

Surgical Management of Glaucoma in Sturge-Weber Syndrome

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Introduction

Sturge-Weber syndrome (SWS) or encephalo-trigeminal hemangiomas is a sporadic, mesodermal phakomatosis characterised by hemangiomas involving many parts of the body, particularly ocular, intracranial and facial cutaneous structures [1-3].

Glaucoma is a common feature, with an incidence of 30%-71% in patients with Sturge-Weber syndrome [4-6]. Many mechanisms of raised intraocular pressure have been described in the past, the most consistent being congenital trabeculodysgenesis, increased episcleral venous pressure and hypersecretion due to ciliary body angioma [7,8].

An increased risk of intra and post-operative complications has been noted with glaucoma filtering procedures in these patients, predominantly due to rupture of the fragile vasculature in the choroidal hemangiomas, leading to expulsive choroidal haemorrhage or exudative choroidal detachment (CD) caused by sudden decompression during or after filtering procedures [9-13]. Prophylactic sclerotomies have been advocated, to be performed prior to ocular decompression, during filtering procedures in order to avoid these complications. The necessity of prophylactic procedures has been questioned. Eibschitz-Tsimhoni et al.[4], in a retrospective study, have reported that none of their 17 patients with SWS who underwent glaucoma filtering surgery without prophylactic posterior sclerotomy developed intraoperative suprachoroidal haemorrhage or choroidal effusion requiring therapeutic intervention [4].

The aim of our study was to evaluate the surgical outcomes in terms of intraocular pressure control, maintenance of visual acuity and complications of glaucoma surgeries, in eyes with Sturge-Weber syndrome and to evaluate risk factors leading to surgical failure.

Methods

Institutional review board and ethics committee clearance for analysis of charts was obtained before commencing the study. A retrospective chart review of all consecutive cases diagnosed with Sturge-Weber syndrome, who underwent glaucoma surgery from January 2003 to December 2013 was performed. The diagnosis of SWS was made clinically on the basis of typical ocular and systemic features. Patients without a minimum follow-up of 2 months were excluded.

The data obtained included age, gender, ocular and systemic features of SWS, visual acuity, baseline intra ocular pressure (IOP), Goldmann applanation tonometry, gonioscopy (Posner four mirror, Ocular instruments.inc. Bellevue, WA), ultra sound pachymetry (Pachette 2, DGH tech. Inc. Shermans Dale, PA), details of medical treatment for glaucoma as well as details of the glaucoma surgery. Post-operative data collected included visual acuity, IOP, number of IOP lowering medications and details of post-operative complications,

including their management at post-op 1 week, 6 weeks and final follow up visit.

Definitions

Glaucoma was defined as the presence of characteristic optic nerve head and/or visual field changes. Optic nerve head features included increased vertical cup: disc ratio, cup asymmetry of more than 0.2, cup: disc ratio of >0.7 associated with focal or diffuse thinning of the neuro-retinal rim and/or focal or diffuse loss of retinal nerve fibre layer seen as wedge defect. Glaucomatous visual field changes were defined by Anderson's criteria as presence of at least 3 contiguous, non-edge points depressed to p<5% and atleast 1 point depressed to p<1%, glaucoma hemi-field test 'outside normal limits' and pattern standard deviation depressed p<5% on standard automated perimetry.

Success and failure

Complete surgical success was defined as IOP ≥ 6 and <18 mm Hg without the need of IOP lowering medications at final follow-up. If additional ocular hypotensive drugs were required to keep the IOP within this range, it was termed as qualified success. Surgical failure included eyes with post-operative IOP of less than 6 mm Hg (defined as hypotony) or more than 18 mm Hg, need for re-surgery (excluding bleb manipulation like removal of sutures, massage or needling), loss of light perception and post-op complications such as persistent exudative choroidal detachment causing significant loss of visual acuity (>2 snellen's lines) at final visit, retinal detachment (at final visit) or phthisis bulbi.

