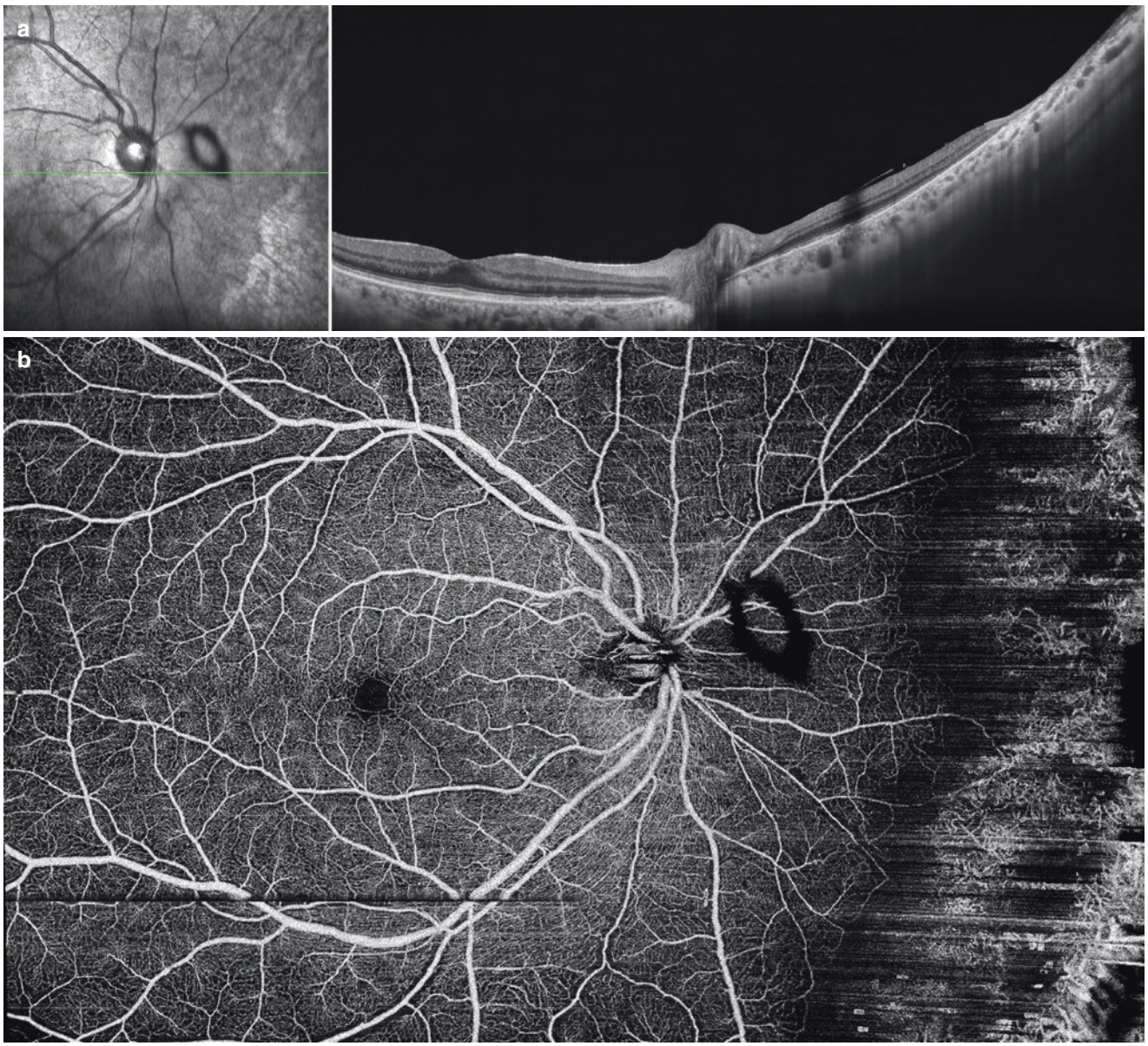

Fig. 11.10 Wide-field SS-OCT and ultra-wide-field SS-OCTA image. A 30-year-old female was diagnosed with retinal vasculitis in the right eye. (**a**) SS-OCT HD single line completely shows the width from the macula to the nasal extent of the optic disc with the depth to the choroidal-scleral junction. Atrophy and thinning of the retinal retina in the nasal mid-periphery region and the loss of RPE could be seen; (**b**) OCTA montage (retina layer) is difficult to delaminate the nasal retina due to the full-layer atrophy, with perspective of the choroidal vessels below. Weiss ring projection was visible near the optic disc

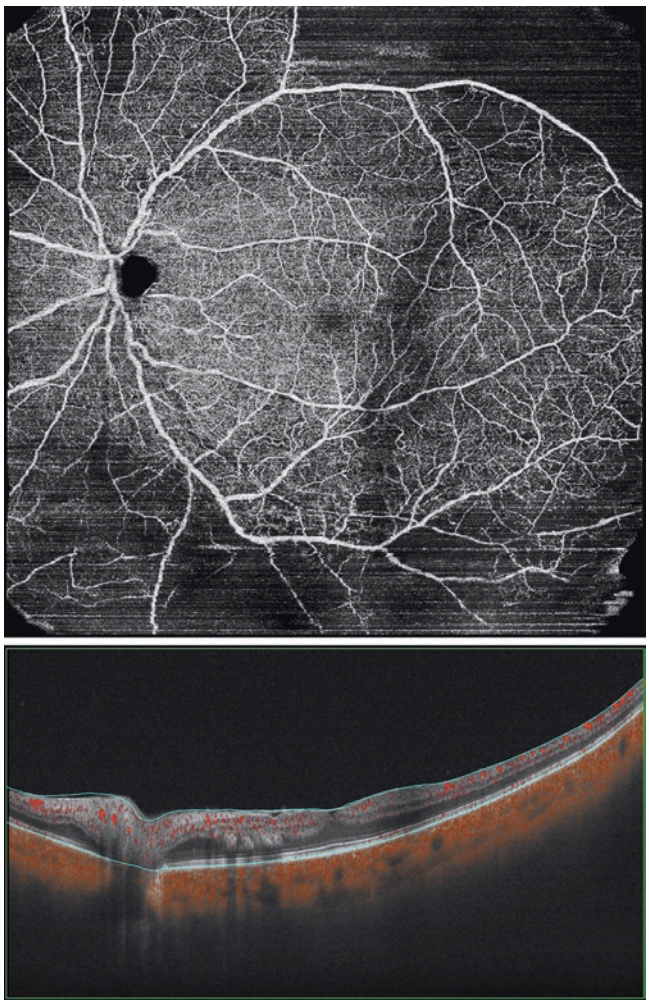

Fig. 11.11 Wide-field SS-OCTA image. A 55-year-old male diagnosed with retinal vasculitis in the left eye. OCTA (12 mm × 12 mm, retina layer) shows the loss of foveal avascular zone, uneven retinal vein diameter, and extensive nonperfusion areas. The corresponding B-scan shows mild macular edema and flattening of the fovea

Vogt-Koyanagi-Harada Disease

Vogt-Koyanagi-Harada disease (VKH) is a common cause of noninfectious panuveitis, which is usually present with bilateral panuveitis. The acute phase is characterized by a panuveitis or posterior uveitis with multifocal serous retinal detachments. In the chronic phases, Dalen-Fuchs nodules and sunset glow fundus are often noted. OCT features include multiple serous retinal detachments, choroidal folds, and choroidal thickening (Figs. 11.12 and 11.13) [6].

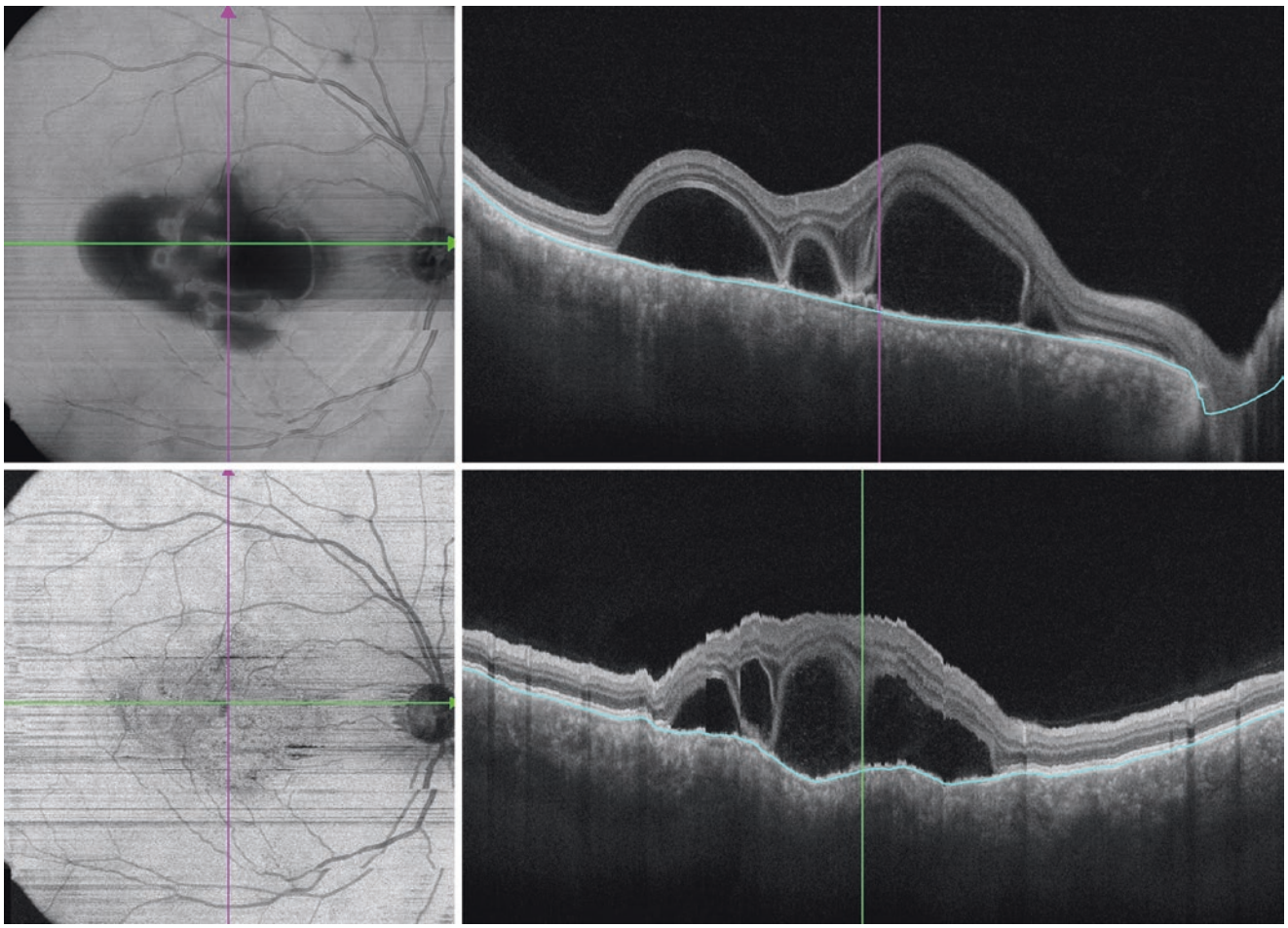

Fig. 11.12 Wide-field SS-OCT image. A 34-year-old female was diagnosed as VKH. For her right eye, SS-OCT (12 mm × 12 mm) shows multifocal serous retinal detachment with RPE folds and choroidal thickening

