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A Case Report of Late-Onset Glaucoma in Sturge-Weber Syndrome

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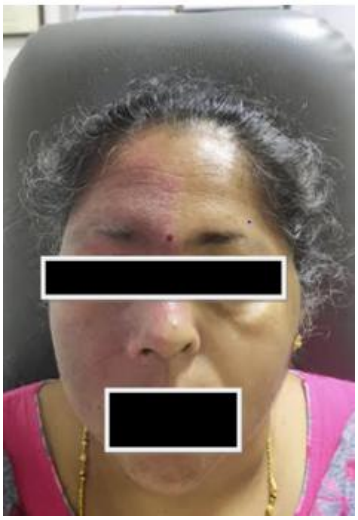
I. Introduction:

Sturge-Weber syndrome has been included in the group of phakomatoses that is characterized by hamartomas involving the brain, skin, and eyes. The characteristic facial port-wine stain involving the first branch of trigeminal nerve and the embryonic vasculature distribution in this area, leads to several ocular complications of anterior segment and can involve the eyelids and conjunctiva. The posterior segment of the eye is also affected with prevalence rate ranging from 30%-70% [1]. Glaucoma is related to anterior chamber malformations, high episcleral venous pressure, and changes in ocular hemodynamics. Glaucoma can be diagnosed at birth, but the disease can develop during childhood and in adults. The management of glaucoma is particularly challenging because of early onset, frequently associated with severe visual field impairment at the time of diagnosis, and unresponsive to standard treatment. Several surgical approaches have been proposed, but long-term prognosis for both intraocular control and visual functions remains unsatisfactory in these patients. Choriodal hemangiomas may also lead to visual impairment thorough exudative retinal detachment and macular edema[2,3].

II. Case-Report

A 53 year female, visited glaucoma clinic on 08.02.2021 with complaints of headache and vomiting since 10 days. Patient has been treated for the same in an eye clinic near her home town and was referred for further management. Patient gave history of anti-glaucoma medications prescribed from the referred doctor. There was no history in decrease in vision in both eyes. Patient was a known case of hypertension since 3 years and had been treated for the same with negative history of epilepsy or other neurological problems. Patient revealed that there is pinkish stain over right side of face since birth.

Patient BCVA was 6/6P with N6 in OD and 6/6 with N6 in OS. There was port-wine stain over the right side of the face. On GAT on the day of presentation IOP was 22 mm of Hg and 16 mm of Hg in RE and LE respectively. Anterior segment examination revealed corkscrew vessels and ill-sustained pupil in RE and within normal limits in LE. Gonioscopy showed open-angle graded according to Shaffer's grading(Grade 4). On fundus examination the CDR in RE was 0.6 with inferior rim thinning, elevated choriodal vessels nasal to disc and in LE the CDR was 0.5-0.6 with healthy neuroretinal rim.



The port wine stain on the right side of the patient's face involved his upper and lower eyelids and cheek.

Patient was investigated including MRI Brain, Standard Automated Perimetry, Pachymetry and OCT for assessing RNFL Thickness and fovea. Perimetry revealed superior arcuate scotoma in RE and early biarcuate scotoma in LE. OCT showed bipolar RNFLD in RE with early IT RNFLD IN LE. Pachymetry was WNL.

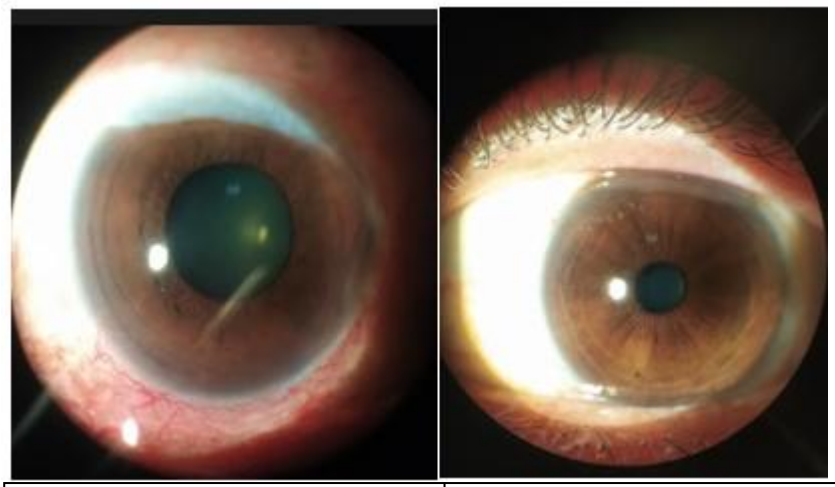
Patient was started on combination of Carbonic Anhydrase Inhibitors and Adrenergic agonist (Brinzolomide and Brimonidine) with Prostaglandin analogue (Bimatoprost) with continuation of antihypertensive medications. Patient was asked to review with MRI reports and for IOP check-up after 3 weeks.

