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Successful treatment of choroidal hemangiomas in Sturge-Weber syndrome using external beam radiotherapy

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Abstract

Sturge-Weber syndrome (SWS) is a rare congenital disorder with diverse manifestations, including diffuse choroidal hemangiomas causing visual impairment. This case report describes a 14-year-old girl with retinal detachment due to diffuse choroidal hemangioma, treated effectively using external beam radiation therapy (EBRT). The treatment achieved a significant 55% reduction in choroidal thickness after the first phase and near-complete remission after supplementary therapy. This case underscores EBRT's efficacy and highlights the potential of adaptive radiation strategies for managing complex SWS-related ocular complications. Early intervention remains crucial for optimizing patient outcomes.

Keywords Sturge-Weber Syndrome, Radiotherapy, Choroidal hemangioma

Background

Sturge-Weber (SW) syndrome is a rare, non-hereditary congenital disorder affecting the skin, eyes, and brain. Abnormal blood vessel formation can lead to a port-wine stain on the face, glaucoma, and leptomeningeal hemangiomas, contributing to seizures and stroke-like episodes [1, 2]. Other presentations include visual deficits and intellectual impairment, with diffuse choroidal hemangioma occasionally leading to retinal detachment [3]. SW syndrome has three classifications: Type I involves neurological and ocular issues with a port-wine stain, Type II has only a port-wine stain, and Type III includes neurological symptoms without the stain. Prognosis is often poor for cases with retinal detachment. Treatment varies based on symptoms, often aiming to manage and prevent complications. This report presents a young SW syndrome patient with retinal detachment from diffuse choroidal hemangioma, treated effectively with external beam radiation therapy (EBRT).

Case presentation

A 14-year-old girl with Sturge-Weber syndrome (SWS) had been followed at China Medical University Hospital for 12 years. Her birth history was uneventful, with no family history of similar disorders. Symptoms began at 2 years and 5 months with her first seizure—a prolonged right-sided partial seizure progressing to secondary generalization, necessitating long-term valproate treatment.

At age three, she developed uncontrolled intraocular pressure (IOP) in the left eye (20 mmHg), requiring anti-glaucoma medications, including atropine, azarga, brimonidine eyedrops, and oral acetazolamide. Despite treatment, glaucoma persisted. She also experienced recurrent aseptic meningitis, likely due to leptomeningeal hemangiomas associated with SWS.

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During her latest visit, she reported progressive blurry vision in the left eye. Physical examination revealed a unilateral port-wine stain from her right forehead and eye to her nose and lips, noted since birth and progressively enlarging. Ophthalmologic evaluation showed best-corrected visual acuity (BCVA) of 1.0 (right eye) and 0.02 (left eye), with IOPs of 21 mmHg (right eye) and 25 mmHg (left eye). Fundoscopy revealed a thickened, folded choroid in the left eye with a "tomato ketchup" appearance, consistent with diffuse choroidal hemangioma, accompanied by bullous retinal detachment (Fig. 1a).

The patient was primarily followed by a glaucoma specialist, with imaging focused on glaucoma-related assessments. The only available fundus color image was from 2019, as routine retinal and choroidal evaluations were not performed. In 2023, following a significant decline in vision, ultrasonography was performed. OCT was attempted but was not feasible due to severe retinal detachment, limiting direct assessment of retinal and choroidal changes.

Imaging, including B-scan ultrasonography (Picture 1b) and pre-treatment MRI (Fig. 2a), confirmed a diffuse choroidal hemangioma with retinal and vitreous hemorrhage, with an initial choroidal thickness of 12.14 mm. Given the significant choroidal thickening and progressive visual deterioration, EBRT was considered the most appropriate treatment to prevent further vision loss.

Treatment was planned using intensity-modulated radiation therapy (IMRT) with a Varian Halcyon linear accelerator, delivering a total dose of 30 Gy in two phases: 20 Gy in 10 fractions, followed by an additional 10 Gy in 5 fractions. Most reported EBRT protocols for diffuse choroidal hemangioma, including Randon et al. [4], have used 20 Gy in 10 fractions as a standard approach. In our case, after the first phase, CT imaging (Fig. 2b) showed a reduction in choroidal thickness to 5.41 mm, but residual thickening persisted. Following a three-month interval, organ-at-risk (OAR) constraints were reassessed, and it was determined that an additional radiation boost could be safely administered. After discussion with the patient's family, we proceeded with the second phase, totaling 30 Gy, to further reduce tumor volume while minimizing radiation-related complications. Post-treatment CT imaging (Fig. 2c) demonstrated a near-complete tumor response. External beam radiotherapy treatment plan was shown (See Fig. 3).

