

Treatment results of glaucoma associated with Sturge-Weber syndrome

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ABSTRACT

Purpose: To evaluate the efficacy and safety of different treatment options in glaucoma associated with Sturge-Weber syndrome.

Materials and Methods: The files of the patients who were diagnosed as Sturge-Weber syndrome associated glaucoma and followed up for at least 6 months in Ege University Glaucoma Department between 2007 and 2023 were reviewed retrospectively. Demographic data, examination findings, treatments, intraocular pressure and complications before and after treatment were evaluated.

Results: 29 eyes of 22 patients (14 females, 8 males) were included in the study. The mean age was 16.8 ± 21.2 (0.02-71) years, the follow-up period was 68.1 ± 55.9 (6-192) months. The mean intraocular pressure was 24.2 ± 7.6 (8-46) mmHg at the first visit, 16.3 ± 5.6 (6-30) mmHg at the last visit. While 15 eyes (51.7%) were followed up with topical antiglaucomatous treatment alone, glaucoma surgery was needed in 14 eyes (48.2%) due to high intraocular pressure despite maximum medical treatment. Dorzolamide-timolol combination was preferred in 18 of 24 eyes (75%) receiving topical treatment as the most commonly preferred agent for medical treatment. Trabeculectomy was performed in 9 (64%) eyes as the most commonly preferred surgery and intraocular pressure was measured as <22 mmHg in 8 of 9 eyes (88.8%). Among the early postoperative complications, hypotony was observed in 7 eyes, choroidal detachment in 3 eyes, and hyphema in 1 eye. Complications completely regressed with medical treatment. There were no destructive complications such as retinal detachment, endophthalmitis, phthisis bulbi.

Conclusion: Glaucoma is the most common ocular complication in Sturge-Weber syndrome. Although it is a condition that requires a high rate of surgery, the treatment results are satisfactory.

Keywords: Glaucoma; Sturge-Weber syndrome; neurocutaneous syndrome; medical treatment; glaucoma surgery

INTRODUCTION

Sturge-Weber syndrome (SWS) is a sporadic congenital neurocutaneous disease characterized by abnormal vascularization of the brain, skin and the eye.¹ It occurs as a result of a somatic activating mutation in GNAQ and the incidence has been reported as 1/20.000-1/50.000. Facial capillary malformation (port wine stain), glaucoma and leptomeningeal angioma are the main clinical features of the complete form.² Ocular involvement, especially glaucoma and choroidal hemangioma (CH), is observed in half of the patients. Glaucoma is the most common

ocular involvement and resulted from the anterior chamber malformations, increased pressure in the episcleral veins and changes in ocular hemodynamics.^{3,4,8} Glaucoma associated with SWS is usually congenital but can also develop in adults.^{4,8} Although there is no exact age limit, patients diagnosed under 2 years of age are categorized as early-onset glaucoma. Early-onset glaucoma, which accounts for 60% of glaucoma associated with SWS, has lower success rates with medical and surgical treatments compared to primary congenital glaucoma.³ The management of glaucoma associated with SWS requires both medical

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and surgical interventions for better control.⁴ Treatment should be planned on a case-by-case basis, depending on intraocular pressure (IOP), the patient's age, the stage of the disease and the type of glaucoma.^{4,10} Surgical options include angle procedures such as trabeculotomy, filtration procedures such as trabeculectomy and glaucoma drainage device implantation surgeries or combination procedures.⁵⁻⁹ The risk of serious complications, such as hemorrhage increases, especially in the presence of diffuse CH and it should be taken into account while deciding the surgical method.⁵ Also it is recommended to maintain a stable IOP during and after surgery in order to avoid hypotony and choroidal detachment (CD).⁶ Cyclodestructive procedures in combination with topical treatment are an effective and safe treatment option.^{11,12}

The aim of this study was to evaluate the efficacy and safety of different treatment options in glaucoma associated with SWS.

METHODS

The study was conducted after the approval of the Ege University Ethics Committee and Board and by the tenets of the Helsinki Declaration. The research clinic is a tertiary, referral hospital situated in the western part of the Turkey and specialized in glaucoma.

