

New findings of swept-source optical coherence tomography angiography in Sturge-Weber syndrome with circumscribed choroidal hemangioma: a case report

Xinyu Yan

The Second Hospital of Hebei Medical University

Fengling Han

The Second Hospital of Hebei Medical University

Feiyan Ma

The Second Hospital of Hebei Medical University

Yuan Qi

The Second Hospital of Hebei Medical University

Ruonan Guo

The Second Hospital of Hebei Medical University

Chendai Niu

The Second Hospital of Hebei Medical University

Liya Liu

liuliya81@sina.com

The Second Hospital of Hebei Medical University

Case Report

Keywords: Optical coherence tomography angiography (OCTA), Sturge-Weber syndrome, Circumscribed choroidal hemangioma, laser photocoagulation, anti-VEGF

Posted Date: May 20th, 2026

DOI: <https://doi.org/10.21203/rs.3.rs-9581945/v1>

License: © ⓘ This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Additional Declarations: No competing interests reported.

Version of Record: A version of this preprint was published at Journal of Medical Case Reports on August 30th, 2026. See the published version at <https://doi.org/10.1186/s13256-026-06518-6>.

Abstract

Background

Circumscribed choroidal hemangioma is not associated with systemic diseases, while it can also occasionally occur in Sturge-Weber syndrome patients. Swept-source optical coherence tomography angiography (SS-OCTA) offers a non-invasive imaging modality for diagnosis and follow-up analysis of circumscribed choroidal hemangiomas. This study presents an uncommon instance of Sturge-Weber syndrome with circumscribed choroidal hemangioma, diagnosed and monitored over a three-year follow-up period using SS-OCTA.

Case presentation

A 16-year-old female presented with decreased vision in her right eye, with a ipsilateral facial port-wine birthmark. Multimodal imaging, particularly SS-OCTA, supports the diagnosis of circumscribed choroidal hemangioma located in the superotemporal quadrant with subretinal fluid. After initial laser photocoagulation, SS-OCTA showed decreased subretinal fluid at the tumor site but increased SRF over the fovea. Subsequently, the patient received intravitreal injection of anti-vascular endothelial growth factor (anti-VEGF) combined with additional laser photocoagulation. After 3-year follow-up using SS-OCTA, SRF had never recurred and choroidal vascularity index of choroidal hemangioma decreased.

Conclusions

SS-OCTA is helpful for the diagnosis of choroidal hemangioma and can also be used to assess treatment response by analyzing choroidal blood flow.

Background

Sturge-Weber syndrome (SWS) is a non-hereditary neurovascular disease characterized by abnormal vasculature in the brain, skin, and eye. The syndrome presents with a congenital facial capillary malformation that aligns with the trigeminal nerve, also known as port-wine birthmark (PWB). In addition, patients may have associated leptomeningeal vascular malformations seen on contrast-enhanced MRI images and ocular vascular abnormalities[1, 2]. SWS is most commonly associated with diffuse choroidal hemangioma (DCH). Circumscribed choroidal hemangioma (CCH) is typically a solitary lesion which is not associated with systemic diseases, while it is rarely seen in SWS patients. The treatment options for CCH include photodynamic therapy (PDT), laser photocoagulation, radiotherapy, transpupillary thermotherapy, and anti-VEGF therapy, among others[3]. The uniqueness of our case reports the relatively rare combination of typical SWS facial features and imaging features of CCH. In this study, SS-OCTA was employed during the diagnosis and treatment process to observe the changes

in hemodynamics and to quantitatively calculate choroidal vascularity index (CVI), which plays a crucial role in the assessment of disease condition.

Case presentation

A 16-year-old female presented with a one-week history of decreased vision in her right eye. She had been previously healthy and denied any history of seizures. Physical examination revealed a facial PWB involving the right forehead, upper eyelid, and nasal root (Fig. 1A). Best-corrected visual acuity (BCVA) was 20/40 in the right eye and 20/20 in the left eye. Intraocular pressures were 14 mmHg and 11 mmHg, respectively. Anterior segment examination was unremarkable. Dilated fundus examination revealed an orange-red, dome-shaped subretinal mass located at the superotemporal vascular arcade of the right eye, accompanied by exudative retinal detachment extending into the superonasal, superotemporal, and inferotemporal quadrants (Fig. 1B). Ocular ultrasonography demonstrated a solid choroidal mass with abundant internal blood flow (Fig. 1C), further confirmed by color Doppler flow imaging (Fig. 1D). SS-OCTA showed a markedly elevated choroidal mass with associated subretinal fluid (SRF) (Fig. 1E–F). Cranial MRI revealed no neurological abnormalities.

