Lamellar Sclerectomy with Sclerostomy for Choroidal Hemangioma in Sturge-Weber Syndrome

Simanta Khadka¹, Raghunandan Byanju², Sangita Pradhan², Sabina Parajuli³

¹MD, Department of Vitreo-Retina, Nepal Eye Hospital, Kathmandu, Nepal. 
²MD, Department of Vitreo-Retina, Bharatpur Eye Hospital, Bharatpur, Chitwan, Nepal. 
³MD Resident, Bharatpur Eye Hospital, Bharatpur, Chitwan, Nepal.

INTRODUCTION

Sturge-Weber Syndrome (SWS) also known as encephalotrigeminal angiomatosis is a rare phakomatosis without any hereditary predisposition which may occur in a frequency of 1 per 50,000 live births.¹ SWS is characterized by a facial angioma, ipsilateral leptomeningeal hemangioma and ocular manifestations including hemangiomas of the conjunctiva, episclera, iris, ciliary body and choroid.² Mutation in the GNAQ gene which leads to stimulation of cell proliferation and inhibition of apoptosis is associated with SWS.³ Secondary glaucoma in SWS is a challenging entity due to its poor response to standard medical treatment. Several surgical approaches have been advocated, however long-term prognosis in terms of both IOP control and functional recovery remains unsatisfactory in these patients.⁴ Herein, we report a case of SWS with choroidal hemangioma and glaucoma not under control with medical management. The IOP was regulated following lamellar sclerectomy with sclerostomy but the visual recovery was not attained.

CASE DESCRIPTION

A 43-year-old male diagnosed with SWS (Figure 1.A) presented with diminution of vision of 6/36 and IOP of 28 mmHg in right eye (RE). His fellow eye was within normal limit with 6/6 vision and IOP of 16 mmHg. On examination of RE, episcleral hemangioma was present (Figure 1.B) and dilated fundus evaluation revealed saturated reddish orange hue suggestive of choroidal hemangioma and exudative detachment in the inferior and superior pole (Figure 1.C, D). Topical antiglaucoma fixed combination Gtt. Timolol maleate
0.5% + Brimonidine tartrate 0.2% BD was started and the IOP was 19 mmHg after a week. The patient lost follow-up for six months during the COVID pandemic. He then presented with severe visual impairment (perception of light) and massive exudative retinal detachment with 360° kissing choroidal which was obvious just posterior to the lens (Figure 2.A, B). IOP in the affected side was 38 mmHg. Maximum tolerated antiglaucoma medications (oral acetazolamide + Gtt. Timolol maleate 0.5% + Brimonidine tartrate 0.2% + Gtt. Latanoprost 0.005%) were used but the IOP was not brought under control. He was then scheduled for four quadrant penetrating sclerectomy with sclerostomy.

SURGICAL TECHNIQUE

The surgery was performed under peribulbar anesthesia with strict aseptic precaution. A 360° conjunctival peritomy was performed and the underlying tenon’s capsule was dissected. Four bridle sutures were placed underneath the recti muscles. The exposed scleral bed was carefully cauterized over the episcleral hemangioma to prepare an area for scleral dissection (Figure 2.C). A 4*6 mm, 2/3rd partial thickness scleral flap was created approximately 3 mm posterior to the insertion of rectus, 2 mm behind the equatorial region. The scleral flap was excised and a 2*1 mm sclerostomy site was created till the underlying choroidal tissue was visible (Figure 2.D). No attempt was made to drain the subretinal fluid. The sclerectomy and sclerostomy area was left bare. The overlying tenon’s and conjunctiva was closed with absorbable 8-0 vicryl. Similar procedure was repeated in the remaining three quadrants. No any intra-operative complications were encountered during the procedure. Post-operatively oral anti-microbial agent (tab. Ciprofloxacin 500 mg BD), oral steroid 1mg/kg body weight, oral acetazolamide, oral proton pump inhibitors and oral analgesics in as per need basis was continued for the next five days. Oral steroid was tapered off in a weekly interval. Topical antibiotics, steroid drops in a tapering fashion, cycloplegic drops and steroid antibiotic combination ointment were administered from the first postoperative day.

The recovery period was uneventful. Six months’ post-surgery at the final follow-up visit, the IOP was stable at 16 mmHg and the exudative fluid was gradually declining (Figure 2.E) but the visual acuity remained unchanged with perception of light. He then lost follow-up.
DISCUSSION

The prevalence of glaucoma in SWS can be estimated to range from 30% to 70%. The choroidal hemangioma which is one of the ocular manifestations is frequently present on the same side as facial nevus flammeus. Glaucoma is also frequent in the affected side. The mechanism for glaucoma in SWS can be explained by several theories. Anterior segment developmental anomalies and elevated episcleral venous pressure may cause increased IOP in infants. Changes in ocular hemodynamics can also be attributed to glaucoma in SWS. Transudates from the thin-walled choroidal vessels also lead to a hypersecretion. The finding of a normal aqueous outflow facility in many cases is consistent with hypersecretion as a cause of IOP in SWS. Though these choroidal hemangiomas are congenital, exudative retinal detachment may occur at any age and observed in approximately half of patients with SWS.