The files of the patients who were diagnosed as glaucoma associated with SWS and followed up for at least 6 months in Ege University Glaucoma Department between January 2007 and June 2023 were reviewed retrospectively. Demographic data, treatments, IOP and complications

before and after the treatment were evaluated. Patients with insufficient records and who has <6 months of followup were excluded from the study.

Statistical analysis was performed using IBM SPSS Statistics Software (IBM, Illinois, USA). Categorical results are given as frequency and percentage, and continuous variables as mean and standard deviation.

RESULTS

29 eyes of 22 patients (14 females, 8 males) were included in the study. The mean age was 16.8 ± 21.2 (0.02-71) years, and the follow-up period was 68.1 ± 55.9 (6-192) months.

The mean IOP was 24.2 ± 7.6 (8-46) mmHg at the first visit and 16.3 ± 5.6 (6-30) mmHg at the last visit. While 15 eyes (51.7%) were followed up with topical antiglaucomatous treatment alone, glaucoma surgery was needed in 14 eyes (48.2%) due to high IOP despite medical treatment. IOP was measured as <22 mmHg in 12 of 15 eyes (80%) receiving only topical treatment. Carbonic anhydrase inhibitor / beta blocker combinations was preferred in 18 of 24 eyes (75%) receiving topical treatment as the most commonly preferred agent for medical treatment. Prostaglandin analogs was preferred in 15 eyes (62.5%), alpha-2 adrenergic agonists in 7 eyes (29.2%), prostaglandin analog / beta blocker combinations in 2 eyes (8.3%), alpha-2 agonist / beta blocker combinations in 1 eye (4.1%) and carbonic anhydrase inhibitors in 1 eye (4.1%). Topical antiglaucomatous agents have effectively controlled IOP (Table 1). Trabeculectomy was performed in 9 (64%), drainage device implantation surgery in 7 (50%),

Table 1. The number of eyes applied to antiglaucomatous agents and their effects on IOP.

Antiglaucomatous agents	Number of eyes	Pre-treatment IOP mmHg \pm SD	Post-treatment IOP mmHg \pm SD	Values of <i>p</i>
Carbonic anhydrase inhibitor / beta blocker combinations	18	25.8 \pm 7.1	17 \pm 5.4	0.000
Prostaglandin analogs	14	26.7 \pm 7.3	18.1 \pm 5.6	0.000
Alpha-2 adrenergic agonists	7	22.4 \pm 11.9	14.4 \pm 3.8	0.000
Prostaglandin analog / beta blocker combinations	2	29 \pm 1.4	23.5 \pm 0.7	-
Alpha-2 agonist / beta blocker combinations	1	30	20	-
Carbonic anhydrase inhibitors	1	30	23	-
IOP: Intraocular pressure, SD: Standart deviation				

non-penetrating glaucoma surgery in 2 (14%), gonioscopy-assisted transluminal trabeculotomy (GATT) in 2 (14%), trabeculotomy in 2 (14%), and cyclodestruction in 3 (21%) eyes. As shown in Table 2, surgical treatments have effectively controlled IOP. In terms of surgical success, IOP was measured as <22 mmHg in 8 of 9 eyes (88.8%) that underwent trabeculectomy, in 3 of 4 eyes (75%) that underwent trabeculotomy, in 5 of 7 eyes (71.4%) that underwent Ahmed glaucoma valve implantation surgery, and in 2 of 3 eyes (66.6%) that underwent cyclodestructive procedures. While a single surgery was sufficient in 8 of the cases, 2 or more surgical procedures were required in 6 of the cases. Additional topical antiglaucomatous use was

required in 9 (64%) of the postoperative cases. As shown in Table 3, topical and surgical treatments have effectively controlled IOP.

Among the early postoperative complications, hypotonia was observed in 7 eyes, CD in 3 eyes, and hyphema in 1 eye, and the complications completely regressed with medical treatment. Systemic corticosteroid treatment was applied for hypotonia and CD, systemic corticosteroid and cycloplegic agents were used for hyphema. There were no destructive complications such as retinal detachment, endophthalmitis, phthisis bulbi. As shown in Table 4, trabeculectomy surgery is the most preferred surgery and the frequency of complications is relatively low.

Table 2. The number of eyes applied to surgical treatments and their effects on IOP.