Surgical procedures

Outcomes of primary surgery only were evaluated. Trabeculectomy with Mitomycin C (MMC, 0.4 mg/ml, 1 minute exposure) was the most common surgery performed (18, 66.7%), followed by trabeculotomy with trabeculectomy with MMC (0.4 mg/ml, 1 minute exposure) (4 eyes, 14.8%). Three eyes (11.1%) underwent trabeculotomy with trabeculectomy without use of antimetabolites. Trabeculectomy without use of antimetabolites and transcleral cyclophotocoagulation was required in 1 eye each (3.7%).

Statistical methods

Statistical package for the social sciences (SPSS) version 14: SPSS Inc. Chicago: released 2007, was used for the analysis. The Kolmogorov-Smirnov test was applied to test for normal distribution of data. Comparison of continuous data was done with the independent-samples Student's t-test for normally distributed data. The Mann-Whitney test and the Wilcoxon signed-rank test were applied to the non-normally distributed data. Categorical data were analysed with the Chi-square test and Fisher's exact test. The Snellen's chart was used

to record visual acuity and the values were converted to LogMAR (logarithm of the reciprocal of the minimum angle of resolution) scale for the purpose of statistical analysis. A change of 2 Snellen's lines from the pre-operative best corrected visual acuity (BCVA) was considered a significant change. A p-value less than 0.05 was considered statistically significant in the study. Logistics regression analysis was performed to evaluate specific risk factors for developing post-op exudative choroidal detachment. The variables that were evaluated included age, sex ratio, laterality, pre-operative BCVA, baseline IOP, presence of choroidal hemangioma, pre-operative number of IOP lowering medications, and whether prophylactic sclerotomy was performed or not.

Results

Twenty seven eyes of 25 patients diagnosed with Sturge-Weber syndrome underwent glaucoma surgery in our series. The median follow-up was 20 months (range: 2-122 months). The majority of the patients were in the paediatric age group. Mean age at presentation was 10.1 years + 10.3 years (range 1 month to 48 years).

Port wine staining in the distribution of the Trigeminal nerve was present in all patients (100%). Neurological features like convulsions were seen in 5 patients (20%) and radiological evidence (MRI/CT scan) of intracranial vascular abnormality was noted in 4 patients (16%). Twelve patients had lid hemangioma (44.4%) and episcleral hemangioma was present in 9 eyes (33.3%). Twenty one eyes (77.8%) also showed evidence of choroidal hemangioma on fundus examination.

Intraocular pressure and glaucoma

Glaucoma was bilateral in 6 patients (24%). All 27 eyes (100%) had open angles on gonioscopy. Initial intra ocular pressure (IOP) was 34 + 8.2 mm Hg ranging from 15 to 50 mm Hg. At final visit the mean IOP decreased to 15.4 + 7.9 mm Hg (p<0.001). The mean number of IOP lowering medications used pre operatively was 1.9 + 1.4. This decreased to 0.4 + 0.8 at the final visit (p<0.001).

Visual acuity

Seven eyes (25.9%) showed improvement in visual acuity, seven eyes (25.9%) showed worsening and 5 eyes (18.5%) maintained their visual acuity during the follow-up period. Mean best corrected visual acuity (LogMAR) at initial visit was 0.7 + 0.8 and at final follow-up was 0.8 + 0.9 but this difference was not significant (p=0.905). Visual acuity of 4 patients in the pre-verbal group could not be measured.

Surgical outcomes

Intraoperative expulsive choroidal haemorrhage was not encountered during any surgery in our series. Out of the 27 eyes that underwent anti glaucoma surgery, complete success was achieved in 17 eyes (62.9%) and qualified success in 1 eye (3.7%). The reasons for surgical failure included uncontrolled IOP (4 eyes), post-operative retinal detachment (3 eyes), persistent choroidal detachment leading to significant visual acuity loss (>2 Snellen's line) (2 eyes) and ocular hypotony (1 eye). The eye that had hypotony also developed choroidal detachment. One eye had both choroidal detachment and uncontrolled IOP due to secondary angle closure. Kalpan Meier survival analysis (Figure 1) showed a cumulative proportion surviving at the end of 1, 2, 3 and 4 years as 83%, 83%, 74% and 74%, respectively.