The patient underwent multimodal ophthalmic imaging including ultrawide-field fundus photography (UWFP), fluorescein angiography (FFA), indocyanine green angiography (ICGA), SS-OCTA, and ocular ultrasonography. Based on the presence of ipsilateral PWB and choroidal hemangioma, a diagnosis of SWS was made. Initial treatment consisted of laser photocoagulation targeting the choroidal hemangioma. Two weeks later, SS-OCTA showed decreased SRF at the tumor site but increased SRF over the fovea and nasal to the optic disc (Fig. 2A). The patient subsequently received intravitreal injection of anti-VEGF (Conbercept 0.5mg/0.05ml) combined with additional laser photocoagulation. At the three-month follow-up, SRF had completely resolved (Fig. 2B). BCVA in the right eye improved to 20/30 and remained stable, with no recurrence during a 3-year follow-up period. UWFP showed laser photocoagulation scars in the superotemporal retina and a normal macular appearance (Fig. 2C–D).

SS-OCTA revealed distinctive vascular changes. Before treatment, the choroidal layer showed densely packed, club-shaped anastomosing vessels within the tumor, transitioning to normal vascular patterns toward the margins (Fig. 3A). After treatment, blood flow signals were markedly reduced, particularly in areas corresponding to laser spots (Fig. 3B). At the choriocapillaris level, the hemangioma exhibited a well-defined perfusion defect pre-treatment (Fig. 3C), whereas post-treatment images showed loss of choriocapillaris flow with replacement by medium-to-large choroidal vessels (Fig. 3D). Using the SS-OCTA choroidal density map, the tumor were delineated and divided into multiple 1×1 mm grids. CVI was calculated within each grid using the advanced segmentation mode (Fig. 3E–H). The mean CVI within the tumor decreased markedly from 0.42 pre-treatment to 0.15 post-treatment.

Discussion and conclusions

OCTA offers a non-invasive imaging modality for characterization of choroidal hemangiomas, facilitating diagnostic confirmation and longitudinal monitoring of disease progression through hemodynamic parameter quantification, including CVI analysis. Traditionally, the diagnosis of choroidal hemangioma has primarily depended on FFA and ICGA. But it is invasive and not suitable for repeated use during follow-up to monitor tumor changes. However, SS-OCTA overcomes these limitations. Thanks to its longer wavelength, enhanced tissue penetration, and high scanning speed, SS-OCTA offers superior visualization of choroidal vascular structures and is especially advantageous for evaluating choroidal lesions[4].

This article reports a patient with SWS exhibiting characteristic ipsilateral facial PWB and ocular choroidal hemangiomas without abnormal neurological lesions. Choroidal hemangioma is a relatively common ocular manifestation of SWS, typically occurring in eyes with an ipsilateral PWB[5]. Although choroidal hemangioma in SWS often presents as diffused choroidal hemangioma, cases of CCH have also been documented. In 1999, Scott IU et al. first reported a case of SWS featuring DCH in one eye and CCH in the fellow eye[6]. A case of SWS combined with CCH was described in 2017[7]. Recently, Zhang et al. employed ultra-widefield SS-OCTA for detailed analysis of a case of SWS combined with CCH[8]. In this case, the available evidence appears to support the diagnosis of CCH. First of all, the tumor showed well-defined borders and was confined to the superotemporal arcade without other quadrants involvement. Besides, the basal width was 16.8mm measured by B-ultrasound, which exceeds the 12 mm mostly reported for CCH in the literature[9]. But its thickness was 5.4 mm and the diameter-to-height ratio exceeded 2, which is consistent with generally accepted diagnostic criteria of CCH[10, 11]. Furthermore, the sustained response of the choroidal hemangioma to combined therapy—with no recurrence observed during a 3-year follow-up—provides additional clinical supporting the diagnosis of CCH. In this report, SS-OCTA was utilized to assist in diagnosis and long-term treatment monitoring, elucidating the potential mechanisms of therapy and offering a valuable reference for the diagnosis and management of similar clinical cases in the future.

