

CASE REPORT

Surgical Treatment of Children with Raised Intra Ocular Pressures Associated with Sturge Weber Syndrome at Hospital Kuala Lumpur, Malaysia

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INTRODUCTION

Sturge Weber Syndrome is a congenital neuro-oculo-cutaneous disorder characterized by facial capillary hemangioma (port wine stain), abnormal blood vessels of the brain (leptomeningeal angioma) and/or abnormal blood vessels in the eye¹. Children with Sturge Weber Syndrome frequently develop seizures, focal neurological impairment, visual problems and cognitive deficits. Port wine stain, also known as nevus flammeus is typical along the ophthalmic branch of the trigeminal nerve².

The most common ocular manifestations are raised intraocular pressure and choroidal haemangioma, ipsilateral to the facial hemangioma². Glaucoma results from angle anomalies, aqueous overproduction from ciliary body haemangioma, or increased resistance to aqueous outflow secondary to raised episcleral venous pressure. Glaucomatous damage to the optic nerve may be present in 60% of the patients, with age of onset varying from newborn to 41 years³. When elevated intraocular pressure develops in infancy, it may cause glaucomatous optic neuropathy or buphthalmos. Patients in whom the haemangioma involves the upper lid appear to be in particular more susceptible⁴.

Patients who manifest the disease early in life demonstrate gonioscopic abnormalities similar to children with primary congenital glaucoma. Thick uveal meshwork, poorly developed scleral spur, attachment of ciliary muscle direct to trabecular meshwork and anteriorly inserted iris root have been histologically demonstrated⁵. Late onset glaucoma arises from elevated episcleral venous pressure resulting from multiple arterio-venous shunt vessels draining Schlemms Canal. Glaucoma in Sturge Weber Syndrome is difficult to treat with lower success rates than in primary congenital glaucoma⁵. There is no universal accepted single management plan for these cases. Multiple surgical procedures are commonly needed.

The purpose of this study is to evaluate the results of various treatment modalities in the children presenting with Sturge Weber Syndrome to the Paediatric Ophthalmology clinic of the Ophthalmology Department of Hospital Kuala Lumpur.

METHOD

This is a retrospective descriptive study of consecutive patients managed in the Paediatric Ophthalmology clinic in Hospital Kuala Lumpur. The aim of this study is to evaluate the management methods and the surgical modality used for Intra Ocular Pressure (IOP) control in all the cases of Sturge Weber Syndrome in the paediatric age group been seen in the Paediatric Ophthalmology clinic from January 2008 to June 2011.

RESULTS

There were ten cases of Sturge Weber Syndrome were referred for further management to Paediatric Ophthalmology Clinic in view of capillary vascular malformation. The youngest child presented at 1 month of age while the eldest presented at 94 months of age. The median age at the time of surgical intervention was 36 months. The male to female ratio was 3:2. Of these 10 cases, 7 presented with unilateral facial angiomas. Two of these 10 patients presented with choroidal haemangioma while a further two patients presented with ocular melanosis. There were 3 cases of bilateral facial port wine stain; one case required unilateral surgery for IOP control while the other two required bilateral surgery for IOP control.

In total there were ten patients in this retrospective study, for a total of 13 eyes. Two of them were not included in the final surgical outcome as one had defaulted follow up while another was treated conservatively as the ocular pressures were within normal. Only one eye of a bilateral facial angioma required surgical treatment resulting in total of 10 eyes with surgical intervention in our retrospective study.

The mean age of first surgical intervention was 30.8 months. The most frequently performed surgery in this case series was trabeculectomy augmented with Mitomycin-C 0.5%. This was followed by Trabeculotomy and Glaucoma Drainage Implant.

From the total of 8 patients, only 3 patients had undergone a single surgical procedure while one patient had undergone a total of 4 procedures to control the ocular pressures.

Of the 10 eyes that were operated on, the mean pre-operative IOP was 28.4 mmHg while post operatively, the mean IOP achieve was 10.4 mmHg. There was a mean drop of 18 mmHg.