Type of surgery	Number of eyes	Pre-treatment IOP mmHg \pm SD	Post-treatment IOP mmHg \pm SD	Values of p
Trabeculectomy	9	24.3 \pm 5.9	15 \pm 5.4	0.000
Angle procedures	4	27.2 \pm 2.7	19 \pm 3.5	0.002
▪ Ab-externo trabeculotomy	2	27.5 \pm 2.1	17 \pm 1.4	-
▪ GATT	2	27 \pm 4.2	21 \pm 4.2	-
Ahmed glaucoma valve implantation	7	28.9 \pm 11.3	16.6 \pm 8.6	0.002
Cyclodestructive procedures	3	34.7 \pm 9.9	13.7 \pm 8	0.098
▪ Cyclophotocoagulation	3	34.7 \pm 9.9	13.7 \pm 8	0.098
GATT: Gonioscopy assisted transluminal trabeculotomy, CD: Choroidal detachment, IOP: Intraocular pressure, SD: Standart deviation				

Table 3. The number of eyes applied to topical and surgical treatments and their effects on IOP.

Type of treatment	Number of eyes	Pre-treatment IOP mmHg \pm SD	Post-treatment IOP mmHg \pm SD	Values of p
Only medical treatment	15	22.8 \pm 5.8	16.4 \pm 4.8	0.001
Number of active ingredients				
▪ 1	3	16.3 \pm 6.8	13 \pm 1.7	0.522
▪ 2	6	24.5 \pm 5.1	15 \pm 4.9	0.023
▪ 3	10	27.4 \pm 4.3	20.9 \pm 4.7	0.000
▪ ≥ 4	5	26.4 \pm 11.9	14.8 \pm 4.4	0.037
Surgical treatment	14	25.7 \pm 9.2	16.3 \pm 6.5	0.000
Number of surgeries				
▪ 1	8	21.2 \pm 8.2	15.6 \pm 6.8	0.007
▪ ≥ 2	6	31.7 \pm 7.2	17.2 \pm 6.6	0.007
All eyes	29	24.2 \pm 7.6	16.3 \pm 5.6	0.000
IOP: Intraocular pressure, SD: Standart deviation				

Table 4. Complications related with glaucoma surgeries.

Type of surgery	Number of eyes	Complications, n (%)		
		Hypotonia	CD	Hyphema
Trabeculectomy	9	2 (22.2%)	1 (11.1%)	0 (0%)
Angle procedures	4	1 (25%)	1 (25%)	1 (25%)
▪ Ab-externo trabeculotomy	2	1 (50%)	1 (50%)	0 (0%)
▪ GATT	2	0 (0%)	0 (0%)	1 (50%)
Ahmed glaucoma valve implantation	7	4 (57.1%)	1 (14.2%)	0 (0%)
Cyclodestructive procedures	3	0 (0%)	0 (0%)	0 (0%)
▪ Cyclophotocoagulation	3	0 (0%)	0 (0%)	0 (0%)

GATT: Gonioscopy assisted transluminal trabeculotomy, CD: Choroidal detachment

DISCUSSION

Management of glaucoma associated with SWS is difficult and depends on the onset of the disease and the underlying pathophysiology. In early-onset glaucoma, medical treatment is usually tried as first-line treatment, but surgery is required for the majority of patients.³ In a study, antiglaucomatous drugs successfully controlled IOP within a 62-month follow-up period in only seven eyes (31.8%) of 22 eyes mostly with early-onset glaucoma.¹³ Another retrospective study found that seven of the eight patients (87.5%) with early-onset glaucoma associated with SWS treated with beta-blockers and carbonic anhydrase inhibitors required glaucoma surgery to lower IOP.¹¹ In late-onset glaucoma associated with SWS, medical treatment remains the first choice.³ In our study, glaucoma surgery was required in 14 of 29 eyes (48.2%). The remaining 15 eyes were followed up with topical antiglaucomatous therapy alone, and in 12 eyes (80%) IOP was measured as <22 mmHg at the last follow-up. In the case of surgical treatment, trabeculectomy is an important surgical option. In a study in 6 patients with late-onset glaucoma with a mean age of 22.4 years, positive outcomes were reported with trabeculectomy at nine months to nine years of follow-up. Only four patients required additional medical treatment and one patient required re-trabeculectomy.¹⁴

