

# Ocular findings in Sturge-Weber syndrome

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**PURPOSE.** We reviewed the rare ocular findings of Sturge-Weber syndrome (SWS) and the results of implantation of the Ahmed valve in cases associated with glaucoma.

**METHODS.** Seven patients (range 18 to 52 years) diagnosed as SWS were reviewed as clinical findings over two years. An anterior chamber maintainer was placed in patients with glaucoma to maintain stable intraocular pressure and minimize the risk of intraocular hemorrhage due to sudden pressure changes during surgery, when an Ahmed valve is implanted.

**RESULTS.** Episcleral venous vessels were prominent in all cases. Diffuse choroidal hemangiomas were seen in three cases. Nevus of Ota was observed in only one case. Three cases had juvenile glaucoma. One also had buphthalmos. No intraocular hemorrhage or choroidal effusion was observed intraoperatively in valve-implant patients. The surgical treatment of the patients with glaucoma gave favorable outcomes.

**CONCLUSIONS.** Rare ocular findings such as choroidal hemangioma and nevus of Ota are sometimes seen in SWS. Drainage valve implantation, with an anterior chamber maintainer, is a good choice for treatment when surgery is done in cases with glaucoma. This method may reduce the risk of intraoperative suprachoroidal effusion and expulsive hemorrhage by stabilizing intraocular pressure within normal limits during the surgery. (*Eur J Ophthalmol* 2000; 10: 239-43)

**KEY WORDS.** Sturge-Weber syndrome, Glaucoma, Ahmed valve, Diffuse choroidal hemangioma, Nevus of Ota

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## INTRODUCTION

Sturge-Weber syndrome (SWS) is a dermato-oculo-neural syndrome involving cutaneous facial nevus flammeus in the area of the first and/or second division of the trigeminal nerve, ipsilateral diffuse cavernous hemangioma of the choroid, and ipsilateral leptomeningeal hemangioma (1). The majority of SWS patients have sporadic, nonfamilial disease. SWS is the only phacomatosis without a hereditary tendency (2). The classical cutaneous feature of SWS is facial nevus flammeus (1) which is usually unilateral and may be associated with hypertrophy of the involved skin of the face. One of the ocular manifestations of

SWS is diffuse choroidal hemangioma (3), usually on the same side as facial nevus flammeus. The fundus of the affected side typically has a much more saturated red appearance than the fundus on the uninvolved side. Other ocular abnormalities include conjunctival and episcleral telangiectasia, and ipsilateral congenital, infantile, or juvenile glaucoma (4). Glaucoma occurs in about 70% of patients, on the side of the facial hemangioma (1). Patients in whom the lesion affects the upper eyelid are at increased risk of glaucoma (1).

In this study, we reviewed the frequent and rare associated ocular findings of SWS. We implanted an Ahmed valve in three of seven patients with glaucoma and review the results.

## METHODS

Seven patients diagnosed as SWS were included in this study at Firat Medical Center and Military Hospital in Elazığ over a period of two years (1996 to 1998). Three were female and four were male. Mean age was 27.7 years (range 18 to 52). Examination of the eyelids, anterior segment biomicroscopy, tonometry, gonioscopy, computerized visual field analysis, anterior segment and fundus photography were done in all patients. They were also evaluated by CT and neurological examination.

Initially, medical treatment combined with timolol maleate, dorzolamide hydrochloride and apraclonidine was planned for all the cases with glaucoma until surgery. One hour before surgery, mannitol solution 20% was injected intravenously, 1.5-2 g/kg in 30 minutes. Surgery was done under general anesthesia to reduce the risk of periorbital hemorrhage with local anesthesia. The dilated episcleral vessels at the upper nasal quadrant, which were vulnerable to surgical trauma, were all cauterized. Then a partial-thickness, limbal-based scleral flap was prepared, 2.5x3 mm. An incision into the anterior chamber was made with a 20-gauge stiletto knife at the temporal peripheral cornea. Later, an anterior chamber maintainer was placed in the anterior chamber to keep a stable in-

traocular pressure and minimize the risk of intraocular hemorrhage due to sudden changes of pressure during surgery. An incision into the anterior chamber was made with a 23-gauge stiletto knife at the upper nasal posterior limbus where the tube of the valve is implanted. The valve is fixed to the sclera and the scleral flap is closed with sutures. Patients with glaucoma were followed at three-month intervals and other cases at six-month intervals.

## RESULTS

Three cases had lesions on the left side of their faces, and four on the right. All had prominent episcleral venous vessels (Fig. 1). Three had lesions only on the lower eyelids and four had them on both the upper and lower lids. Diffuse choroidal hemangiomas were found in three cases (Fig. 2a-b). Nevus of Ota was identified in only one case. The findings in the anterior segment of this case consisted of vast areas of bluish pigmentation of the conjunctiva and episclera, and a heavily pigmented iris. The main features of the cases with SWS are summarised in Table I.

Three patients had unilateral juvenile glaucoma, with intraocular pressures (IOP) 38, 40 and 36 mmHg, and c/d ratios (cup to disc ratio) 0.7, 0.8 and 0.9 (Fig. 2a).