In our study, structure of retinal pigment epithelium (RPE) and photoreceptor layer was clearly visualized. Star-shaped highly reflective signals were attached to the RPE, with SRF extending to macula. A histopathological study on choroidal hemangioma in patients with SWS showed that these hemangiomas were mainly composed of cavernous, capillary, or mixed vascular types, with cavernous vascular type being the predominant type and capillary vascular type being the least[12]. This is consistent with changes observed at the corresponding blood flow level in this case. Choroidal blood flow layer shows rod-shaped abnormal communicating blood vessels inside the tumor and choriocapillaris slab shows hyporefectivity in the area of the lesion.

Although PDT remains a first-line intervention for subfoveal choroidal hemangiomas, clinical application remains limited by the high cost of photosensitizers and barriers to sustained drug availability. Therefore, the patient initially underwent laser photocoagulation. OCTA revealed RPE atrophy at the photocoagulation sites and regression of SRF over the tumor. However, SRF at the macular fovea increased. The presence of SRF scatters laser energy and impairs effective chorioretinal adhesion,

thereby resulting in a high recurrence rate[9]. So the patient received intravitreal injection of anti-VEGF combined with additional laser photocoagulation. SS-OCTA after treatment revealed that the previously intermingled club-shaped anastomosing intratumoral vessels in the choroidal layer were replaced by hyporefectivity at the laser photocoagulation sites. There was no significant change in the middle and large vessels of the choroid after treatment, but obvious flow voids appeared in the capillary layer. Quantitative analysis demonstrated a marked decrease in CVI within the hemangioma region after treatment. Both morphological assessment and quantitative analysis led to the same conclusion: laser photocoagulation induces occlusion of tumor vessels, especially choriocapillaris, which is consistent with hyporefectivity in the blood slabs demonstrated by SS-OCTA. This observation aligns with the previous report by Chawla et al., which indicated a laser-induced reduction of the overlying choriocapillaris in such lesions[13]. Throughout the three-year follow-up period, there was no apparent restoration of flow in the choriocapillaris in the lasered area and SRF did not occur. Histopathological analyses of laser scars in human subjects have similarly documented persistent choriocapillaris disruption with no evidence of subsequent regeneration[14, 15]. In the treatment of this case of choroidal hemangioma, anti-VEGF therapy may play a certain role, which may reduce tumor-associated vascular leakage, potentially by depleting endothelial fenestrations and altering intercellular adhesion molecules[16]. Liu et al. first confirmed elevated levels of VEGF-A in the aqueous humor of CCH patients, offering direct pathological evidence supporting the clinical application of anti-VEGF therapy[17]. While intravitreal anti-VEGF monotherapy can achieve partial resolution of associated retinal edema, its efficacy in inducing regression of CCH itself remains limited[18]. Emerging clinical evidences demonstrates that intravitreal injection of anti-VEGF combined with laser photocoagulation can effectively control CCH[19, 20], suggesting that combined therapy might be a safe and effective method for the treatment of CCH. On the one hand, intravitreal injection of anti-VEGF reduces vascular leakage and facilitates SRF absorption, thereby enabling more efficient delivery of laser energy to the lesion. On the other hand, intravitreal injection of anti-VEGF may ameliorate laser-associated inflammation, further promoting fluid resolution, and potentially decreasing the risk of recurrent macular edema after additional laser treatment. Therefore, this combined treatment presents a clinically effective and cost-efficient treatment strategy, which validates the efficacy for cases involving the macula.

In summary, this report documents a case of a patient featuring the cutaneous manifestation of SWS co-existing with the CCH. The patient was managed with combined therapy of laser photocoagulation and intravitreal anti-VEGF injections. Throughout the 3-year follow-up period, SRF exhibited no recurrence, validating this treatment as a viable alternative for patients inaccessible to PDT. Furthermore, OCTA not only reveals pathological mechanisms underlying CCH through comprehensive analysis of blood flow dynamics, vascular density and vascular architecture, but also serves as a valuable tool for dynamic monitoring and long-term follow-up assessment. OCTA can also be employed for quantification of hemodynamic changes before and after treatment through calculating CVI within defined regions, which improves the objectivity and comparability of imaging-based evaluations.