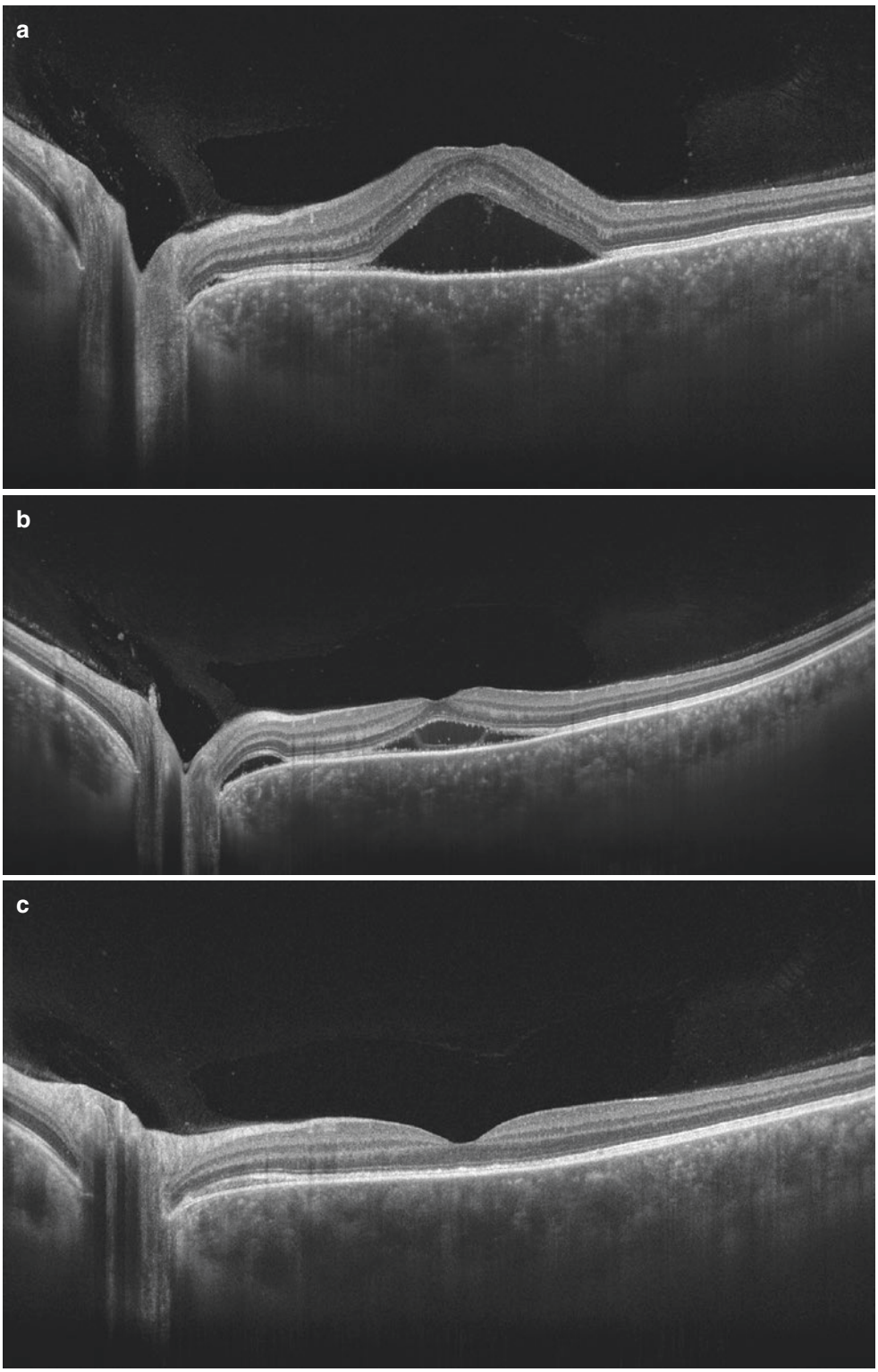

Fig. 11.13 Wide-field SS-OCT single-line scan. For the same patient in Fig. 11.12, (**a**) SS-OCT (12 mm × 12 mm) of her left eye reveals serous retinal detachment with punctate hyperreflexia under the retina; (**b**) Subretinal fluid decreased after 4 days of treatment with glucocorticoid eye drops and peribulbar injection of triamcinolone acetonide; (**c**) Subretinal fluid was completely absorbed after 2 weeks of glucocorticoid treatment

Behçet's Disease

Behçet's disease is a multi-organ chronic autoimmune disease characterized by uveitis, oral ulcers, and skin lesions. Ocular manifestation includes anterior uveitis with hypopyon, retinal vasculitis, and retinitis. The fundus is often characterized by retinal hemorrhage, edema, and yellowish-white exudate. In the acute phase of Behçet's disease, RPE atrophy can be observed on OCT. OCTA shows irregular FAZ, disorganized macular capillary arch rings, etc. (Figs. 11.14 and 11.15).

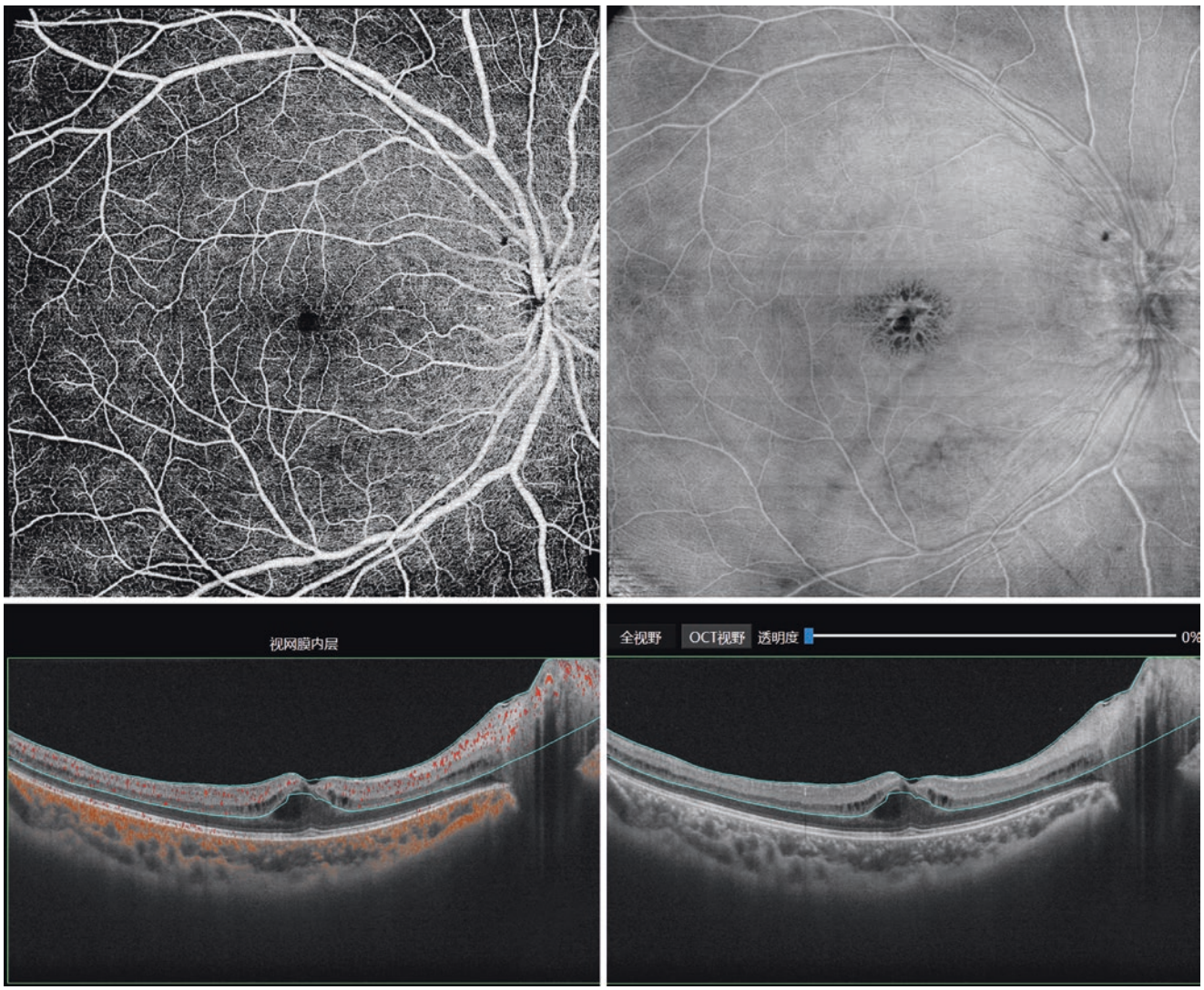

Fig. 11.14 Wide-field SS-OCTA image. A 20-year-old female had sudden vision loss and visual distortion in both eyes for 1 week. SS-OCTA (12 mm x 12 mm) shows irregular FAZ in the inner retina, retinal thickening, macular cystic edema, and choroidal thickening in her right eye

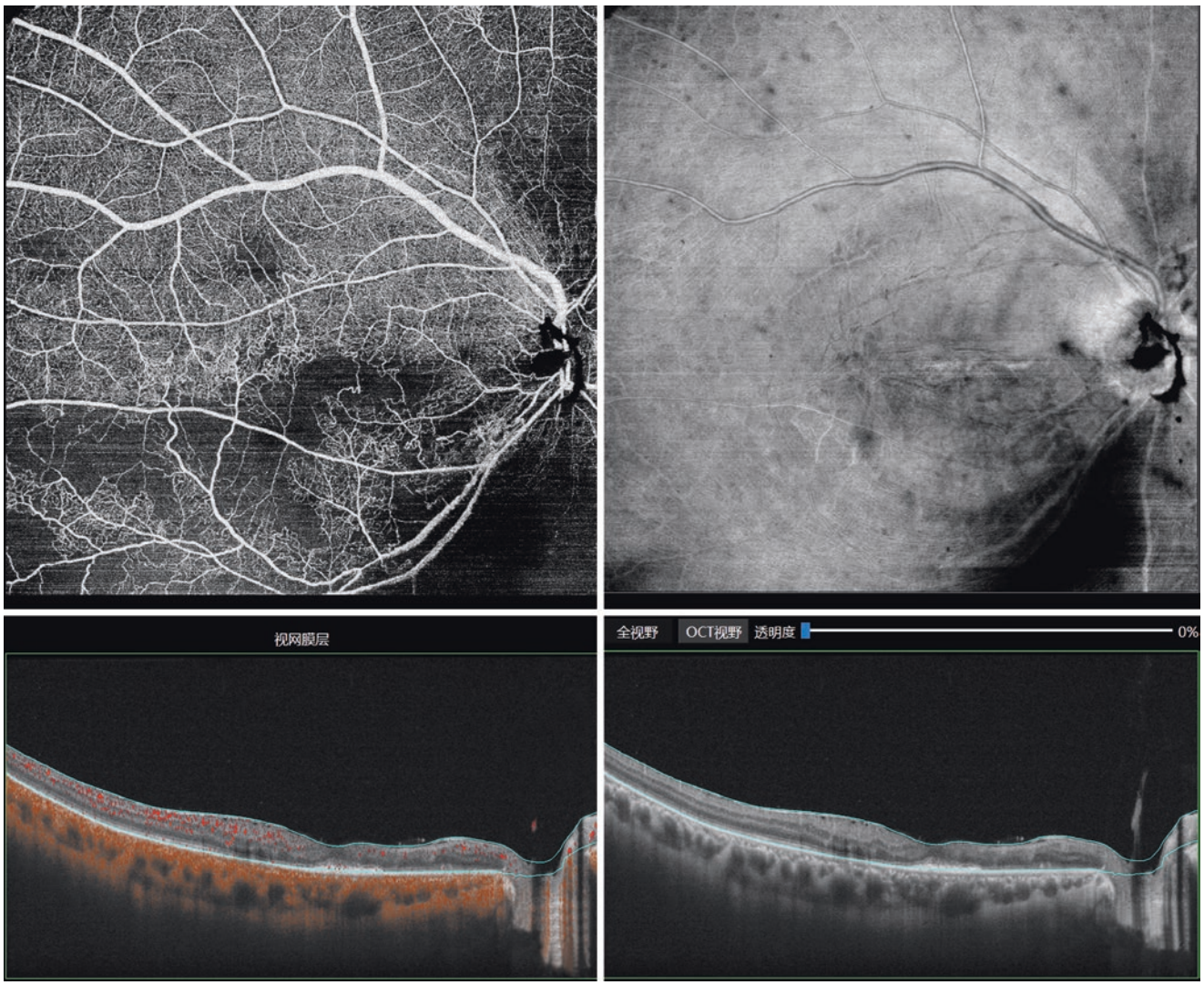

Fig. 11.15 Wide-field SS-OCTA image. For the same patient in Fig. 11.14, SS-OCTA (12 mm x 12 mm) shows sparse blood vessels in the inner layer, with disruption of the macular arch, and irregular FAZ in her right eye. No blood flow signal is shown in the lower part of the retina. B-scan shows retinal atrophy in the right eye, and the retinal structure is disorganized in the inner layer of the central fovea, with discontinuous ellipsoidal bands