Age (Years)	Mean+SD	10.1+10.3
	Range	0.08-48
Sex ratio	(Male:Female)	14:13
Follow up (Months)	Mean+SD	37.9+37.1
	Range	2-122
Duration between presentation and surgery (days)	Mean+SD	132.7+282.6
	Range	1-1122
Systemic findings	Nevus Flammeus	27
	Neurological features	5
	Intracranial vascular abnormality	4
Laterality	Right eye	16
	Left Eye	11
Anterior segment	Lid hemangioma	12
	Episcleral hemangioma	9
	Tortous & dilated episcleral vessels	13
	Iris hemangioma	0
Pachymetry (microns)	Mean+SD	599.9+120.6
	Range	483-943
Glaucoma	Unilateral:bilateral	19:6
	Open angle	27
	Visibility of blood in Sclerotic canal	21
Fundus	Choroidal hemangioma	21
Initial visual acuity (LogMAR)	Mean+SD	0.7+0.8
	Range	-0.1 to 2.5
Baseline intraocular pressure (mm Hg)	Mean+SD	34+8.2
	Range	15-50
Pre-op no. of glaucoma medications	Mean+SD	1.9+1.4
	Range	0-4

Table 1: Demographics and clinical features.

On performing logistics regression analysis to evaluate risk factors for surgical failure, no statistically significant difference was observed between success and failure groups in terms of age (p=0.002), gender (p=0.586), laterality (p=0.782), BCVA (p=0.087), pre-op number of AGMs (p=0.840), presence of choroidal hemangioma (p=0.326), and prophylactic sclerotomy (p=0.326). However, significantly higher baseline IOP (p=0.037) was noted in the failure group as compared to the success group.

Choroidal detachment

Six eyes (22.2%) developed exudative choroidal detachment (CD) after the surgery that persisted beyond 2 weeks. In the eyes that developed CD the baseline IOP was significantly higher when compared to the eyes that did not develop CD ($p=0.026$). None of the eyes in this series had undergone pre-operative medical, laser, radiation or surgical treatment for choroidal hemangioma. A greater proportion eyes that developed post-op CD had choroidal hemangioma (83.3%) as compared to the eyes that did not develop CD (76.2%). However, this difference was not statistically significant ($p=0.146$). Similarly, there was no significant difference between the two groups in terms of age ($p=0.805$), gender ($p=0.918$), visual acuity ($p=0.715$), pre-op number of AGM ($p=0.939$) or prophylactic sclerotomy ($p=0.712$).

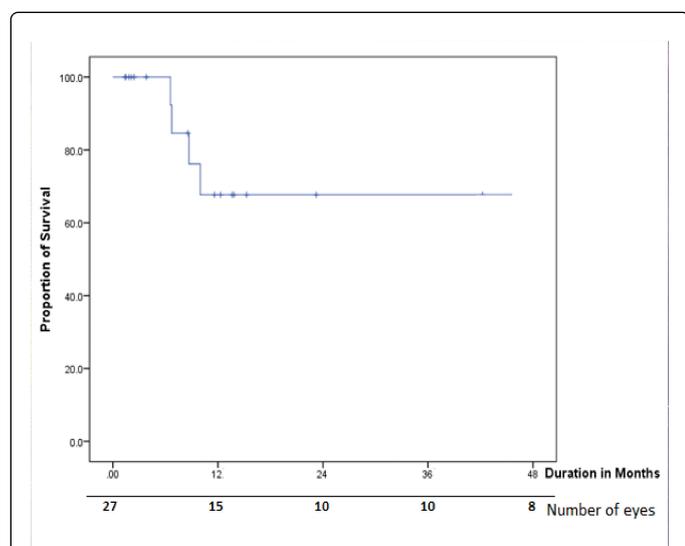


Figure 1: Kaplan-Meier survival graph for surgical success in eyes with glaucoma secondary to Sturge-Weber disease ($n=27$). Survival was defined as no surgical failure. The number of eyes at each follow-up interval is depicted in the table.