In a review study conducted in 42 eyes, surgical success in glaucoma associated with SWS patients was examined and more than one surgery was required in 27 of 42 eyes (64%). Surgical results were classified as successful (no additional medications) in 26 (30%) eyes, sufficiently successful (with antiglaucoma drugs) in 15 (18%) eyes, limitedly

successful (need for re-operation) in 23 (26%) eyes and unsuccessful in 23 (26%) eyes. The surgery with the lowest failure rate was trabeculectomy.¹⁵ Similarly, in our study, trabeculectomy was preferred in 9 of the 14 eyes (64%) that underwent surgery, and only one of these 9 eyes had IOP measured as >21 mmHg at the 9 (1-16) years median follow-up.

Sharan et al reported that out of 24 eyes with glaucoma associated with SWS, IOP control was achieved in 9 (37.5%) eyes with single anti-glaucoma agent, in 9 (37.5%) eyes with more than one anti-glaucoma agent. Trabeculectomy surgery was preferred in 10 (41.6%) eyes and glaucoma drainage device implantation was preferred in 2 (8.3%) eyes.¹⁶

Trabeculotomy surgery has shown promising results in the treatment of glaucoma associated with SWS. In the study of Wu et al., 34 eyes of 32 patients underwent trabeculotomy surgery, and the cumulative rate of overall success was 94.1%, 90.5%, 86.6%, 86.6%, and 86.6%, respectively, in three months, six months, one year, two years, and three years.¹⁷ In our study, 3 of the 4 eyes (75%) that underwent trabeculotomy had IOP <22 mmHg at the 5 (2-9) years median follow-up.

Combined trabeculotomy and trabeculectomy is also an another surgical option in patients with glaucoma associated with SWS, but was not preferred in our clinic for these eyes. In a study conducted in 10 eyes of 9 patients, postoperative IOP was observed below 16 mmHg in all eyes after surgery.¹⁸ In another study, the success rate at 42 months after trabeculotomy and trabeculectomy was found

to be 61.1%.¹⁹ In the study of Sood et al., the long-term (mean 12 years) qualified success rate was observed as 41.7% in 22 eyes of 20 patients who underwent combined trabeculotomy and trabeculectomy.⁷

In our study, in 5 of the 7 eyes (71.4%) that underwent Ahmed glaucoma valve implantation, IOP measured within normal limits (<22 mmHg). Kaushik et al reported the cumulative success rate of Ahmed glaucoma valve implantation for approximately 2 years as 75%.²⁰ Hamush et al, reported a similar success rate (79%) in with Ahmed glaucoma valve surgery for the post-operative first 2-years, however, it was decreased to 59% and 30% in 42th and 60th months, respectively.²¹

Cyclodestructive procedures are usually limited to eyes with low visual potential or a high risk of intraoperative complications.²² In our study, IOP was observed within normal limits in 2 of 3 eyes that underwent diode laser cyclophotocoagulation as cyclodestructive procedures. There were no destructive complications such as retinal detachment, endophthalmitis, phthisis bulbi.

SWS patients are at higher risk of severe choroidal effusion or expulsive bleeding after surgery.³ Although the underlying mechanism of choroidal effusion is similar to the occurrence of effusion observed with a significant decrease in IOP after glaucoma surgery, yet patients with SWS have a faster occurrence of choroidal effusion, more massive effusion, and a higher prevalence after filtration surgery.^{23,24} Iwach et al. reported intraoperative choroidal effusion in 24% of cases undergoing trabeculectomy, including 40% of five cases in the early-onset group and 17% of 12 cases in the late-onset group.²⁵ The presence of CH puts the eyes at greater risk for both intraoperative and postoperative choroidal effusion. Pandey et al. reported that 83.3% of eyes with choroidal effusion had CH.²⁶ In our study, only 3 (%) of the 29 eyes developed postoperative CD and the findings regressed with medical treatment using oral corticosteroids within weeks (range 1-6).

Glaucoma is the most common ocular complication associated with SWS and can be seen in patients of all ages. Although it is a condition that requires surgery at a high rate and many eyes require re-operations, the results of the treatment are satisfactory with careful management of post-operative complications.

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