CASE REPORT

Enhanced Visibility of the Retinal Nerve Fiber Layer Defects by the Underlying Diffuse Choroidal Angioma in Sturge Weber Syndrome with Glaucoma

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Abstract

Aim: To describe a unique finding in a patient with Sturge–Weber syndrome (SWS) accompanied by glaucoma.

Background: Cases with SWS may have an associated choroidal angioma and glaucoma. The localized retinal nerve fiber layer (RNFL) defects induced by the associated glaucoma are better detected against the red background of the associated choroidal angioma.

Case description: A 15-year-old boy was presented with left-side SWS. The case was subjected to full clinical examination, intraocular pressure (IOP) measurements, color fundus photography, ultrasonography, visual field assessment, and optical coherence tomography (OCT) of the peripapillary RNFL and ganglion cell layer (GCC) analysis. The main clinical finding was the enhanced detection of defects of the RNFL against the red color of associated choroidal angioma in this glaucomatous eye.

Conclusion: There is enhanced clinical visibility of RNFL defects in presence of associated diffuse choroidal angioma in Sturge–Weber glaucoma.

Clinical significance: Easier clinical detection of RNFL defects in SWS glaucoma associated with diffuse choroidal angioma.

Keywords: Case report, Choroidal angioma, Retinal nerve fiber layer defects, Secondary glaucoma, Sturge–Weber syndrome, Trans-scleral diode cyclophotocoagulation (DCPC).

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Background

The critical feature of glaucoma is an ongoing thinning of the RNFL as a result of retinal ganglion cell loss. Slit lamp biomicroscopy of the peripapillary region could allow clinical detection of the RNFL defects. Such findings are confirmed and quantified with the OCT. RNFL defects are recognized as dark bands radiating from the optic disc margin and wider than the superficial retinal vessels. Generalized thinning of the RNFL, in advanced glaucoma, are harder to detect compared to the localized defects which are easier to be assessed with red-free green photographs.[1] However, the clinical detection of RNFL defects is not always easy and requires training.

The present case report demonstrated a case with SWS associated with secondary glaucoma and a diffuse choroidal angioma, in which the RNFL defects were easily detected clinically. This fully documented case will be of interest to the readers due to its rarity and significance. The patient's family consent was obtained for publication, photo presentations, and subsequent surgical intervention. Personal identifying features are avoided in this report. The tenets of the Declaration of Helsinki were followed and the Ethical Committee of the Giza Eye Center approved the work.

Case Description

A 15-year-old boy was referred to with an uncontrolled IOP in the left (LT) eye. Irrelevant medical and family history and no past interventions.

Examination revealed features of unilateral SWS on the left side of the face. The skin showed capillary angiomas in the distribution of the ophthalmic division of the trigeminal nerve with dilated episcleral vessels on the surface of the LT eye (Fig. 1A) and gonioscopy revealed reddish coloration of the trabecular meshwork. The best corrected visual acuities (OU) were 1.0 *oculus uterque* [right (RT) eye: +0.25, −0.75 @5°, (LT) eye: +1.5, −0.25 @115°]. IOP was 18 mm Hg on the RT eye and 40 mm Hg in the LT eye using Goldmann applanation tonometry, under a combination of topical B-blockers and carbonic anhydrase inhibitors eye drop applied twice daily only to the LT eye. The slit lamp examination was normal. Optic nerve head examination showed thinning of the neuroretinal rim. B-scan ocular ultrasonography showed diffuse choroidal thickening in all quadrants (Fig. 1B).

Color fundus photography showed diffuse redness with enhanced visibility of the RNFL defects, being darker in color and easily distinguishable, from the surrounding retina (Fig. 2).

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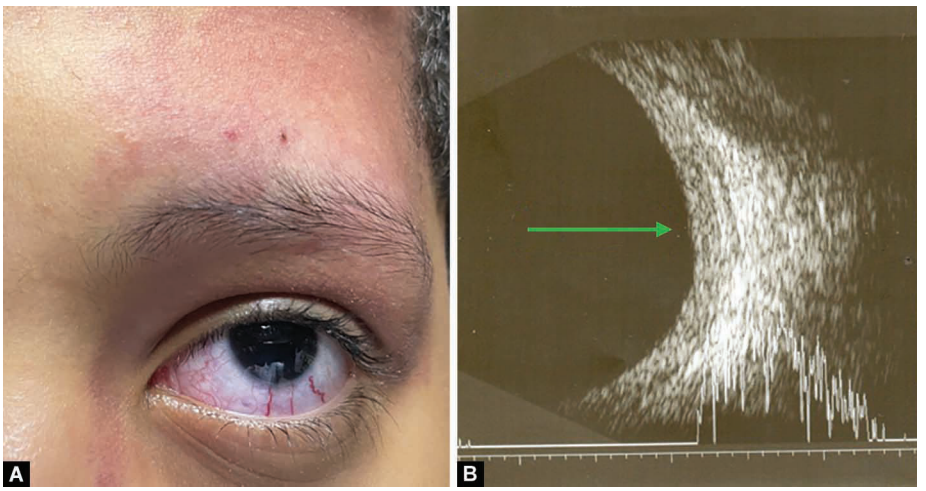
Patient consent statement: The author(s) have obtained written informed consent from the patient's parents/legal guardians for publication of the case report details and related images.:



Figs 1A and B: (A) Port wine stains affecting the skin of the forehead, upper and lower eyelids on the left side together with dilated episcleral vessels; (B) Showed the thickened choroid by the diffuse angioma (green arrow)

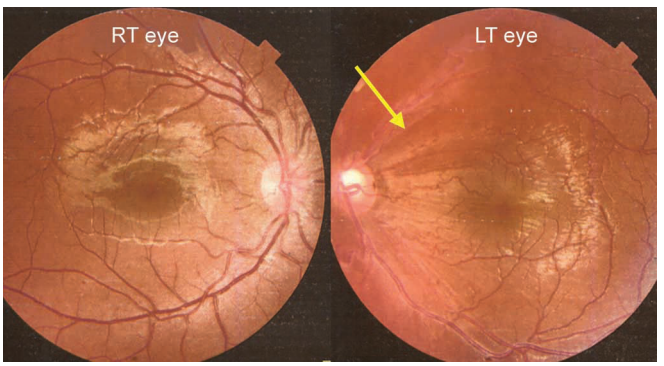


Fig. 2: Color fundus photography. RT fundus is normal. LT eye shows diffuse red color of the fundus, thinning of the neuro-retinal rim and obvious RNFL defects (yellow arrow)

Diagnostic Assessments

Optical coherence tomography (OCT) of the peripapillary RNFL [Optovue, Fremont, California (CA), US] showed thinning (average total is 94 µm, average superior 74 µm, and inferior 85 µm) together with a mild reduction of the GCC thickness in the macular region. The 30-2 visual field-testing showed moderate glaucomatous damage (MD −5.99, outside normal glaucoma hemifield testing with superior and inferior arcuate defects).

Therapeutic Intervention

The uncontrolled glaucoma was managed by trans-scleral diode cyclophotocoagulation. A total of 24 nonoverlapping spots were applied circumferentially sparing the 3 and 9 clocks' meridians. The laser parameters were 1500 MW power and 3 seconds. Laser treatment was done under general anesthesia using the fiber-optic Iris G-probe (OcuLight SLx, Iris Medical Instruments, Mountain View, CA, US).

Follow-up and Outcomes

At the 12-month visit, the IOP reached 20 mm Hg under a combination of topical β-blockers and carbonic anhydrase inhibitors eye drops applied twice daily only to the LT eye. At this point, a topical α-agonist is added and the patient is awaiting further evaluations and a possible laser redo (Fig. 3).

Discussion

Sturge–Weber syndrome (SWS) is a congenital hamartomatous vascular disorder involving the skin, eyes, and brain. The most common presentation is a facial cutaneous angioma involving parts of the distribution of the trigeminal nerve. Choroidal hemangioma on the same side (40%) and possibly IOP elevations and glaucoma (30%) also commonly occur.[2]

Glaucoma may be present in 30–70% of the cases, such incidence increases with the presence of capillary angiomas of the same side eyelid, episclera, and conjunctiva. Glaucoma is usually unilateral and often diagnosed early in life, but it can develop during adolescence or adulthood in 40% of cases. Bilateral facial hemangiomas could be associated with bilateral glaucomas.[3]

Postulated mechanisms for the development of glaucoma in those patients include—firstly, the existence of anterior chamber angle anomalies, secondly, the elevated episcleral venous pressure (EVP) induced by the episcleral angiomas.[4]

The incidence of diffuse choroidal angioma is 20–70% of patients with SWS, almost unilateral to the side of cutaneous angiomas. However, bilateral cases have been described.

Choroidal angioma may assist in the pathogenesis of glaucoma and increase its incidence. Diffuse choroidal angiomas are detected clinically as a bright red-orange color appearance of the fundus and confirmed with imaging.[5]

The present case demonstrated the features of SWS syndrome and is associated with diffuse choroidal angioma (as seen clinically and confirmed by ultrasonography) and secondary glaucoma mostly due to elevated EVP and the presence of diffuse choroidal angioma (the age of onset and presence of blood in Schlemm's canal). The presence of diffuse of choroidal angioma allowed for better detection of the RNFL defects induced by glaucoma; they appear darker and reddish in contrast to the hardly detected dark wedges induced by the RNFL defects in normal glaucoma subjects. In another ward, the diffuse choroidal angioma shows up more through the overlying areas of RNFL defects.