Of the operated eyes, none of the eyes required anti glaucoma medication on subsequent follow up. In our retrospective study, only one patient with Sturge Weber syndrome who presented at 48 months had good IOP control with anti-glaucoma medication.

DISCUSSION

In our retrospective study, child with facial capillary vascular malformation and uncontrolled IOP less than 12 months of age, underwent trabeculotomy as treatment of choice while others who presented age one year and older, trabeculectomy and drainage implant were the choice of treatment. Trabeculotomy is the primary surgical procedure in early-onset glaucoma related Sturge Weber Syndrome⁵. In our study 2 out of 3 eyes who underwent trabeculotomy as primary surgical

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Age in Months	Gender	Race	Presentation Age	Angioma	Surgical Age	Type of 10 surgery	Both Eye Operation (other eye)	Ocular Manifestation	IOP RE
42	Female	Chinese	1	Bilateral	10	Trabeculectomy	Trabeculectomy	Congenital Ocular Melanosis	23
24	Female	Indian	2	Unilateral	3	Trabeculectomy	.	None	10
31	Male	Malay	1	Bilateral	1	Trabeculectomy	Valve Implant	Choroidal hemangioma	24
57	Male	Malay	48	Unilateral	48	None	.	Choroidal hemangioma	38
43	Male	Malay	36	Unilateral	39	Trabeculectomy	.	None	22
13	Female	Malay	1	Unilateral	.	None	.	None	10
90	Male	Indian	76	Unilateral	82	Trabeculectomy	.	None	32
46	Female	Malay	18	Bilateral	36	Valve Implant	None	Congenital Ocular Melanosis	32
130	Male	Malay	94	Unilateral	127	Trabeculectomy	.	None	37
8	Male	Indian	1	Unilateral	2	Trabeculectomy	.	None	13

IOP LE	More than 1 surgery	Total Procedures Done	Last IOP RE	Last IOP LE	Additional Topical Medication	Procedures Done	Before OP	After OP	Drop IOP
28	Others	4	10	10	No	RE resuture+LE 5FU	23	10	13
22	Valve Implant	2	14	12	No		28	10	18
34	Valve Implant	2	17	4	No		22	12	10
20	None	.	.	.	No	Defaulter	24	17	7
34	Trabeculectomy	2	14	10	Yes	Other Eye/ RE Revision	34	4	30
16	None	0	10	11	No	Other Eye/ RE revision	34	10	24
22	None	1	14	2	No	GutMaxide	22	2	20
24	None	1	18	12	No		32	18	14
14	Others	3	6	12	No	a/c reform+ Autologo	37	6	31
34	None	1	12	20	No		34	20	14

Note: The last IOP recorded was at 2 months or more after the last procedure done on the eye.

procedure required additional surgery to control intraocular pressure (IOP) but none had complication from the surgery.

In our case series, 5 of 10 eyes underwent trabeculectomy with Mitomycin-C as treatment of choice. In our study, 4 out of 5 operated eyes who received trabeculectomy as initial treatment required second surgery to correct the complication of the procedure. The complications in this series noted were retracted conjunctiva, shallow anterior chamber secondary to excessive drainage and choroidal effusion. Except for two limited choroidal effusion that resolved with time, all complications had further surgery to correct the problems as soon as they were detected.

CONCLUSION

The management of Sturge Weber related glaucoma in children, has always been a therapeutic challenge. Surgical approach remains the treatment of choice in controlling the secondary glaucoma. Trabeculectomy is recommended as primary surgical option for early onset glaucoma, especially children less than 1 year of age. However sustained control of IOP is difficult. Trabeculectomy augmented with anti-metabolite has been proven to be the best surgical option to control IOP despite complication that could arise from sudden

changes in pressure. In our center glaucoma drainage devices are usually reserved for patients whose trabeculectomy is failed. It is important to avoid sudden drastic reduction in IOP intraoperatively to avoid potential complications.

The management of Sturge Weber Syndrome associated raised intraocular pressure is managed surgically in most instances by the Paediatric Ophthalmology team at Hospital Kuala Lumpur. The target intraocular pressure can be achieved with surgical management alone without the use of anti-glaucoma medication, although more than one surgical procedure may be required to achieve this.

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