Follow-up imaging demonstrated a marked reduction in choroidal thickness to 5.41 mm after the initial phase of radiation, representing a 55% shrinkage (Fig. 2b). Optical coherence tomography (OCT) showed intraretinal fluid and bacillary layer detachment (BALAD) (Fig. 1c). Upon completion of the second phase of treatment, imaging revealed a near-complete response (Fig. 2c), with significant flattening of the retinal layers (Fig. 1d). Upon follow-up appointments, the patient expressed previous low vision has improved to some luminance contrast. However, due slow response from the patient, it was difficult to determine the extent of vision improvement.

Informed consent was obtained for this case report, and ethical approval was granted by China Medical University Hospital.

Discussion

Early detection and monitoring of choroidal hemangioma in SWS patients remain critical to preserving vision. In this case, the absence of long-term retinal imaging prior to disease progression contributed to delayed diagnosis and treatment. This highlights the need for regular fundus examinations, particularly in SWS patients with glaucoma, where retinal evaluation may not always be prioritized. External beam radiation therapy (EBRT) is an effective treatment for diffuse choroidal hemangioma (CH) in SWS patients. Traditional options, such as scatter photocoagulation, cryotherapy, and brachytherapy, aimed to slow tumor progression and preserve vision but often achieved limited success, particularly in cases with subretinal fluid [3].

In this case, EBRT achieved a significant 55% reduction in choroidal thickness after the first phase, consistent with prior studies. Randon et al. [4] also reported stabilization of visual acuity and reduced tumor thickness with low-dose radiation (20 Gy in 10 fractions), though mild cataracts occurred in some patients. Notably, our patient developed no cataracts, but long-term monitoring remains crucial to detect late complications. Previous studies have highlighted potential long-term risks of EBRT, such as radiation-induced cataracts, ocular surface disorders, optic neuropathy, and retinopathy [5, 6]. This underscores the need for individualized radiation dose planning to optimize therapeutic outcomes while preserving visual function. Continued ophthalmic follow-up is essential to detect and manage late-onset radiation complications.

Proton therapy, an alternative modality, offers precise tumor targeting while sparing surrounding tissues due to its Bragg peak, making it a promising option for pediatric ocular tumors [7].

To our knowledge, this is the first reported case of total remission of diffuse choroidal hemangioma following an additional course of radiation therapy. Unlike cases achieving only partial remission after a single treatment, this patient's response suggests that supplementary radiation may be beneficial for similar cases. This outcome underscores the importance of adaptive treatment strategies in managing complex SWS presentations.