Punctate Inner Choroidopathy (PIC)

Punctate inner choroidopathy (PIC) is a rare inflammatory disease of the eye that often affects young, myopic women. Patients present with decreased visual acuity, flashing, and scotoma. Funduscopic examination reveals multiple yellowish-white punctate lesions without intraocular inflammation. PIC is usually a benign disease, and most patients can be observed and kept followed-up. However, PIC is frequently associated with choroidal neovascularization (CNV) and subretinal fibrosis, which may lead to severe visual impairment. Inflammatory lesions without CNV, which are located near the fovea, can be treated with systemic or peri/intraocular steroids or immunomodulators and immunosuppressants. If secondary CNV develops, intravitreal injection of anti-VEGF agents can be used (Figs. 11.16, 11.17, 11.18, 11.19, 11.20, 11.21, 11.22 and 11.23) [7–9]

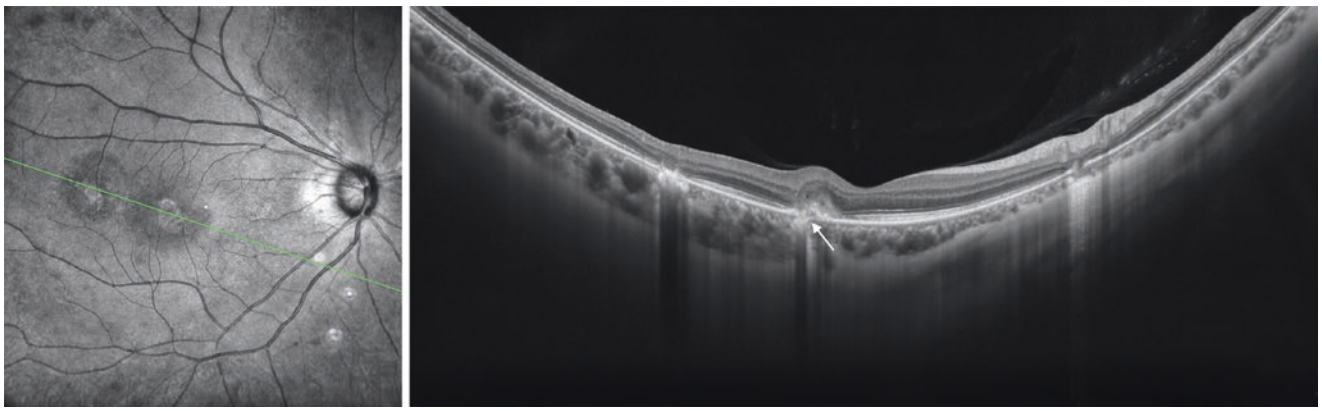

Fig. 11.16 SS-OCT single line scan. SS-OCT (12 mm single line scan) shows two active chorioretinal lesions with loss of ellipsoid zone, interdigitation zone, and external limiting membrane. The subfoveal lesion shows hyperreflective signal in the outer retinal layer with interruption of RPE (white arrow)

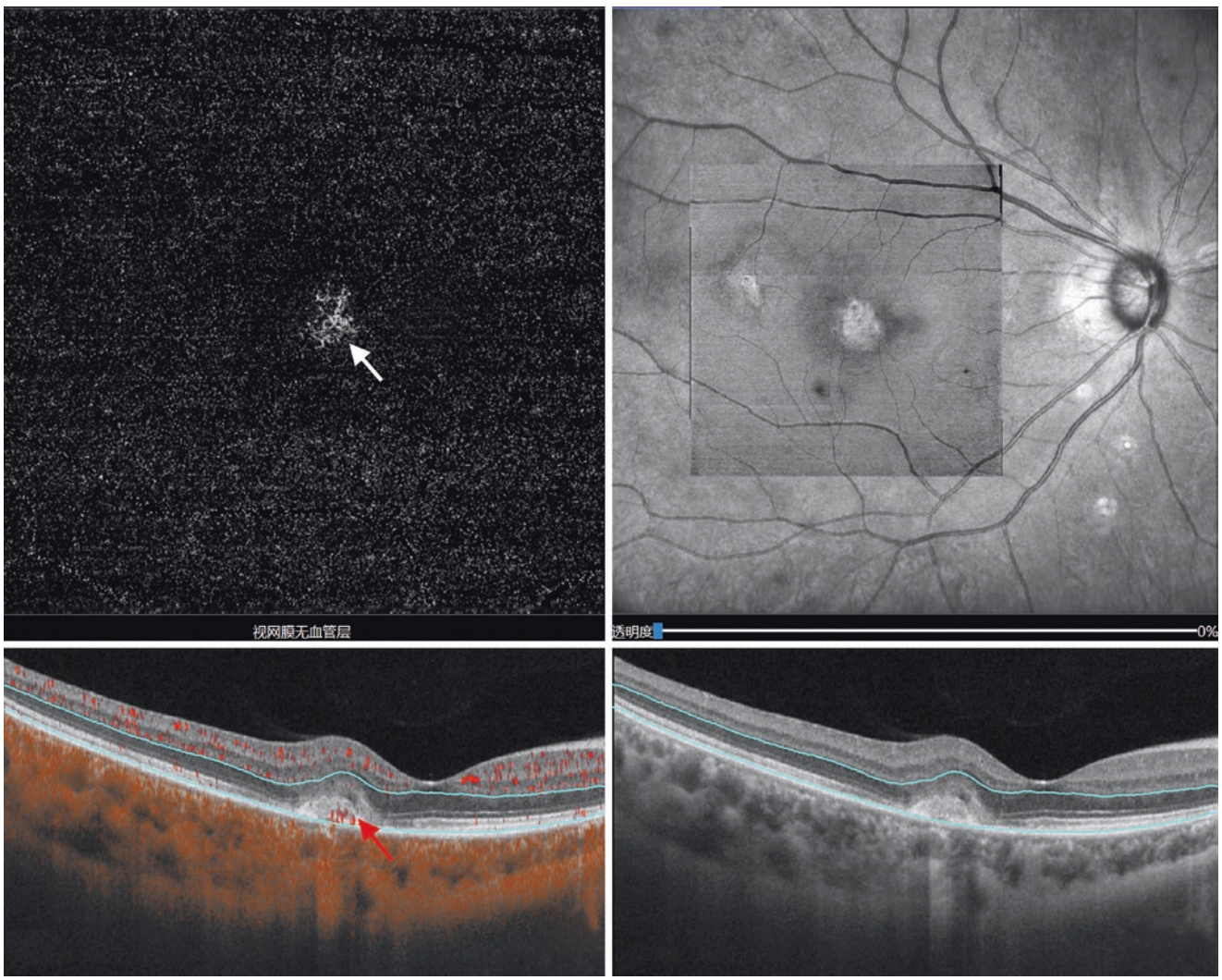

Fig. 11.17 SS-OCTA images. 6 x 6 mm OCTA of the same patient in Fig. 11.16 was helpful to identify the presence of secondary choroidal neovascularization. A secondary CNV can be seen in the avascular layer (white arrow). In the B-scan image, an area of hyperreflectivity with blood flow signals above RPE layer can be seen corresponding to the CNV (red arrow)

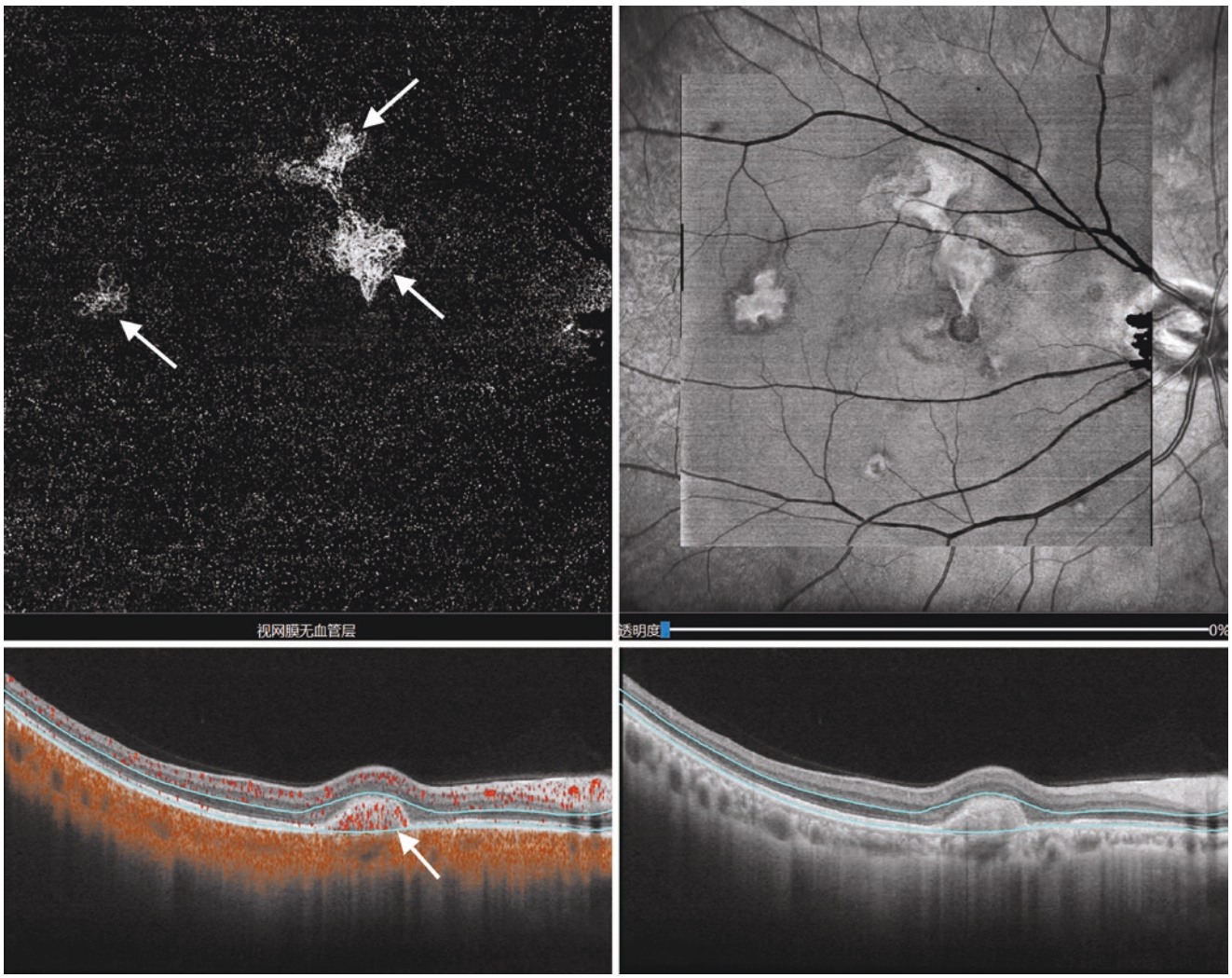

Fig. 11.18 SS-OCTA images of the macula. A 27-year-old male presented with vision loss and distortion in the right eye for 1 year. Color fundus photograph shows multiple grayish white lesions in the posterior pole of right eye. OCTA (9 × 9 mm retinal avascular layer) shows multiple gray-white lesions secondary to CNV (white arrows), and the corresponding B-scan shows red blood flow signals located above the RPE