There were also several limitations. First, the limitation of SS-OCTA, particularly its restricted penetration depth, compromises detailed visualization of deeper vascular plexuses within the tumor. Most

importantly, there is a deficiency persisting in diagnostic criteria for choroidal tumors utilizing SS-OCTA imaging. Therefore, large-sample randomized controlled trials are needed to validate this therapy's efficacy and establish standardized guidelines.

Abbreviations

SS-OCTA

swept-source optical coherence tomography angiography

OCTA

optical coherence tomography angiography

SWS

Sturge-Weber syndrome

PWB

port-wine birthmark

DCH

diffuse choroidal hemangioma

CCH

circumscribed choroidal hemangioma

PDT

photodynamic therapy

CVI

choroidal vascularity index

BCVA

best-corrected visual acuity

SRF

subretinal fluid

UWFP

ultrawide-field fundus photography

FFA

fluorescein angiography

ICGA

indocyanine green angiography

RPE

retinal pigment epithelium

Anti-VEGF

anti-vascular endothelial growth factor

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was obtained from the patient's parent or legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

All data generated or analysed during this study are included in this published article.

Competing interests

The authors declare that they have no competing interests.

Funding

This study was sponsored by Hebei Provincial Medical Science Research Project Program (20211081 and 20240041).

Authors' contributions

X.Y. Yan collected the data and drafted the manuscripts. F.Y. Ma provided revision suggestions. L.Y. Liu treated and admitted the patients. All the authors revised the manuscript and approved its submission.

Acknowledgements

Not applicable.

Additional information

Publisher's note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Supplementary Information

All data generated or analysed during this study are included in this published article.

References

1. Dingenen E, Segers D, De Maeseneer H, et al. Sturge-Weber syndrome: an update for the pediatrician. *World J Pediatr.* 2024;20:435-43.
2. Yeom S, Comi AM. Updates on Sturge-Weber Syndrome. *Stroke.* 2022;53:3769-79.

3. Lupidi M, Centini C, Castellucci G, et al. New insights on circumscribed choroidal hemangioma: "bench to bedside". *Graefes Arch Clin Exp Ophthalmol*. 2024;262:1093-110.
4. Gündüz AK, Mirzayev I, Kasimoglu R, et al. Swept-source optical coherence tomography angiography findings in choroidal and retinal tumors. *Eye (Lond)*. 2021;35:4-16.
5. Silverstein M, Salvin J. Ocular manifestations of Sturge-Weber syndrome. *Curr Opin Ophthalmol*. 2019;30:301-5.
6. Scott IU, Alexandrakis G, Cordahi GJ, et al. Diffuse and circumscribed choroidal hemangiomas in a patient with Sturge-Weber syndrome. *Arch Ophthalmol*. 1999;117:406-7.
7. Tripathy K, Das A, Subhadarshani S. Sturge-Weber Syndrome with Choroidal Hemangioma. *Indian Dermatol Online J*. 2017;8:225-6.
8. Zhang Z, Zhu R, Yang L. Choroidal hemangioma in Sturge-Weber syndrome elucidated by ultra-widefield swept-source optical coherence tomography angiography. *BMC Ophthalmol*. 2025;25:492.
9. Yang Z, Tian D, Xie Z, et al. Clinical features, diagnosis, management, and prognosis of circumscribed choroidal hemangioma. *Surv Ophthalmol*. 2025;70:389-400.
10. Witschel H, Font RL. Hemangioma of the choroid. A clinicopathologic study of 71 cases and a review of the literature. *Surv Ophthalmol*. 1976;20:415-31.
11. Krohn J, Rishi P, Frøystein T, et al. Circumscribed choroidal haemangioma: clinical and topographical features. *Br J Ophthalmol*. 2019;103:1448-52.
12. Helmi HA, Alkatan HM, Al-Essa RS, et al. Choroidal hemangioma in Sturge Weber syndrome: Case series with confirmed tissue diagnosis. *Int J Surg Case Rep*. 2021;89:106626.
13. Chawla R, Tripathy K, Sharma A, et al. Swept source optical coherence tomography-angiography of choroid in choroidal hemangioma before and after laser photocoagulation. *Indian J Ophthalmol*. 2017;65:751-4.
14. Wilson DJ, Green WR. Argon laser panretinal photocoagulation for diabetic retinopathy. Scanning electron microscopy of human choroidal vascular casts. *Arch Ophthalmol*. 1987;105:239-42.
15. Stitt AW, Gardiner TA, Archer DB. Retinal and choroidal responses to panretinal photocoagulation: an ultrastructural perspective. *Graefes Arch Clin Exp Ophthalmol*. 1995;233:699-705.
16. Hang A, Feldman S, Amin AP, et al. Intravitreal Anti-Vascular Endothelial Growth Factor Therapies for Retinal Disorders. *Pharmaceuticals (Basel)*. 2023;16.
17. Liu K, Guo L, Cheng Y, et al. Cytokine Concentrations in Aqueous Humor of Eyes With Circumscribed Choroidal Hemangioma. *Front Endocrinol (Lausanne)*. 2022;13:916789.
18. Elkayal HE, Sagoo MS, Negretti GS. INTRAVITREAL BEVACIZUMAB FOR THE TREATMENT OF CHOROIDAL HEMANGIOMAS. *Retina*. 2025;45:501-8.
19. Zhang WY, Chen J, Liu HM, et al. Bilateral circumscribed choroidal hemangioma: a case report. *Int J Ophthalmol*. 2024;17:2141-4.
20. Lai K, Gong Y, Xu F, et al. Effectiveness and Safety of Intravitreal Injection of Conbercept as an Initial Treatment for Exudative Circumscribed Choroidal Hemangioma. *Ophthalmologica*. 2020;243:436-