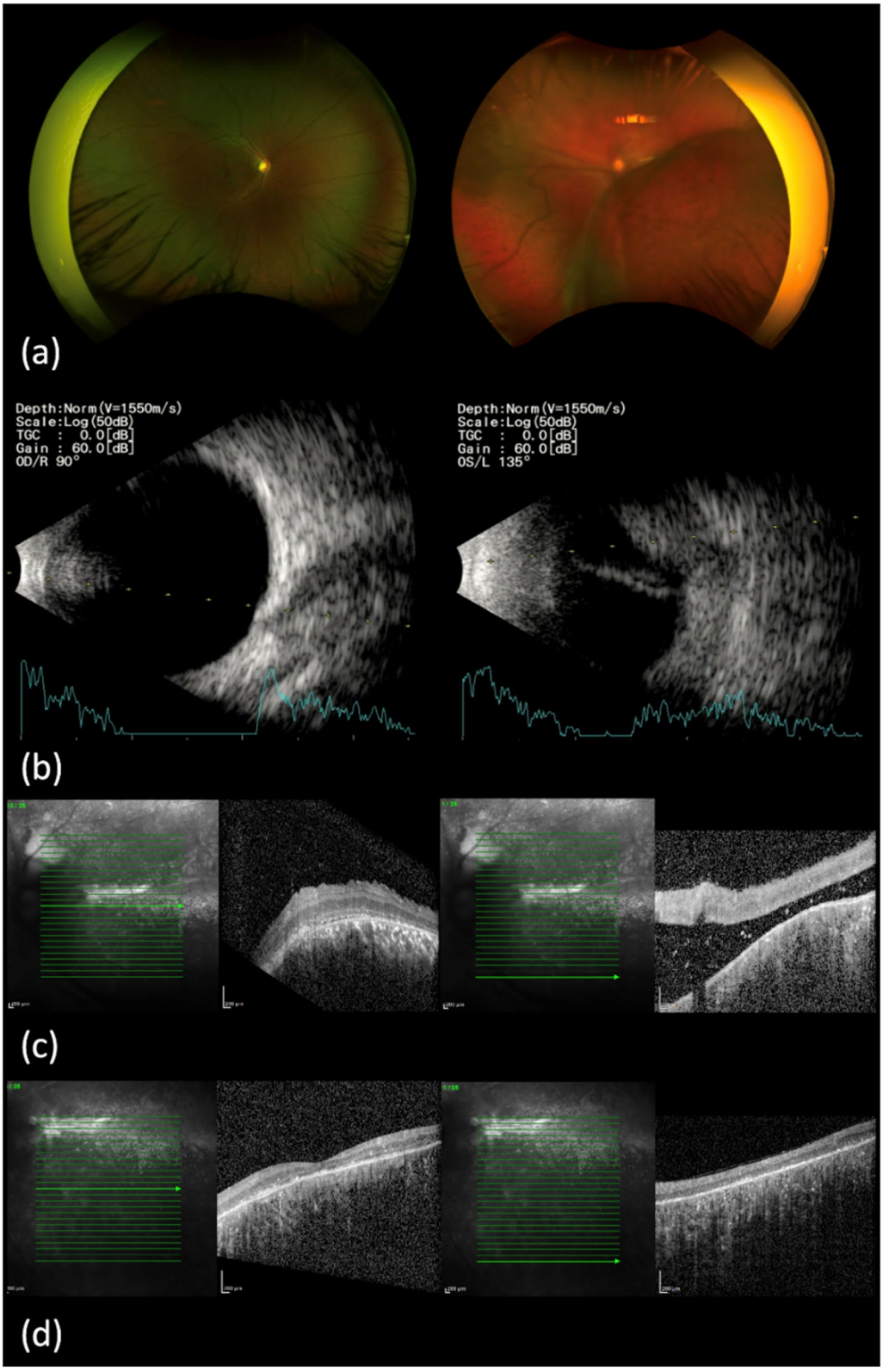

Fig. 1 (**a**) RE / LE of the fundus examination: LE Thick and folded choroid, with dark and saturated red area similar to a "tomato ketchup" appearance. (**b**) RE / LE of the ultrasound B scan: diffuse thickening of the choroid and retinal detachment in the LE. (**c**) OCT after initial phase: Bacillary layer detachment (BALAD) and intraretinal fluid still observed after the initial phase of treatment. (**d**) OCT after second phase: restoration of retinal morphology with resolution of the intraretinal fluids, flattening of the retinal layers, and no hyperreflectivity seen

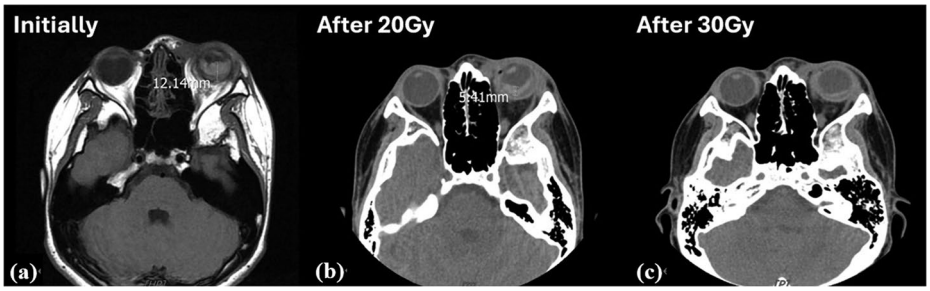

Fig. 2 Presents imaging results from different modalities at various treatment stages. (**a**) Pre-treatment MRI showing thickness and enhancement of the retina of the left eyeball with angiomatosis; initial choroidal thickness of 12.14 mm. (**b**) Post-first-phase CT after 20 Gy, demonstrating a reduction in choroidal thickness to 5.41 mm. (**c**) Post-second-phase CT after a total dose of 30 Gy, showing near-complete response. Apparent differences in lesion size are due to variations in imaging modality and slice thickness rather than true changes in tumor dimensions