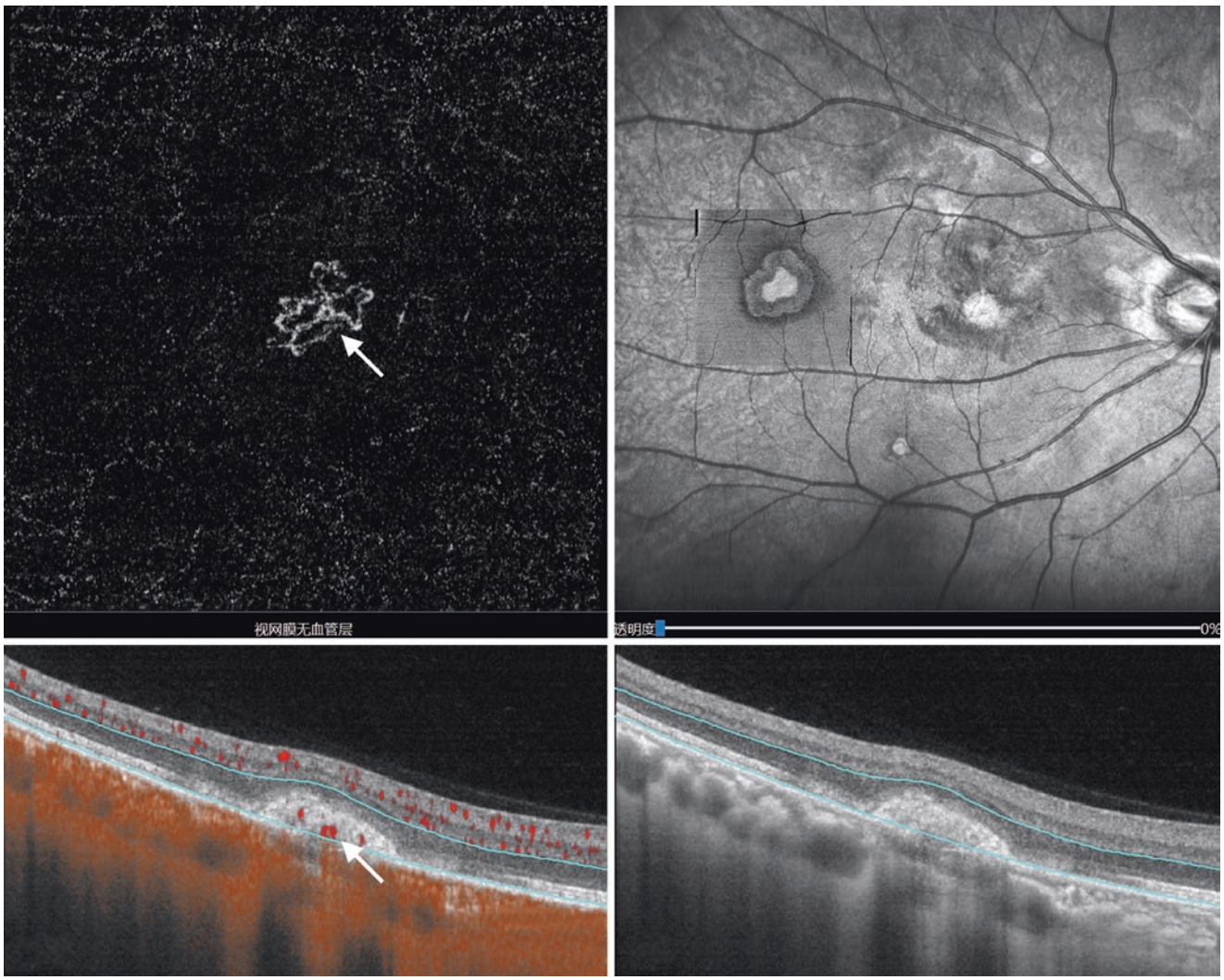

Fig. 11.19 SS-OCTA image in the temporal side of the macula. OCTA (3 mm × 3 mm retinal avascular layer) of the same patient in Fig. 11.18, shows more detailed vasculature of the CNV located to the temporal of foveal (white arrow), and the corresponding B-scan shows red blood flow signals above the RPE layer

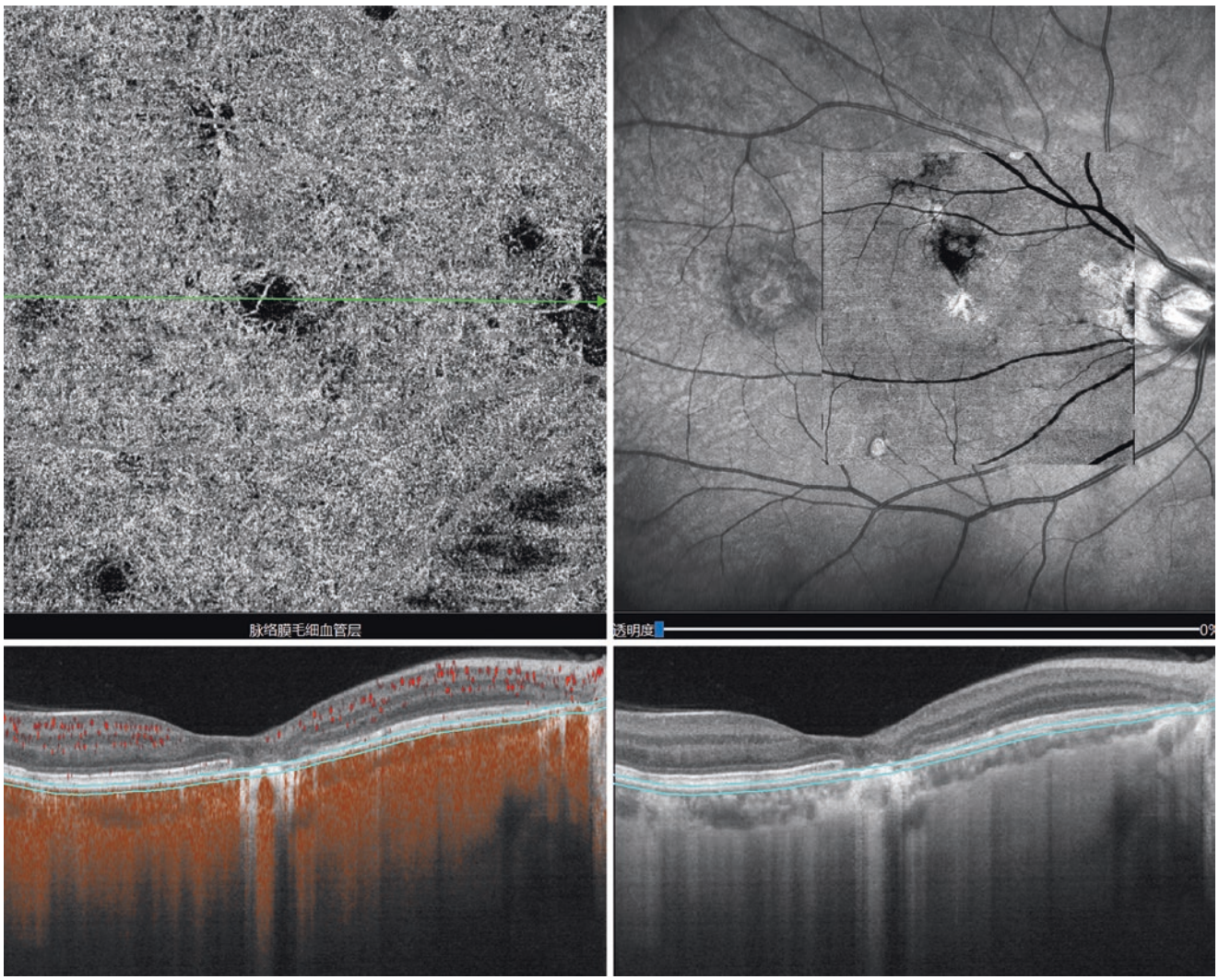

Fig. 11.20 SS-OCTA of the macula. OCTA (6 x 6 mm choroidal capillary layer) of the same patient in Fig. 11.18 shows foveal atrophy with loss of outer retina and choroid

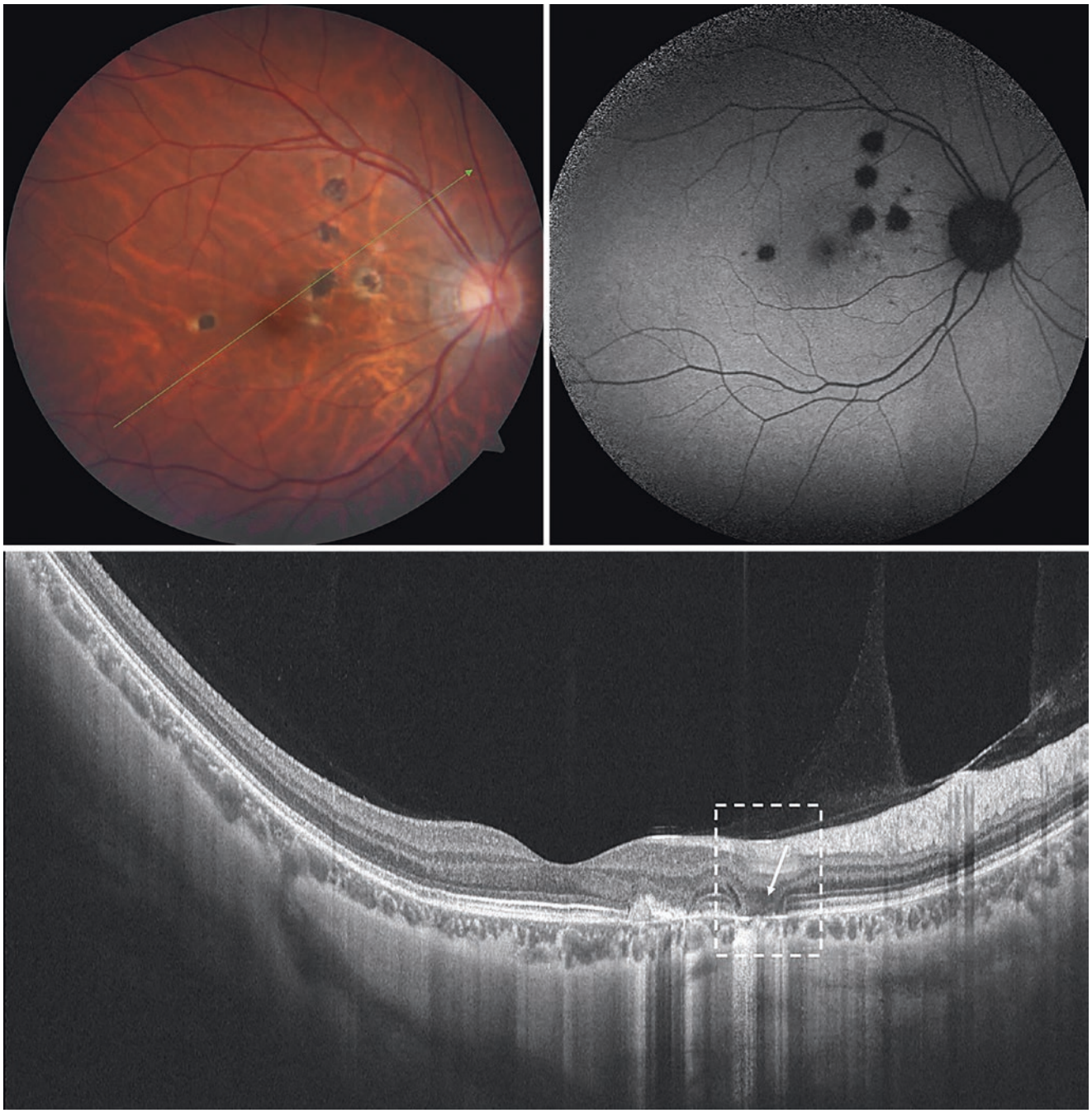

Fig. 11.21 Color Fundus Photograph, Autofluorescence, and SS-OCT images. A 37-year-old male presented with vision loss in both eyes for 3 years and visual distortion in the left eye for 1 month. Color fundus photograph of the right eye shows multiple pigmented lesions in the posterior pole. Fundus autofluorescence shows low autofluorescence at the corresponding lesions. SS-OCT of the posterior pole showing interrupted RPE (white arrow), with herniation of the outer plexiform layer and inner retina to the choroid (V-shaped appearance) through the break of RPE and BM (dashed box)

