Case Report

Serous Retinal Detachment Resolution following Trabeculectomy in a Patient with Sturge–Weber Syndrome

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Abstract

Purpose: To report a case of Sturge–Weber syndrome (SWS) complicated with uncontrolled glaucoma and serous retinal detachment (SRD) in the left eye that evolved with complete resolution after trabeculectomy.

Methods: We report the case of a 10-year-old boy with SWS complicated with uncontrolled glaucoma and SRD in the left eye. In primary evaluation, he presented with a left-sided nevus flammeus affecting upper eyelid and best corrected visual acuity of 20/50 on the affected eye. Fundus examination revealed glaucomatous optic nerve neuropathy and diffuse choroidal hemangioma with overlying SRD, which were confirmed with spectral domain optical coherence tomography. Right eye was unremarkable.

Results: The patient underwent trabeculectomy with mitomycin-C on the affected eye. Two weeks later, he presented with normalization of the intraocular pressure and substantial resolution of subretinal fluid (SRF), which improved to complete resolution of the SRD at 2 months of follow-up. In addition, there was an improvement of visual acuity from 20/50 to 20/40.

Conclusion: This is the first report to describe a case of SWS associated with SRD and resolution of SRF after trabeculectomy.

Keywords: Glaucoma, Serous retinal detachment, Sturge–Weber syndrome, Trabeculectomy

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Introduction

Sturge–Weber syndrome (SWS) or encephalo-trigeminal hemangiomatosis is a relatively uncommon mesodermal phakomatosis, characterized by ipsilateral hemangioma involving many parts of the body, notably the ocular tissues. Choroidal hemangioma occurs in approximately 50% of patients with SWS and may undergo serous retinal detachment (SRD).[1] Further, glaucoma is the most common ocular complication in SWS, occurring in up to 71% of the cases.[2]

Detection and prompt treatment of SRD is crucial to avoid permanent visual loss. Although previous studies have described clinical improvement with different therapeutic strategies, such as photocoagulation, photodynamic therapy (PDT), radiotherapy, brachytherapy, oral propranolol, and bevacizumab,[3] there is no definitive treatment for SRD associated with diffuse choroidal hemangioma (DCH) in SWS.

In this case report, we describe a young patient with SRD secondary to DCH and SWS who evolved to complete remission of the subretinal fluid (SRF) hereinafter a successful trabeculectomy. Written informed consent was obtained from study participant, and research was conducted with the approval and ethical review.

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Case Report

A 10-year-old male patient with SWS, diagnosed at birth, was referred to our clinic due to uncontrolled intraocular pressure (IOP). He presented with a left-sided nevus flammeus affecting the ophthalmic and maxillary division of the trigeminal nerve, but no central nervous system involvement, as demonstrated by magnetic resonance imaging. Ophthalmic history included mild amblyopia in the left eye and ongoing treatment with topical dorzolamide hydrochloride 2%/timolol maleate 0.5%. The family denied any previous systemic treatment or surgical procedures.

On clinical examination, best corrected visual acuity (BCVA) was 20/20 in the right eye (−0.50 D Sph-0.50 D Cyl ×180) and 20/50 in the left (+2.00 D Sph-1.25 D Cyl ×170). Gonioscopy revealed open angles without abnormalities bilaterally. IOP was 16 mmHg in the right eye and 28 mmHg in the left eye. On dilated fundus examination, the left optic nerve had a 0.5 cup-to-disc ratio, with diffuse and localized neuroretinal rim loss, corroborated by spectral-domain optical coherence tomography (SD-OCT). Additionally, left eye presented DCH with the classic "tomato ketchup" appearance, overlying macular SRD. SD-OCT imaging of the left eye confirmed the presence of SRF [Figure 1a], and enhanced depth imaging demonstrated a thicker choroidal bed than the right eye. The patient underwent trabeculectomy with mitomycin-C (subconjunctival injection of 30 μg/0.1 ml) on the left eye, with 400 ml administration of mannitol at 20% concentration 30 min before the surgery.

Two weeks after surgery, one laser suture lysis (out of 5) was performed. At this visit, fundoscopic examination evinced resolution of the SRD while hypo-hyper pigmentation alterations, in a *leopard-spot* pattern, took place [Figure 2]. SD-OCT imaging demonstrated restoration of foveal depression and confirmed expressive resorption of the SRD with a small amount of remaining fluid. Foveal scan enabled the identification of hyperreflective foci alterations of the retinal pigment epithelium (RPE) – photoreceptor layer, associated with attenuation of the ellipsoid zone [Figure 1b].

At 2-month follow-up, BCVA of the left eye was 20/40 (-1.25 D Cyl ×170). The SD-OCT revealed a retina completely attached to the RPE, disappearance of hyperreflective foci alterations, and restoration of the ellipsoid zone [Figure 1c]. The patient was free of eye drops, and IOP values varied between 12 and 14 mmHg on several measurements.

Discussion

We were unable to find in the literature other studies reporting remission of the SRD after trabeculectomy. This can be considered a clinically relevant finding, one that precludes the patient from being submitted to additional procedures, which is especially important when considering infant patients. In other words, if trabeculectomy is needed due to glaucoma conditions in a patient with SRD associated with SWS, it may be worth doing that before using other alternatives treatments for SRD, such as PDT.