Figures

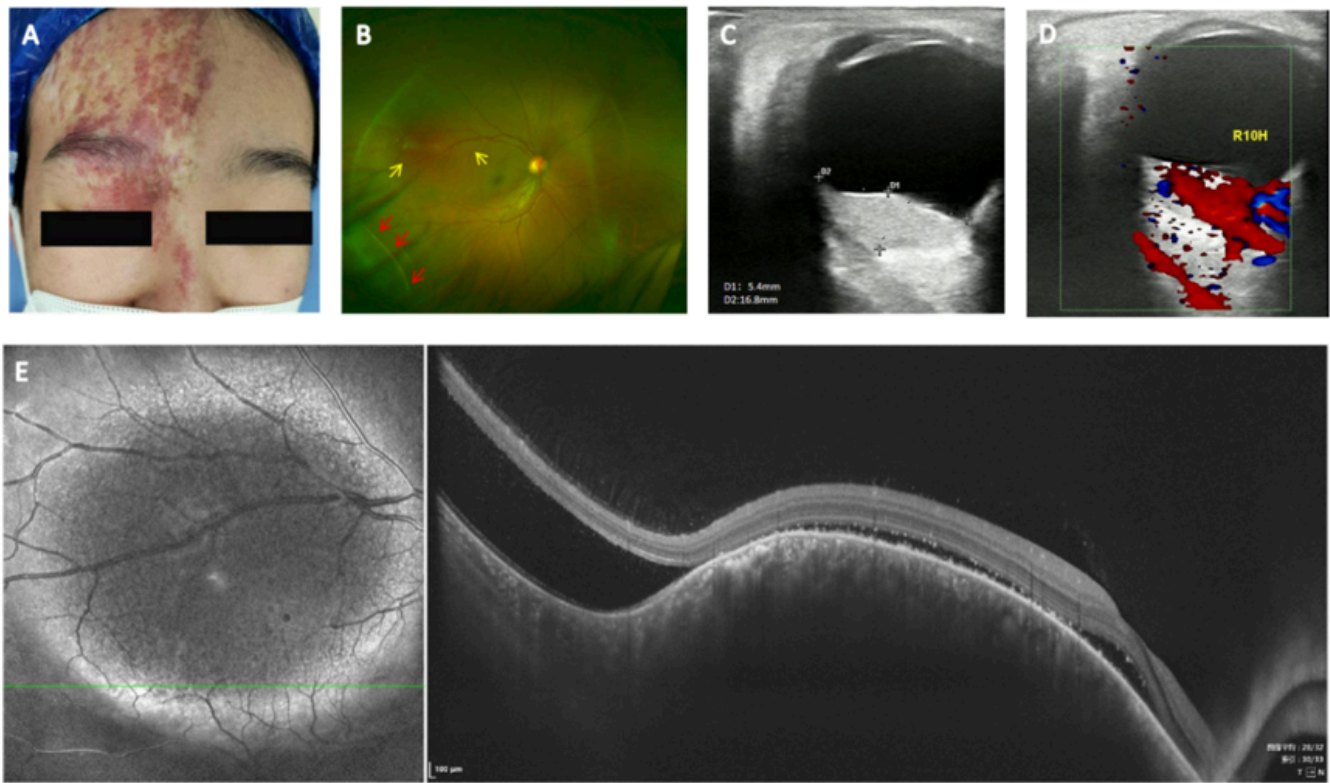


Figure 1

Multi-model imaging: A. Cutaneous examination reveals extensive port-wine stain distribution involving the right frontal region, upper eyelid, and nasal root. B. UWFP demonstrates a well-circumscribed orange-red lesion (maximum tumor diameter denoted by yellow arrows) in the superotemporal quadrant, accompanied by inferior exudative retinal detachment (delineated by red arrows). C. Ocular ultrasonography identifies a solid elevated mass (height: 5.4 mm; basal width: 16.8 mm) with moderately homogeneous internal echoes at the superotemporal ocular wall. D. Color Doppler flow imaging (CDFI) confirmed prominent intralesional blood flow. E. SS-OCTA near-infrared imaging illustrates the choroidal tumor's transverse elliptical configuration. F. Corresponding SS-OCTA section reveals a highly