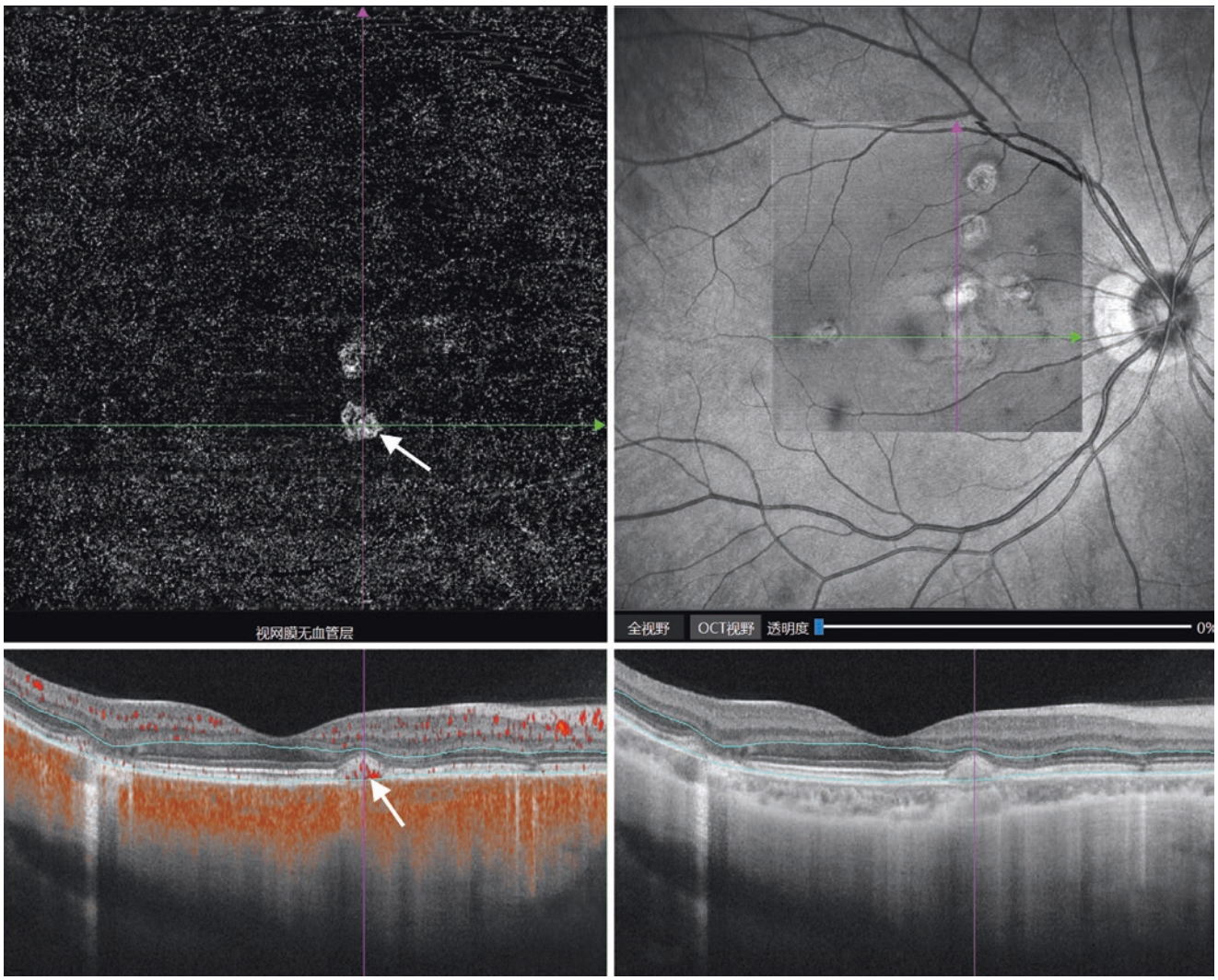

Fig. 11.22 Wide-angle SS-OCTA image. OCTA (9 mm x 9 mm) of the same patient in Fig. 11.21, retinal avascular layer shows a partial lesion secondary to CNV (white arrow), and the corresponding B-scan shows red blood flow signals located above RPE

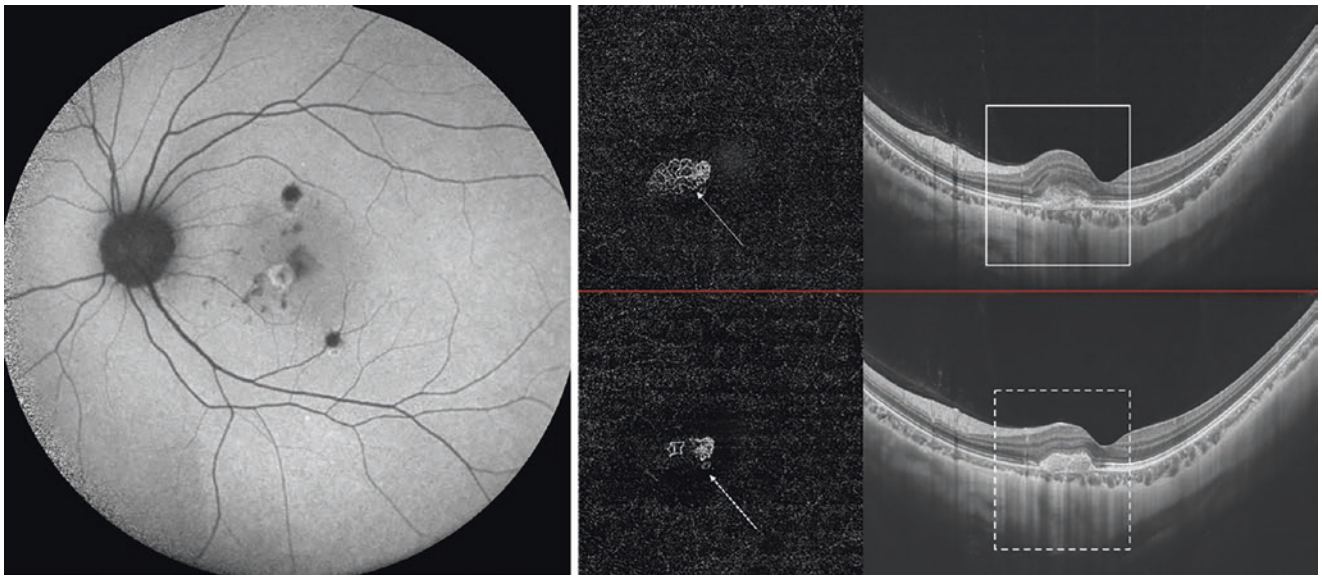

Fig. 11.23 Comparison of SS-OCTA and SS-OCT images before and after anti-VEGF treatment. Multiple patchy low autofluorescence lesions were observed in the left eye, and a round high autofluorescence lesion was found in the center of macula. OCTA (6 mm × 6 mm) of the retinal avascular layer shows a subfoveal secondary CNV (solid arrow), and B-scan OCT shows hyperreflectivity with blurred subretinal border of RPE and external limiting membrane disruption (solid box). One month after anti-VEGF agent injection, OCTA (6 mm × 6 mm) of the avascular layer showed a marked reduction of CNV (dashed arrow), and B-scan OCT showed a well-defined CNV lesion with a visible external limiting membrane and ellipsoid zone (dashed box)

Acute Zonal Occult Outer Retinopathy (AZOOR)

Acute zonal occult outer retinopathy (AZOOR) is a rare idiopathic inflammatory disorder with monocular or binocular onset and unknown etiology [10]. It usually affects young healthy women with myopia who develop acute visual field loss and photopsia due to broad damage of outer retina. 74% of patients have visual acuity of 0.5 or better, and the retina appears normal on funduscopic examination at the early stages of the disease, with regional retinal pigment epithelial atrophy appearing as the disease progresses. 75% of patients have an enlarged blind spot observed in the visual field test, and 99% have abnormal electroretinography. Several treatments for AZOOR have been suggested, including systemic corticosteroids, systemic immunosuppressants, and different antibiotics, but none have been proven to be effective (Figs. 11.24 and 11.25)