It is not completely clear how filtering surgery could have interfered favorably reducing SRF. We hypothesized two possible explanations. First, the increase in rate of aqueous humor filtration could have lowered levels of vascular endothelial growth factor (VEGF) on the eye, resulting in reduction of the inflammatory cascade, thereby reducing vascular leakage and enabling SRF absorption. This rationale is supported by previous work showing that injection of anti-VEGF was capable of reducing SRD associated with SWS and circumscribed choroidal hemangioma.[3]

Second, it is also possible that injection of mannitol preoperatively could have helped to establish fluid balance in retinal-choroidal system and acted as a trigger to achieve ocular homeostasis and maintenance of the retina attached.

Mannitol has a short-term effect, and even after 2 months of follow-up, SRF did not recur. Therefore, the use of mannitol

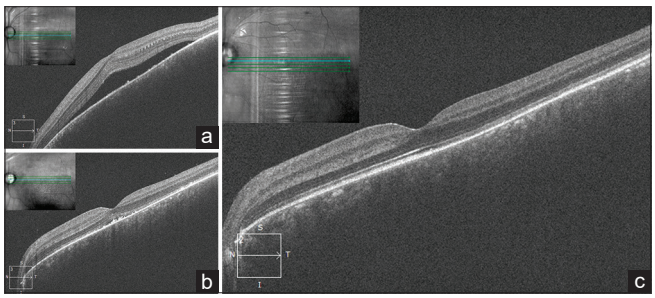

Figure 1: (a) Spectral-domain optical coherence tomography (SD-OCT), horizontally oriented, demonstrates a serous retinal detachment. (b) SD-OCT two weeks after the trabeculectomy procedure demonstrates expressive resorption of the serous retinal detachment with a small amount of remaining fluid associated with focal alterations of the retinal pigment epithelial – photoreceptor layer. (c) SD-OCT two months after the trabeculectomy procedure demonstrates a retina completely attached to the retinal pigment epithelium, disappearance of hyperreflective foci alterations, and restoration of the ellipsoid zone

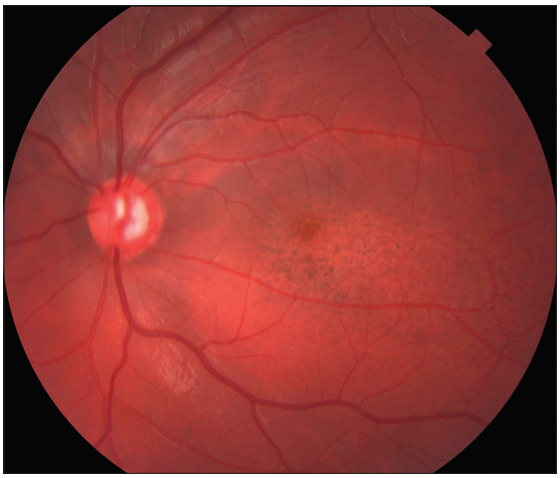

Figure 2: The left fundus image shows 0.5 cup-to-disc ratio, diffuse choroidal hemangioma, and hypo-hyper pigmentation of the foveal area

may have reduced the SRD allowing the pumping capacity of RPE to keep the retina attached.

Patients with DCH are at higher risk of developing spontaneous or drug-induced SRD, and this seems to be related to alterations in retina and choroid vessels. These patients tend to have thickened choroids, irregular choroidal vessels, and thinner outer retina.[4] Our patient presented choroidal thickness of 469 μm, which was thicker than the right eye (295 μm).

Changes on the external retina and RPE were also observed in this case after the resolution of the SRD. At 1-month postoperative, fundus examination revealed an interesting pattern on the posterior pole with hypo-hyper pigmentation of the foveal region. We also identified hyperreflective foci alterations of the RPE – photoreceptor layer on SD-OCT, which correlated with fundoscopy findings. Abdolrahimzadeh *et al*.[5] described this OCT finding as small white dot-shaped "micro-drusen-like" alterations of the retina.

We recognize that this is a single case and that other factors, such as the intrinsic fluctuation of the SRF, could have acted as confounders.[6] However, the large amount of reabsorbed fluid as well as the rapid and lasting resolution of the SRF following the trabeculectomy makes this fact less likely. Further, SRF has been associated with glaucoma. This finding can occur in patients with acquired optic pit due to the defect in lamina cribrosa, and the fluid is characteristically continuous to the optic nerve. Our patient did not present optic pit in retinography, angiography, nor OCT, and SRF was limited to the subfoveal region, not reaching the peripapillary region.[7,8]

When it comes to rare diseases, such as SWS, prospective studies can be time and resource consuming and, eventually, unfeasible. Since trabeculectomy achieved IOP control and possibly resolved the SRF, it could be considered the elected treatment for managing SRD in patients with clinically uncontrolled glaucoma secondary to SWS, before trying other forms of invasive therapies. Further studies should attempt to better investigate this relationship.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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