On review after 3 weeks the pressure in RE was 40 mm of HG and in LE WAS 20 mm of HG, prostaglandin was discontinued with addition of beta-blocker (Timolol) with oral CAIS for a period of 10 days and decide to start oral propranolol for choriodal lesion

Further on follow-up for RE choriodal hemangiomas physician opinion was sought to start oral beta-blockers.

On 21.07.2021 after three months the IOP was still high in RE about 26/24 mm of Hg with no significant changes expect with decrease in choriodal elevation noted on RE fundus examination with oral propranolol. On considering persistent elevation of IOP in RE patient was advised RE Trabeculectomy with MMC under guarded visual prognosis explaining pros and cos of surgery with requirement of routine follow-up

Patient was operated on 31.07.2021 with proper surgical protocol with proper measure taken to prevent choriodal effusion during surgery including iv mannitol 100 ml infusion before surgery and patient tolerated the procedure well. On post-op examination, there was diffuse bleb formed and was discharged with proper post medications and with oral Carbonic Anhydrase inhibitors, antiglaucoma medications.



Right eye of the patient with RAPD with dilated episcleral vessels , and elevated bleb .Left eye was normal

RIGHT EYE	LEFT EYE

After 15 days patient visited glaucoma clinic, the bleb in right eye was early cystic (H2E2V0) formed and IOP was controlled .

Patient was asked to follow-up after 2 month was advised to continue oral propronolol and decide to stop topical AGM next visit.

III. Discussion:

Sturge-Weber Syndrome, a phakomatosis that has no known hereditary pattern. Individuals with this congenital anomaly have angiomatous involvement of the meninges, and brain, which causes seizures, mental retardation and cerebrocortical atrophy. Glaucoma has been reported between 30%-70%. Although facial angioma is unilateral, between 10%-30% are bilateral. Patients with upper-lid involvement are more common likely to develop glaucoma. Vascular malformation of the conjunctiva, episclera, choroid, and retina are also found[4,5].

In 1973, Weiss proposed two mechanism of glaucoma in SWS. In congenital glaucoma, increased episcleral venous pressure from episclera angiomas combined with abnormal angle development leads to development of the glaucoma[2]. In later onset with normal appearing angle glaucoma may occur due to elevated episcleral venous pressure which is seen majority of patients.

Propranolol was discovered to induce involution of cutaneous, orbital and ocular hemangiomas. Mechanisms include an early effect through vasoconstriction from reduced nitric oxide release; an intermediate effect by down regulation of VEGF, and matrix metalloproteinase and late effect through apoptosis of proliferating endothelium. Oral propranolol seems to be an effective method to minimize the development of sight-threatening choriodal effusion after glaucoma surgery[4,5,7].

Regardless of the mechanism of glaucoma, controlling of IOP depends on the patient presentation. In later onset glaucoma, topical medications are often first line of treatment, but frequently fail eventually to control IOP. Limited success with laser trabeculoplasty has been reported. Trabeculectomy with MMC and Aqueous drainage devices has been successful in controlling IOP.

Filtering surgery on patients with SWS carries the risk of massive intaroperative choriodal effusion or suprachoriodal hemorrhage[5,6].

IV. Conclusion:

Glaucoma is the most common ocular complication in SWS. The most frequent is congenital glaucoma, but it can occur in children and adults, making careful ophthalmic follow-up.

Glaucoma management is challenging in SWS due to its early development and poor response to standard medications. Surgery is frequent to obtain long-term control of IOP in order to avoid visual function loss. To the date, despite the wide range of available medical and surgical approaches to treat both the congenital and late-onset form of glaucoma, the development of this ocular complication still represents the worst prognostic factor for vision loss in SWS patient. As with all glaucoma, early detection and treatment are important for the patient's useful vision.

DISCLOSURE:

The authors report no conflict of interest in this work.

References:

[1]. Kainat Akhter, BA and Sarwat Salim, MD .Sturge –weber syndrome and secondary glaucoma. Ophthalmic Pearls. Eyenet
[2]. Weiss DI. Trans Ophthalmology.2000:107(11):2105-2110
[3]. Mandal AK ophthalmology. 1999;106(8):1621-1627.
[4]. Rodolleda G Ophthalmology. 200;108(12);2152-2153
[5]. Flavisco Mantelli, Alice Bruscolini. Ocular manifestation if Sturge-Weber Syndrome: pathaogensis, diagnosis, and manangement. Clinical ophthalmology 2016:10.871-878.
[6]. Basela E. Sturge-Weber syndrome. Semin Cutan Med Surg.2004;23:87-98.
[7]. S Kaushik, P Kataria, G Joshi. Perioperative Propranolol: a useful adjunctive for glaucoma surgery in Sturge-Weber syndrome

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