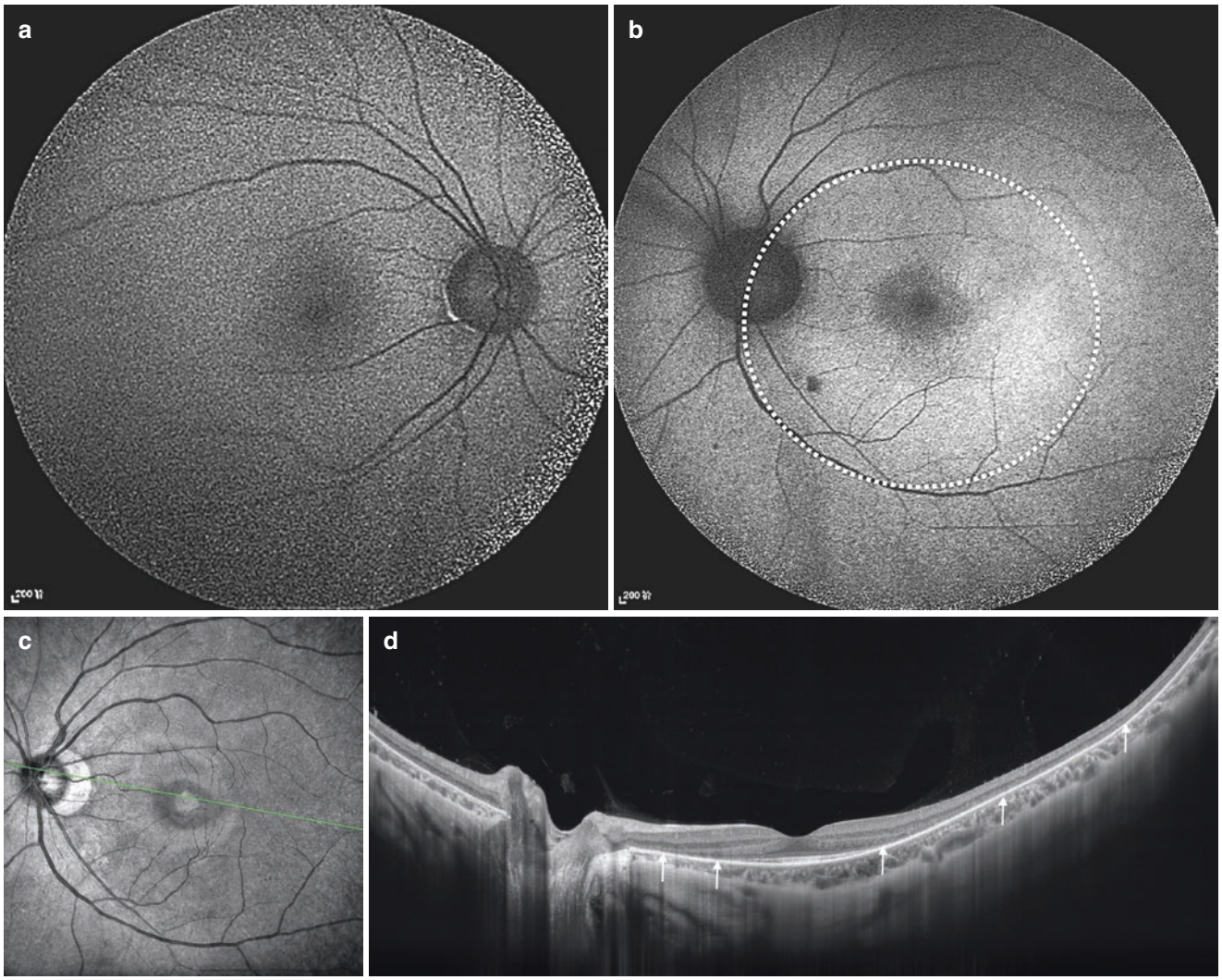

Fig. 11.24 Fundus autofluorescence and wide-angle SS-OCT single-line scan. A 36-year-old female, presented with photopsia in the left eye. Her visual acuity is 20/20 in both eyes. (**a**, **b**) Fundus autofluorescence shows significantly enhanced autofluorescence around the macular region in the left eye compared to the right eye (dotted circle). (**c**, **d**) SS-OCT shows the absence of ellipsoid zone, interdigitation zone, and external limiting membrane of the macular (white arrow)

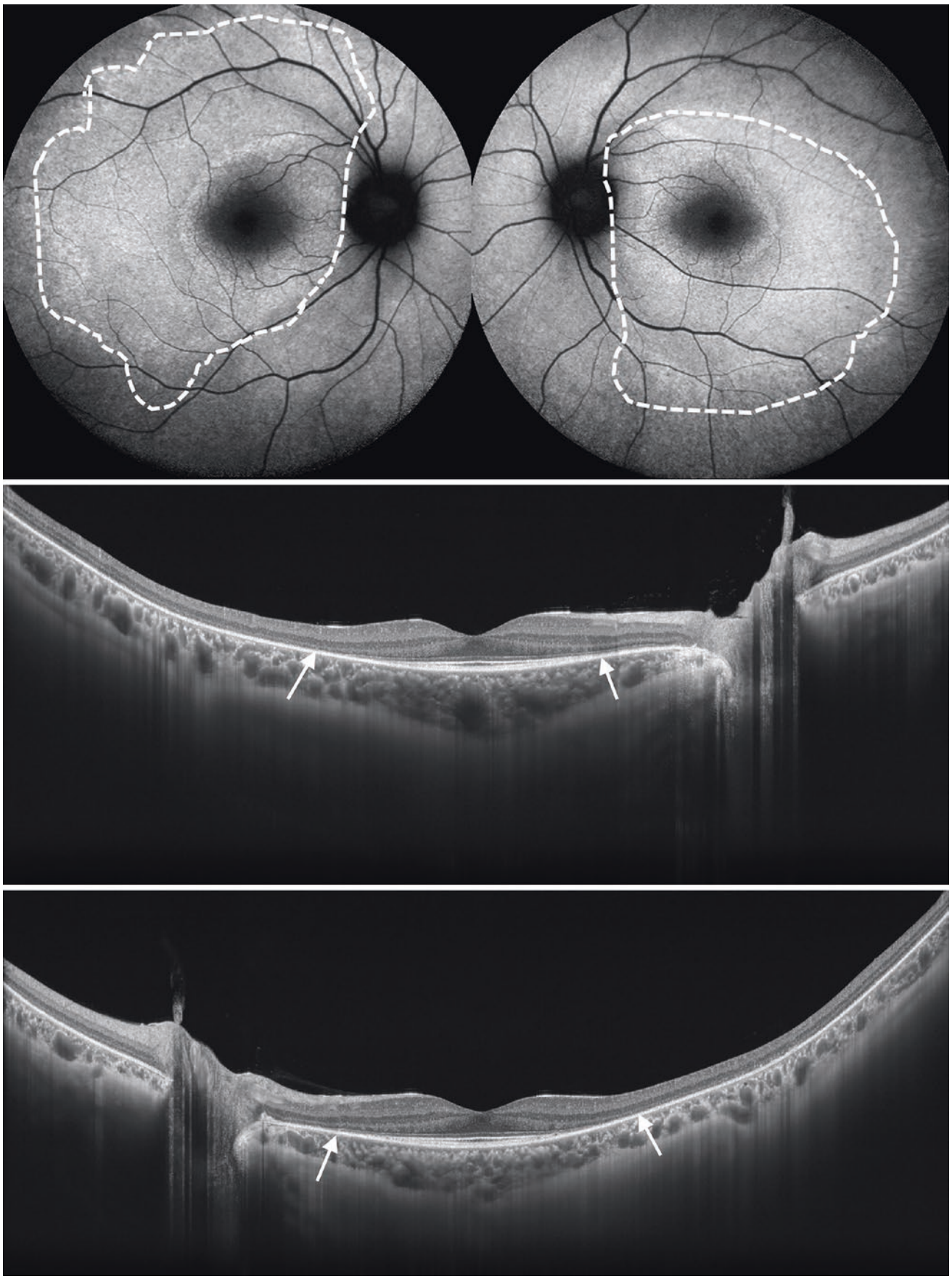

Fig. 11.25 Fundus autofluorescence and wide-angle SS-OCT single-line scan image. A 38-year-old female presented with bilateral photopsia. Her visual acuity is 20/20 in both eyes, Fundus autofluorescence showed demarked enhanced autofluorescence of macular in both eyes (dashed circles). SS-OCT showed the absence of ellipsoid zone, interdigitation zone, and external limiting membrane around the fovea (white arrows)

Multiple Evanescent White Dot Syndrome (MEWDS)

Multiple evanescent white dot syndrome (MEWDS) is an acute inflammatory disorder in which patients presented with unilateral visual field defect, blurred vision, and photopsia, commonly seen in young myopic women. MEWDS is usually self-limited, with recovery of vision and resolution of the lesion within a few weeks (Figs. 11.26, 11.27, 11.28 and 11.29).

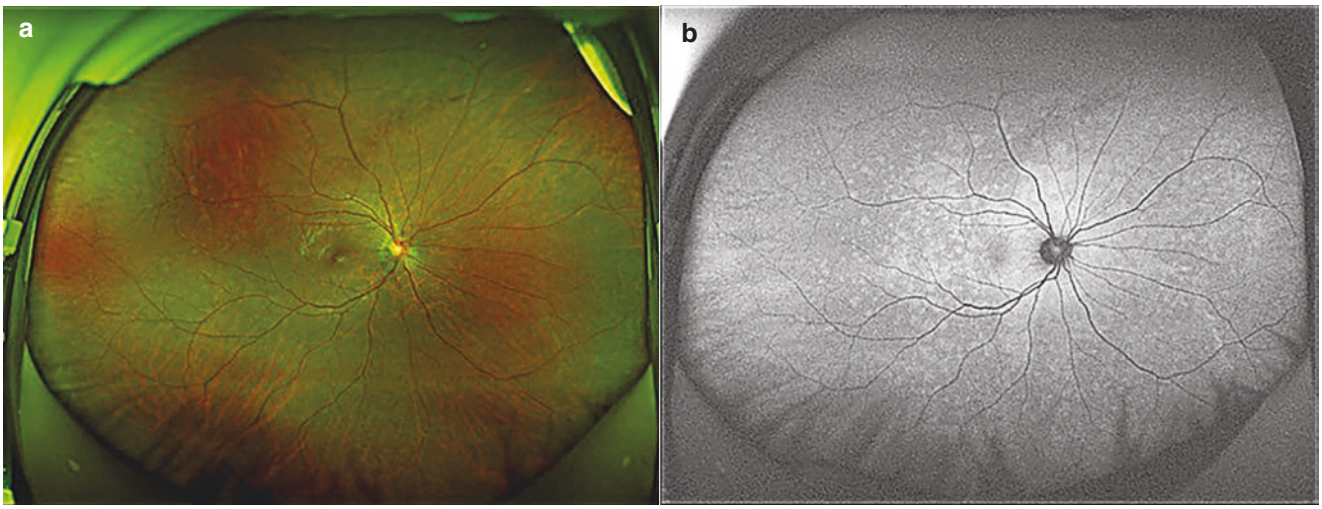

Fig. 11.26 Ultra-wide-field fundus photograph and fundus autofluorescence. A 28-year-old patient, with a sudden visual field loss in the right eye. Her best corrective visual acuity was 0.6, and visual field test showed temporal visual field defect in the right eye. (**a**) Ultra-wide-field color photograph demonstrates multiple small whitish dots throughout the posterior pole of the right eye. (**b**) Ultra-wide-field autofluorescence shows more hyperfluorescent spots than that can be identified on color photograph