The diffuse choroidal angioma could also explain the hyperopic refraction in the LT eye due to axial length shortening.

The aim of the current case is to document the clinical findings rather than to focus on management.

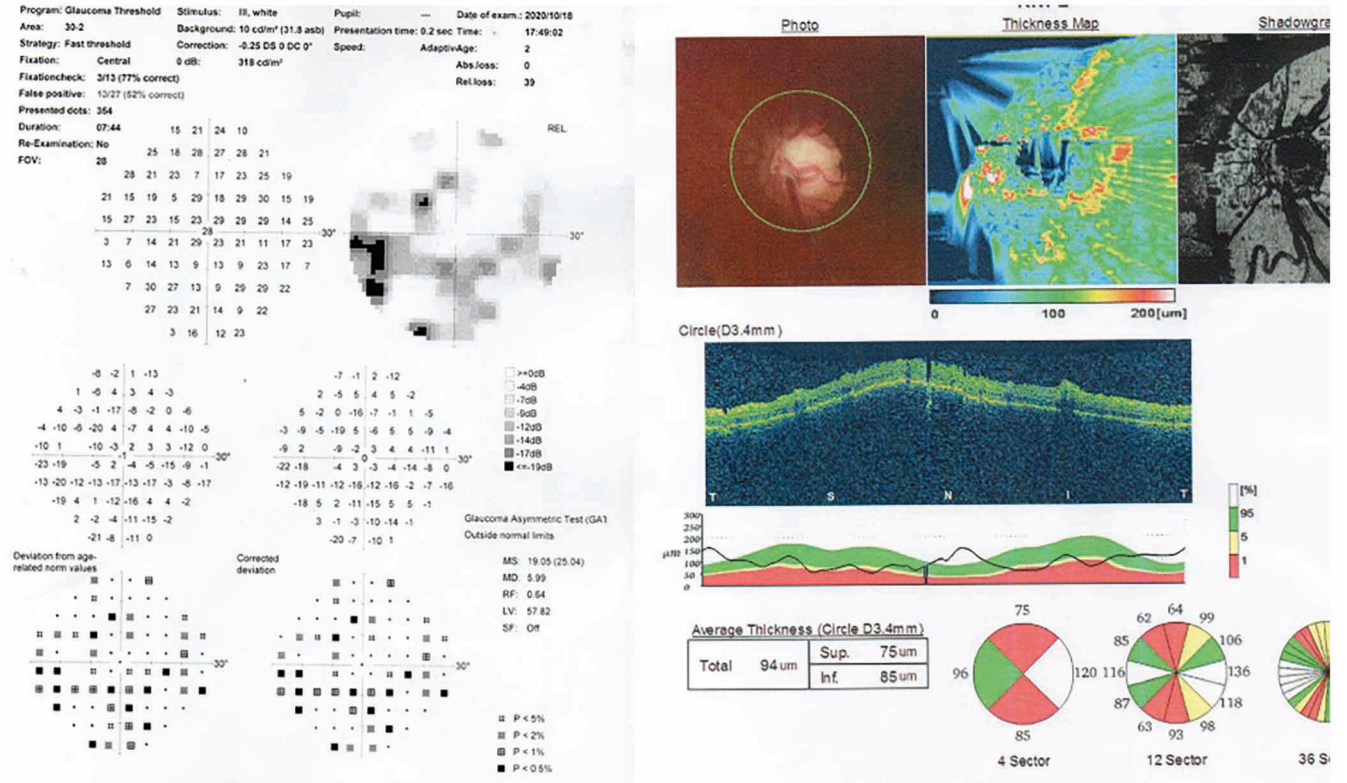


Fig. 3: The visual field and peripapillary GCC of the LT eye

To the best of the available knowledge, the finding reported in this manuscript is novel to the literature and hasn't been published before.

Conclusion

The presence of diffuse choroidal angioma in SWS may assist in the clinical glaucoma diagnosis by allowing better visibility of the RNFL defects induced by the associated glaucoma and *vice versa*, whereas the enhanced visibility of the RNFL defects alerts to the diagnosis of associated diffuse choroidal angioma in SWS cases.

Clinical Significance

Easier clinical detection of RNFL defects in SWS glaucoma associated with diffuse choroidal angioma. Such a fully documented case will be of interest to the readers due to its rarity and significance.

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