Discussion

Glaucoma is the most common cause of visual loss in patients with Sturge-Weber syndrome [5]. Various mechanisms have been proposed in the past with trabeculo-dysgenesis being responsible in the early onset glaucoma and increased episcleral venous pressure contributing to the raised intraocular pressure later on [7].

The onset of glaucoma at birth or during infancy is a common feature [5]. This was also evident in our study with 85.2% of case belonging to the pediatric age group. Most of the glaucoma in SWS presents unilaterally, though bilateral glaucoma has also been reported [6,13]. In our series, 76% patients had unilateral glaucoma.

Visibility of blood in the Schlemm's canal was seen in 21 eyes (77.8%). However, only 13 (48.1%) eyes had dilated and tortuous episcleral vessels, suggesting that raised episcleral pressures may be present (and contribute to glaucoma) even without the presence of dilated episcleral vessels evidenced on slit-lamp biomicroscopic examination. A study by Shiao T, et al. showed 3 eyes (60%) had increased episcleral venous pressure without increase in intraocular

pressure or dilated episcleral vessels [5]. Their study also had 2 eyes (66%) with glaucoma and increased episcleral venous pressure but no dilated episcleral vessels. Hence, gonioscopy should be performed in all cases of childhood glaucoma as it might be crucial in determining the mechanism of raised IOP particularly in cases where other features of SWS like facial hemangiomas are subtle or absent.

Trabeculectomy was the most common surgery performed ($n=19$, 70.4%) followed by combined trabeculotomy with trabeculectomy (25.9%). Phelps proposed that as the site of maximum resistance to the aqueous outflow in eyes with hemangiomas would be distal to the trabecular meshwork; angle surgeries like goniotomy or trabeculectomy alone would not be enough to lower the IOP [8]. Surgical success in childhood glaucoma is quite variable and varies with the type of surgery and associated anterior segment abnormalities. Success rates reported in the literature were lower in patients with developmental glaucoma associated with syndromes such as aniridia and SWS. Our series had an overall success rate of 66.7%. This was comparable with previous studies by Agarwal HC et al. (61.1%) [16]. Combined trabeculotomy with trabeculectomy (85.7%) had a better success rate than trabeculectomy alone (63.2%) in our series.

On performing a logistic regression to analyse the risk factors for surgical failure and development of post-operative exudative choroidal detachment, baseline IOP was found to be a statistically significant risk factor for both surgical failure and development of choroidal detachment. This indicates that sudden decompression of the eye could be a major factor leading to the development of choroidal detachment and failure of filtering procedures.

There are very few reports of intraoperative expulsive choroidal haemorrhage in eyes with SWS undergoing glaucoma surgery [9]. None of the eyes in our series developed choroidal haemorrhage, although choroidal hemangioma was present in 21 eyes (77.8%) including 3 eyes in which prophylactic sclerotomy was not done. Eibschitz-Tsimhoni, et al. also demonstrated that none of their 17 eyes with SWS or Klippel-Trenaunay Weber syndrome developed supra choroidal haemorrhage in spite of no pre/intra-op prophylactic measures (like sclerotomy) being undertaken [14,15]. They also debated the relevance of posterior sclerotomy in preventing post-op exudative choroidal detachment though 6 patients developed CD in their series. None of them required any therapeutic intervention. Another study of 19 eyes reported an incidence of post-operative CD in 16.7% eyes [16]. In our series, out of the 21 eyes in which posterior sclerotomy was performed 5 (23.8%) developed post-op CD, while out of the 6 eyes in which posterior sclerotomy was not done, 1 eye (16.7%) developed CD. This difference was not significant statistically.

Limitations

Limitations common to retrospective analysis was also present in our study. The utility of a prophylactic sclerotomy cannot be addressed without performing a randomized controlled trial, which could be difficult due to the rarity of the disease. The sample size in our study was not adequate to correctly compare the surgical outcomes and post-operative follow up in few eyes was not long enough to comment on the success of filtering procedure. However, our sample size was fairly large when considering the rarity of this disorder and the success rates of our surgical procedures compare well with those reported in the literature.

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