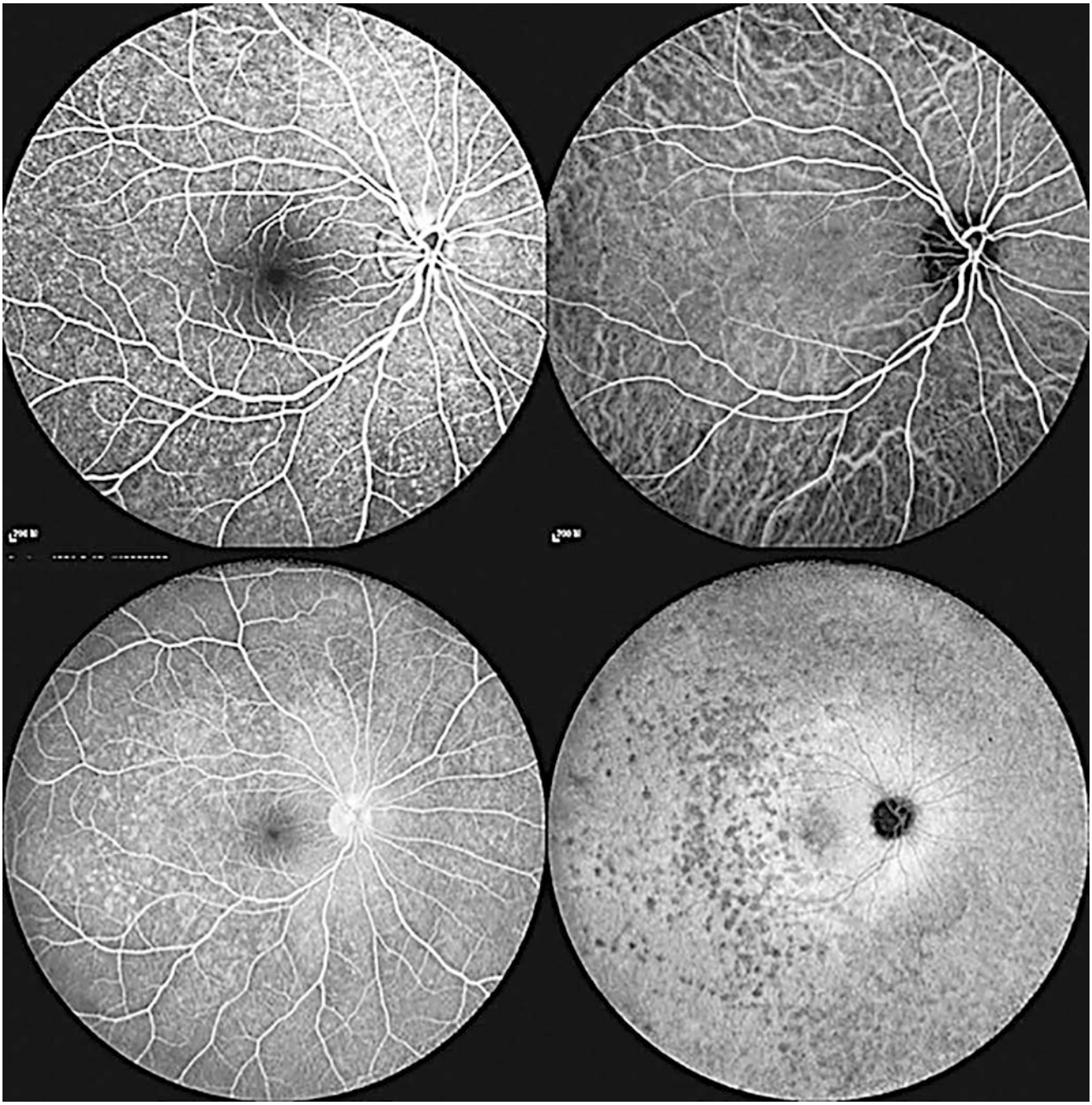

Fig. 11.27 FFA and ICGA images. In the same patient as in Fig. 11.26, FFA shows multiple punctate hyperfluorescent spots throughout the posterior pole of the right eye in early phase (upper left), with larger spots in the near peripheral region, and hyperfluorescence in the optic disc in the late phase (lower left); A corresponding ICGA shows widespread multiple hypofluorescent lesions best visualized in the late phase of ICGA

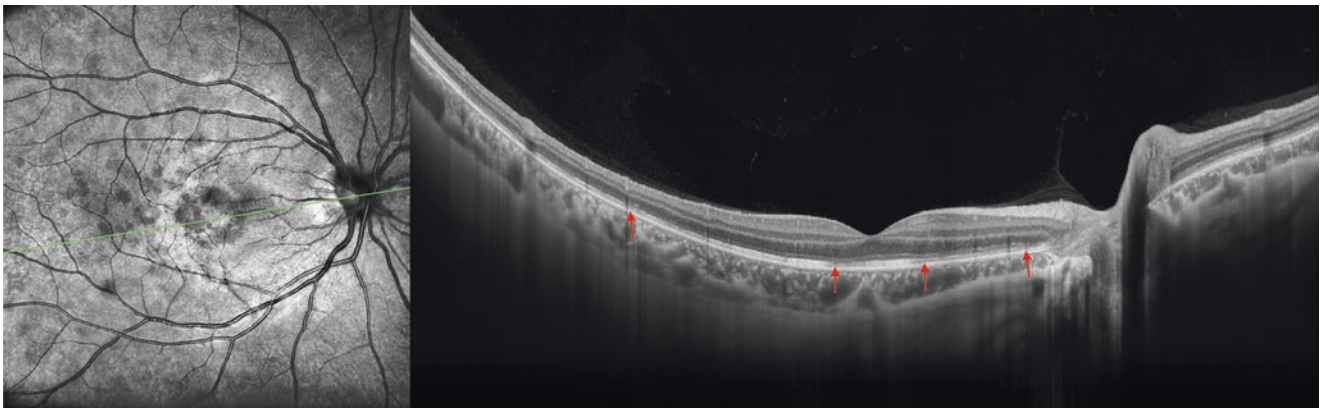

Fig. 11.28 Ultra-wide-field SS-OCT HD single-line scan. As in patient of Fig. 11.26, SS-OCT single-line scan shows an interrupted ellipsoid zone in the right eye (arrow)

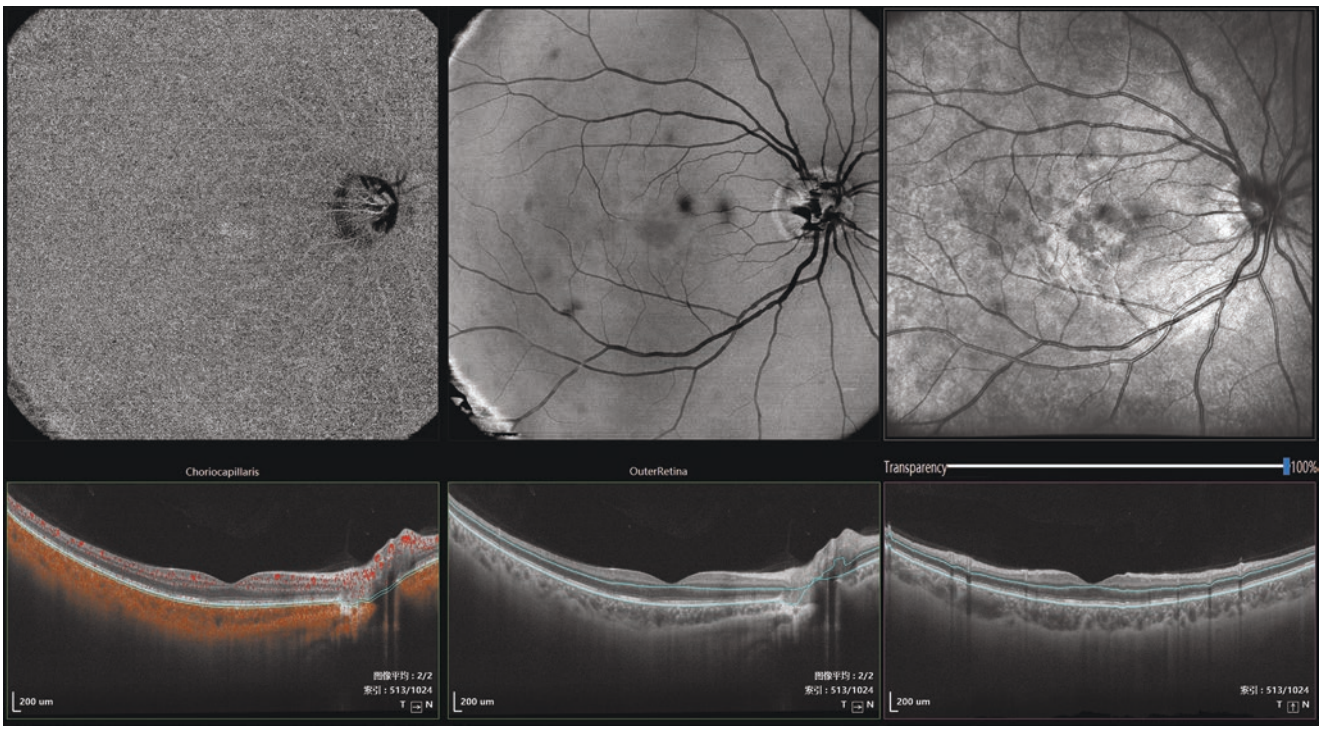

Fig. 11.29 SS-OCTA, en face SS-OCT, and cSSO images. In the same patient as in Fig. 11.26, OCTA (12 mm × 12 mm) shows the choroidal capillary layer without significant abnormalities; en face OCT shows a partial punctate lesion located in the outer nuclear layer; confocal scanning fundoscopic (cSSO) images clearly show the lesion located in the deep retinal layer

Overlapping "White Dot" Syndromes

Idiopathic inflammatory disorders that manifest as multifocal white dots or spots in the fundus are called "white dot syndromes," including multifocal choroiditis (PIC or multifocal choroidopathy, MFC), MEWDS, acute macular neuroretinopathy (AMN), and AZOOR. Some patients can present with more than one of these white dot syndromes, and usually referred to as overlapping "white dot" syndromes (Fig. 11.30, 11.31, 11.32, and 11.33)

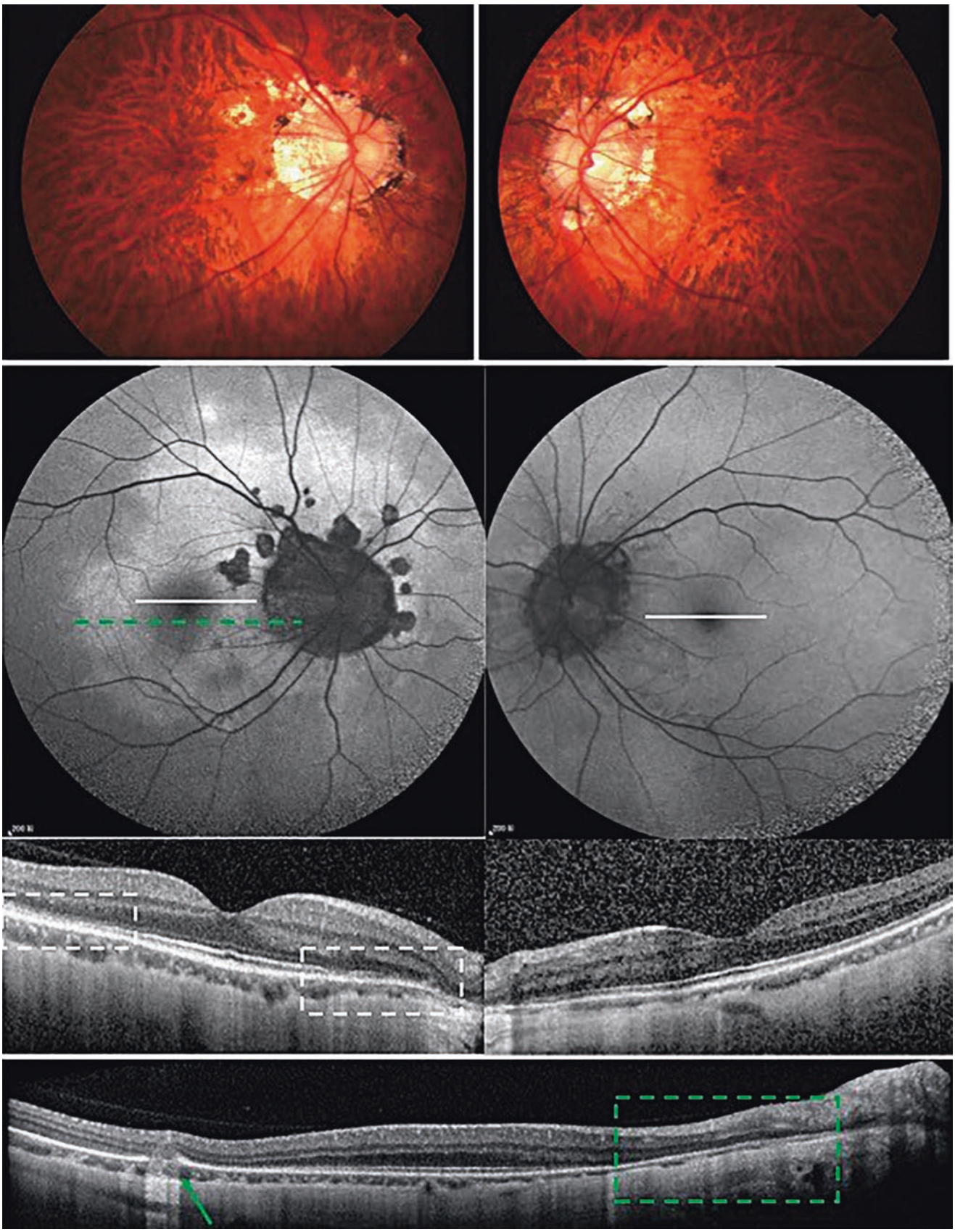

Fig. 11.30 Color fundus photograph, fundus autofluorescence, SD-OCT images. A 36-year-old male with high myopia presented with photopsia in the right eye for 1 month, color fundus photography shows multiple yellow-white spot lesions in the tessellated fundus of both eyes and a zonal defect consisting of peripapillary atrophy in the right eye. Fundus autofluorescence shows diffuse hyperfluorescence throughout the posterior pole of the right eye. SD-OCT shows obscured ellipsoid zone in the right eye (white dashed box), a chorioretinal lesion with RPE and EZ destruction located temporal to the fovea, which is an indication of inflammatory lesion (green arrow)