The management of glaucoma in SWS is still debatable. Medical treatments in SWS patients may not ensure a good long-term control of glaucoma, hence a surgical approach is mandatory in these type of cases. The surgical approach may be tailored according to the age at presentation. Goniotomy and trabeculotomy represent the most appropriate surgical procedures to overcome the malformed anterior chamber angle under 4 years of age. Similarly, trabeculotomy–trabeculectomy have been suggested as the viable alternative in infants and children. In adults, filtering surgery like trabeculectomy is most commonly performed. However, filtering procedures have been associated with severe complications such as expulsive choroidal hemorrhage, bleeding, prolonged hypotony and high risk of bleb failure. Hence we avoided this procedure in our case and also the pathogenesis for glaucoma is different in our context.

Diffuse choroidal hemangioma is benign vascular tumor and B-scan ultrasonography can also be utilized to confirm the diagnosis. Nonetheless, magnetic resonance imaging is deferred in our case to demonstrate diffuse choroidal hemangioma and also to rule out leptomeningeal hemangioma because of the financial constraints of the patient. Exudative retinal detachment associated with SWS is caused by accumulation of serous fluid beneath the neurosensory layers. This condition is not only difficult to manage but often follows a relapsing course. The glaucoma in our case is secondary to massive exudation due to diffuse choroidal hemangioma which caused the forward propulsion of the lens-iris diaphragm. There are different proposed treatment options for symptomatic serous retinal detachment associated with choroidal hemangiomas including photodynamic therapy, plaque brachytherapy, external beam and proton beam radiation and stereotactic radiosurgery. Nonetheless due low resource setup, we could not apply these methods.

Low dose ocular irradiation with external beam radiation therapy (ERBT) had shown a good result to decrease the tumor thickness, reattach the retina, and stabilize the visual acuity. ERBT is deemed most efficient in these type of cases with limited adverse effects of cataract, radiation retinopathy, keratitis and soft tissue burn. However, due to unavailability of the services in this region and refusal of the patient for referral to other center we were left to decide upon the management based on our expertise and available resources.

We managed our case with partial thickness sclerectomy combined with sclerostomy after discussing the prognosis and possible complications. We preferred this technique because of our previous experience with management of uveal effusion syndrome. The advantage of bypassing the penetration into the anterior chamber is prevention of sudden intraoperative and postoperative hypotony which could prevent potential sight-threatening complications of choroidal effusion and hemorrhage. Moreover, the sclerectomy facilitates the drainage of uveal exudation in the immediate postoperative period and it also decompress vortex veins indirectly by relaxing the scleral tension. The subconjunctival filtering bleb following sclerectomy is more diffuse and flatter than trabeculectomy which facilitates
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REFERENCES


gradual fluid resorption. This procedure also bypasses the episcleral venous pressure, the main factor implicated in the pathogenesis of glaucoma in SWS occurring in childhood or adulthood. Sclerostomy is also feared with complications in this type of condition. We preferred this technique to facilitate the prolonged drainage of the exudative fluid. Similarly, non-drainage of the fluid by penetration of the uveal tissue is preferred to prevent the bleeding. Complications reported with this procedure are intraoperative hemorrhage, hypotony, hyphema, vitreous hemorrhage, rhegmatogenous retinal detachment and uveal prolapse. Episceral hemangioma could significantly hinder the dissection of scleral flap. This factor should be considered preoperatively before making the sclerectomy. Available literature supports the outcome of sclerectomy in significantly reducing the IOP. The damage to the foveal photoreceptor layer is associated with the duration of detachment. Degenerative foveal changes, secondary fibrous metaplasia, and cystoid macular edema lead to irreversible functional loss as a consequence of persisting exudation. Hence functional outcome may not be achieved in chronic cases.

A relative short follow-up period precludes us from making a definitive statement about the efficacy of this procedure. However, we believe that this could be an effective surgical option for the management of clinically significant exudative retinal detachment associated with diffuse choroidal hemangioma in SWS especially in lower resource setup.

ACKNOWLEDGEMENT

We would like to thank the patient for giving us consent to publish the case report and accompanying images.

ETHICAL CONSIDERATION

Written and informed consent has been obtained from the patient for the publication of the case details and accompanying images. The identity of the patient has been anonymized throughout the text. Institutional approval was not required to publish case details.
CASE REPORT

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