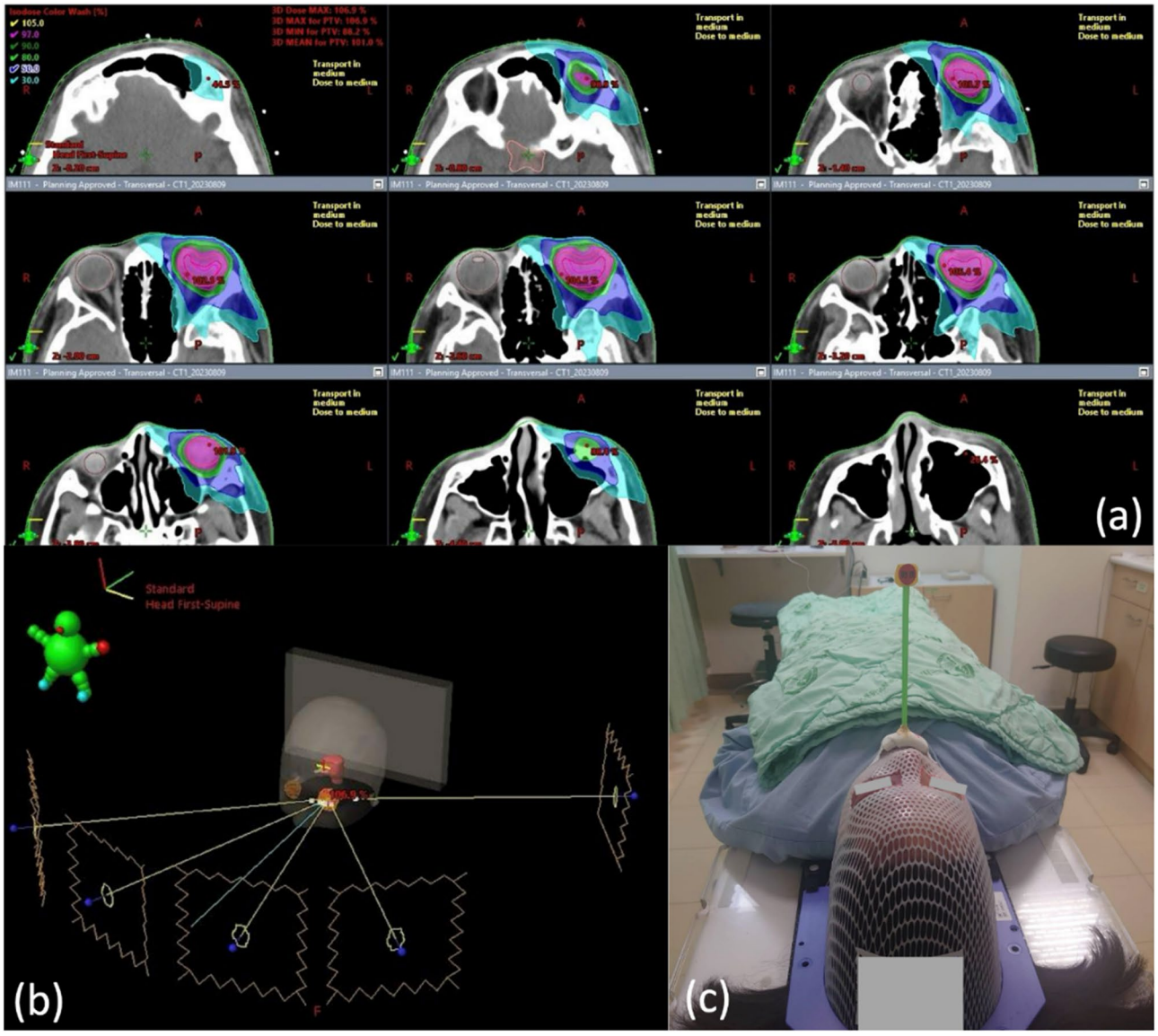

Fig. 3 External beam radiotherapy treatment plan. (**a**) Dosimetry (**b**) Portal design (**c**) head cast with a target placed anterior to the lips

Conclusion

EBRT is effective for treating diffuse choroidal hemangiomas in Sturge-Weber syndrome, offering significant tumor reduction and potential for complete remission with supplementary therapy. Early intervention remains key to optimizing outcomes.

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Author contributions
Conceptualization and Design: CN Chu and TW Su were responsible for conceptualizing the study and designing the research protocol. Data Collection and Analysis: IL Chou collected the clinical data and conducted the primary analysis. TW Su and CN Chu verified the data accuracy. Manuscript Drafting and Review: TW Su and CN Chu drafted the initial manuscript. IL Chou, HC Fan, and SN Chen provided critical revisions and approved the final version. Supervision and Oversight: SN Chen and CN Chu supervised the study design, data interpretation, and manuscript preparation. Guarantor: CN Chu serves as the guarantor, ensuring the integrity and accuracy of the entire work.

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Data availability
All data generated or analyzed during this study are included in this published article.

Declarations

Ethics approval and consent to participate
All methods were performed in accordance with the relevant guidelines and regulations. Informed consent was obtained from all subjects. Informed consent was obtained from all subjects. Ethical approval has been obtained from the ethics committee of China Medical University Hospital. (project ID number CMUH113-REC1–082).

Consent for publication
All authors confirm that they have reviewed the manuscript and consent to its publication.Written informed consent was obtained from the patient for publication of this case report.

Competing interests
The authors declare no competing interests.

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