elevated sub-retinal pigment epithelium (RPE) mass with poorly defined inferior border, surrounded by SRF accumulation extending to the foveal avascular zone.

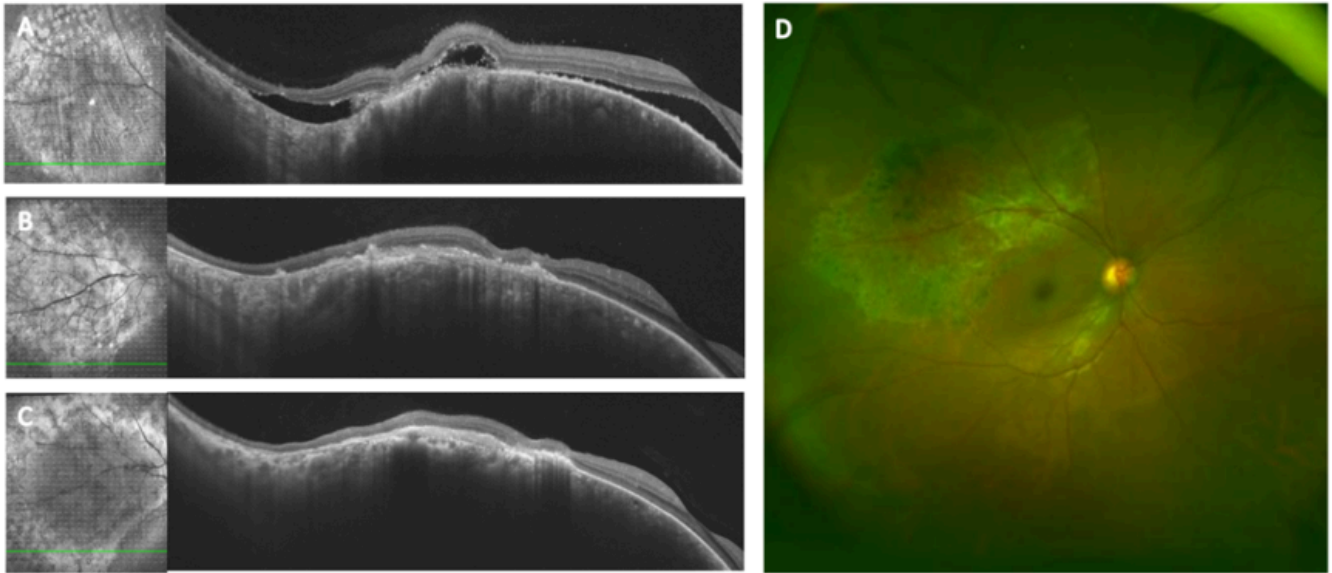


Figure 2

B-scan findings of OCTA and SLO during the follow-up period. A. Post-initial retinal laser photocoagulation, SS-OCTA demonstrates partial resolution of SRF overlying the tumor, while the SRF at the macular fovea increased. B. Following combined anti-VEGF therapy and supplemental photocoagulation, complete SRF absorption is achieved. C. Three-year follow-up reveals durable therapeutic efficacy with stabilized tumor dimensions and no evidence of active progression. D. UWFP documents residual laser photocoagulation scars overlapping the regressed tumor mass. No subretinal fluid is observed and the structure of the macular fovea is clear.