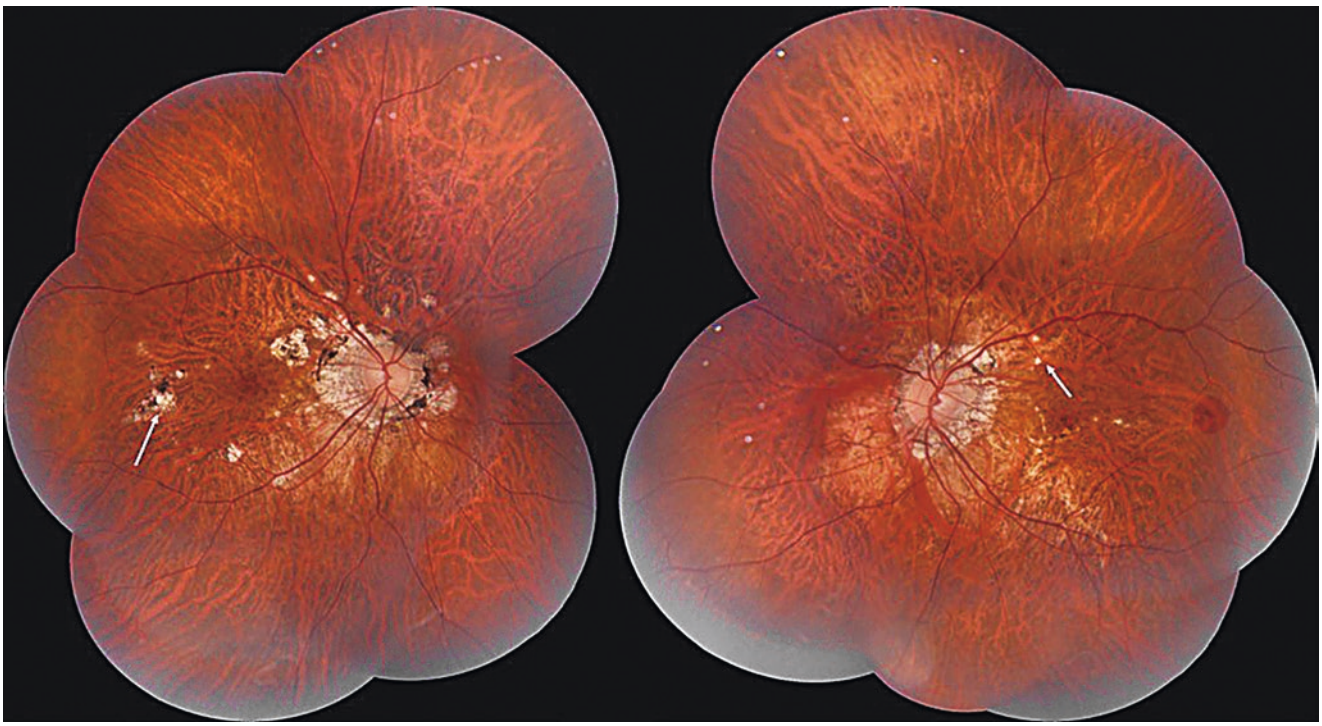

Fig. 11.31 Color fundus photograph. After 4 years of follow-ups, the patient developed more numbers of yellow-white lesions in both eyes, some of the lesions had pigment proliferation, and the choroid around the optic papilla developed diffused atrophy

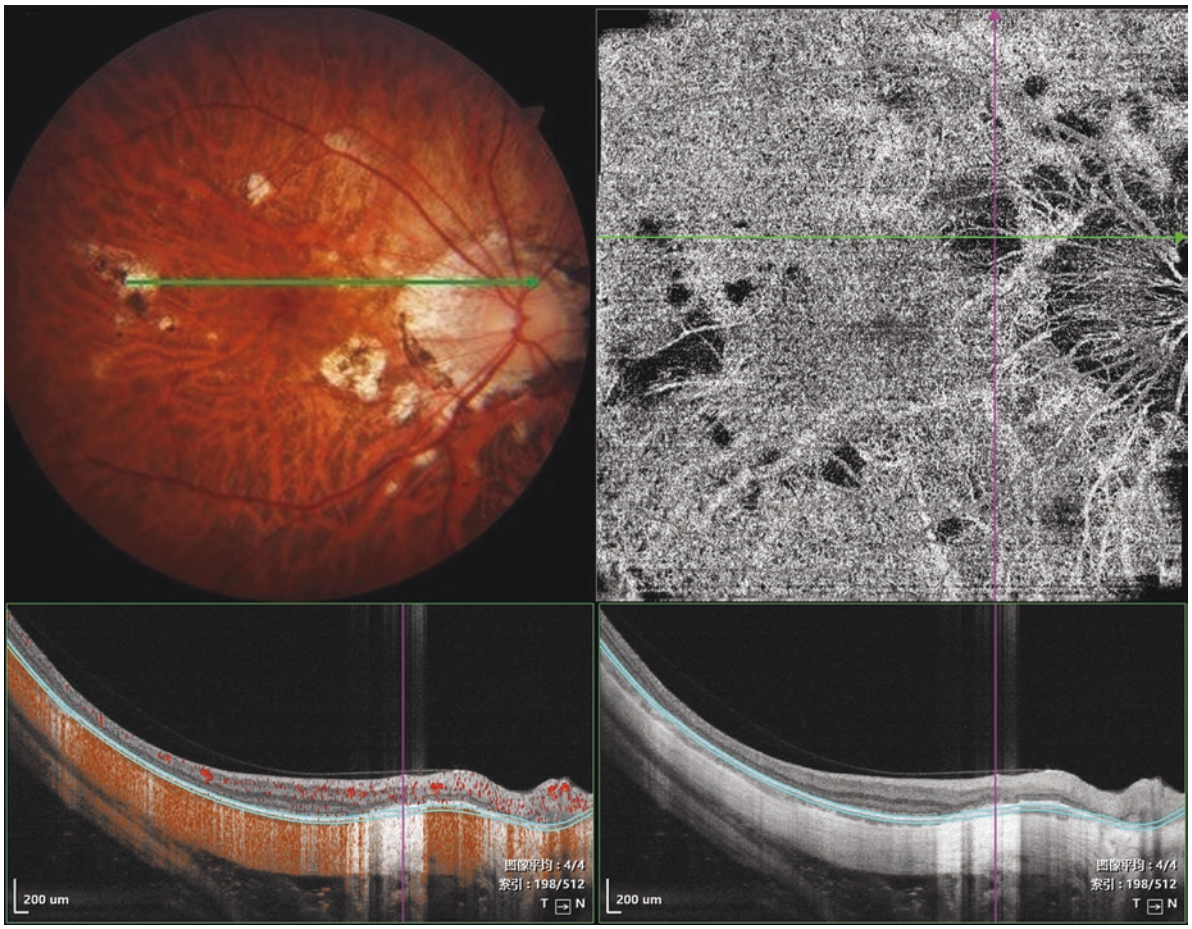

Fig. 11.32 Color fundus photograph, SS-OCTA images. After 4 years of follow ups, the patient in Fig. 11.30 developed more numbers of yellow-white lesions of the right eye, in which the choroid capillary layer shows atrophy and no blood flow signal in SS-OCTA 9 mm scan

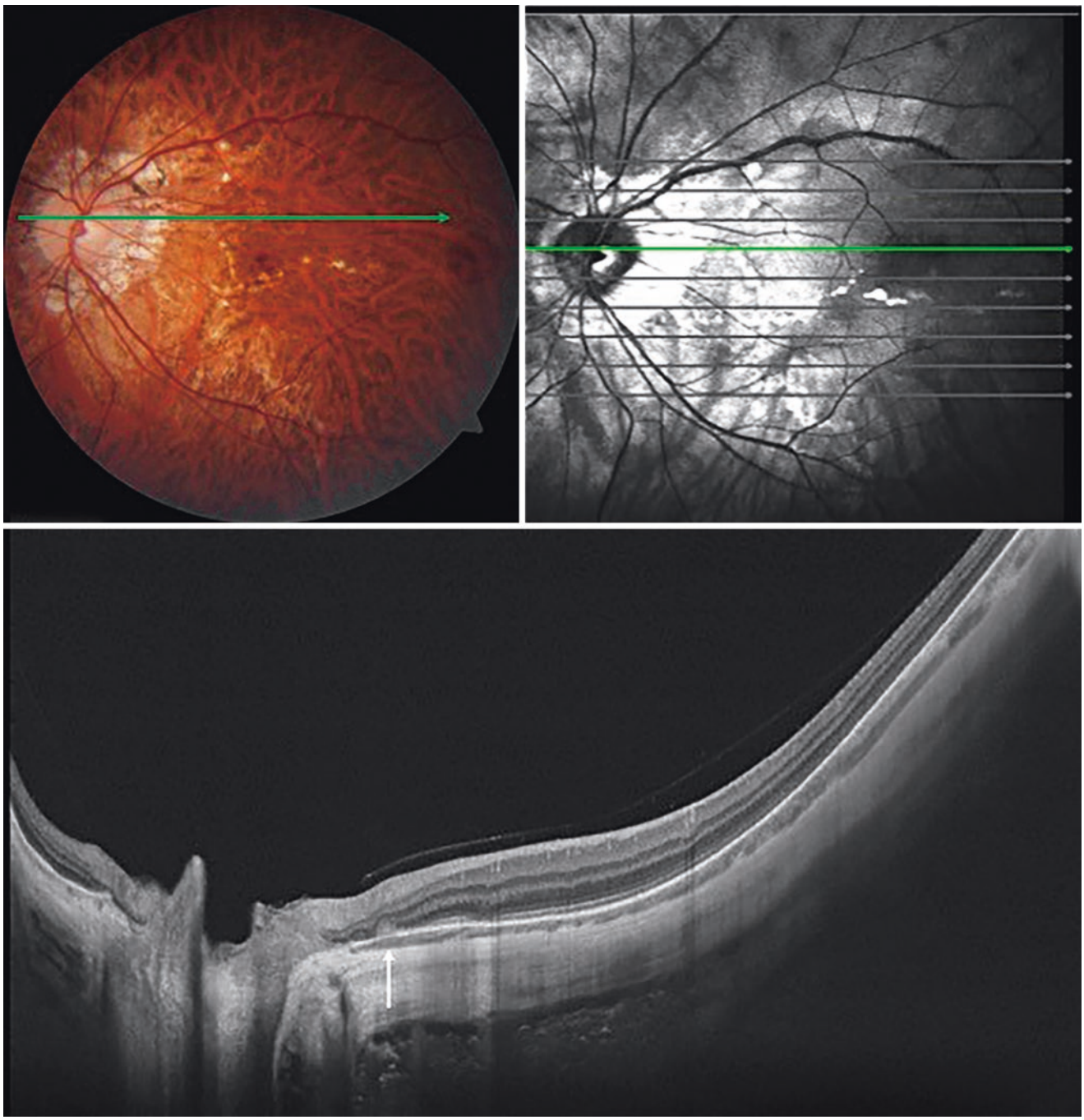

Fig. 11.33 Color fundus photograph, SS-OCTA images. After 4 years of follow-ups, the patient in Fig. 11.30 developed a new subretinal puncture lesion adjacent to the optic disc in the left eye, accompanied with EZ and RPE layer destruction

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