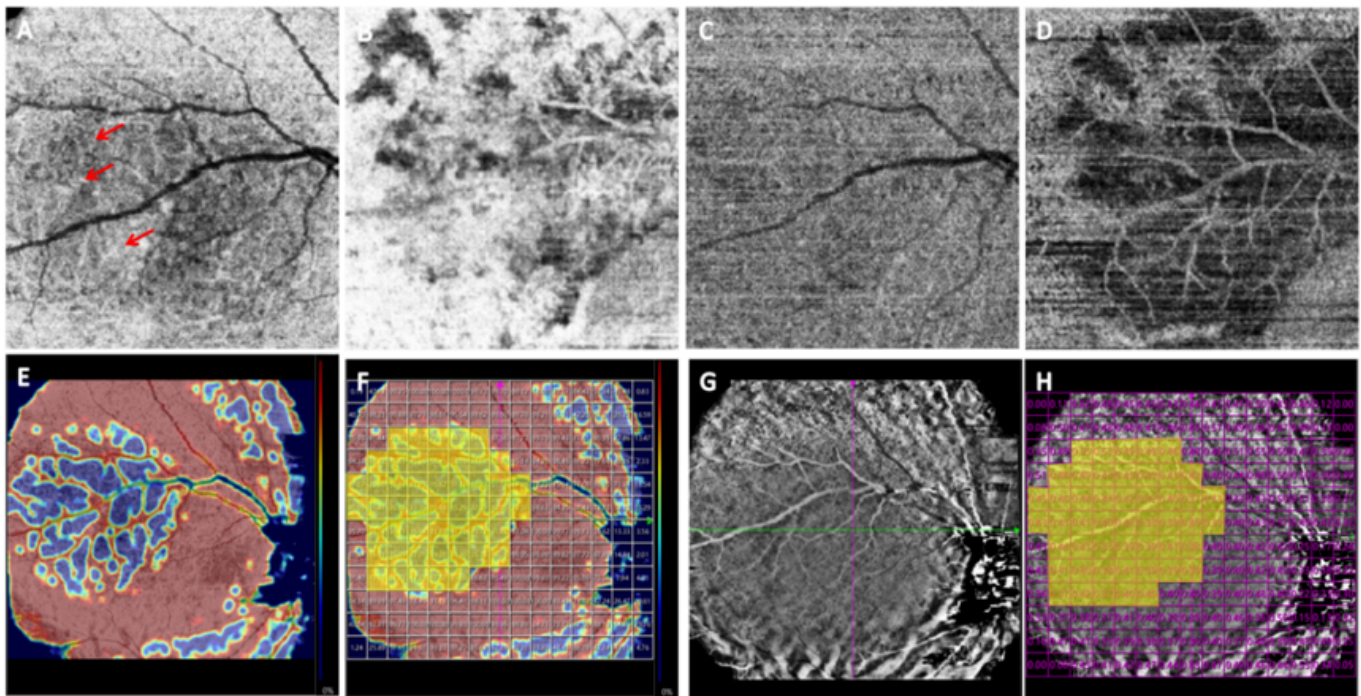


Figure 3

Corresponding en-face angiography images. A-D: SS-OCTA images before and after treatment. A. The choroidal blood flow layer shows rod-shaped abnormal communicating blood vessels inside the tumor (delineated by red arrows), while the tumor boundary is surrounded by normal choroidal vasculature. B. SS-OCTA through the same area 3 years after co-treatment shows disruption of abnormal choriocapillaris. C. Choriocapillaris slab shows hyporeflectivity in the area of the lesion. D. After co-treatment, SS-OCTA reveals choriocapillaris slab reduced signal flow and predominance of crisscrossing medium and large vessels. E-H: Methods for quantification blood flow within the tumor. E, F. Tumor visualization: Using the software provided by the equipment, 1mm×1mm grids are placed over the choroidal vascular layer to locate the tumor area. If the edge of the tumor occupies $\geq 1/2$ of the grid area, the data at this location will be retained. The yellow area in Figure F represents the tumor area. G, H. Advanced analysis. The advanced segmentation mode shows the choroidal vascular index, and the yellow area at the same position is selected for CVI data analysis.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- [CAREchecklistEnglish.pdf](#)