**TABLE I - OCULAR FINDINGS IN CASES WITH STURGE-WEBER SYNDROME**

Case no.	Age sex	Side involved	Visual acuity	Glaucoma	Initial IOP	c/d	Treatment	Final IOP	Associated findings	Follow-up (yrs)
1	21, M	Left	20/20	Juvenile	38	0.7	Valve implantation	17	DCH Buphthalmos	2.5
2	22, M	Left	20/20		14	0.2			PEV	2.0
3	52, F	Right	20/40	Juvenile	40	0.8	Valve implantation	17	PEV	1.5
4	22, F	Right	20/20		13	0.3			PEV	1.5
5	22, M	Right	20/20		16	0.3			DCH	1.5
6	18, F	Left	20/20		14	0.3			Ota Nevus	1.0
7	37, M	Right	20/40	Juvenile	36	0.9	Valve implantation	14	DCH	1.0

IOP: intraocular pressure, c/d: cup-to-disc ratio, DCH: diffuse choroidal hemangioma, PEV: prominent episcleral vessels

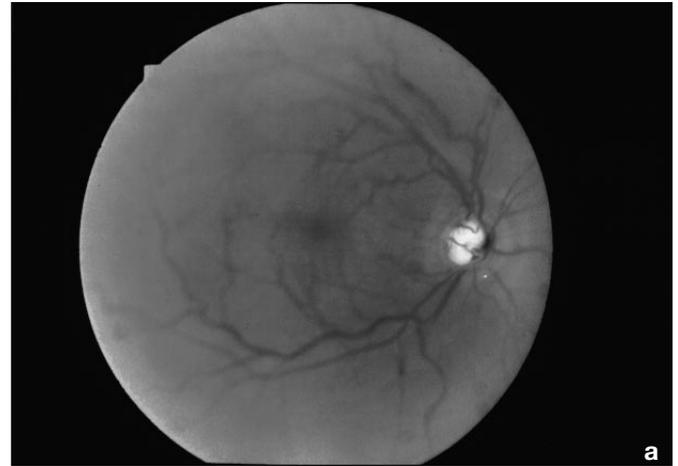


**Fig. 1** - Facial nevus flammeus and prominent episcleral vessels in case 1, with Sturge-Weber syndrome.

There was blood in Schlemm's canal in the gonioscopy and a definite concentric narrowing in the visual field. One patient had a buphthalmic eye and the horizontal corneal diameter measured 14.5 mm. IOP in the patients with glaucoma could not be reduced to less than 25 mmHg with medical treatment. No intraocular hemorrhage or choroidal effusion was observed intraoperatively. Postoperative IOP in these cases was around 16 mmHg without any medication. The c/d ratios and visual fields of these patients did not progress. We did not observe any complications caused by the Ahmed valve during follow-up.

## DISCUSSION

Classic SWS comprises the triad of nevus flammeus in the distribution of the trigeminal nerve that respects the vertical midline, ipsilateral glaucoma and intracranial angiomas. In patients with glaucoma, both the upper and lower eyelids are usually involved in the facial telangiectasis (5). Cibis et al, report that glaucoma occurs in about one third of patients with SWS (6). We observed glaucoma in 43% of our cases. The increased pressure is the result of either an angle anomaly similar to that seen in primary congenital glaucoma or raised episcleral venous pressure (1). This high pressure can cause open-angle glaucoma because of obstruction of the outflow of aqueous into the venous drainage system (7) and results in engorged episcleral veins. The high pressure lets the Schlemm's canal



**Fig. 2** - Asymmetric fundus appearance in case 7, with diffuse choroidal hemangioma. **a)** Right fundus has a saturated red color caused by diffuse choroidal hemangioma. Note the extensive deep cupping of the optic disc. **b)** Left fundus has a more orange choroid than the right eye. The optic disc appears normal.

fill with blood, that can be seen by gonioscopy. All these clinical findings were present in our cases with glaucoma.

The treatment of glaucoma in patients with SWS is usually difficult and the best way to treat it is still controversial. In cases whose angle is similar to that of primary infantile glaucoma, goniotomy is a reasonable first step (5). In cases with increased episcleral venous pressure, a fistulization procedure such as trabeculectomy or drainage implant may be more successful (9, 10). Initial treatment of all kinds of glaucoma is usually medical but in resistant cases filtration surgery and drainage implants are inevitable despite the high risk of intraoperative bleeding and suprachoroidal effusion in 17-68% of cases (1, 10-12). An

anterior chamber maintainer was used to stabilize the anterior chamber and to control IOP, and is suggested as a precautionary measure (13). Some surgeons recommend the placement of two or three posterior sclerotomies to prevent choroidal expansion and intraoperative hemorrhage prior to entry into the anterior chamber (5, 11).

Our cases with glaucoma were at a terminal stage and we had immediately planned surgical treatment in our study. Ahmed valve implantation was the surgical method of choice. We avoided trabeculectomy for several reasons, one being the high risk of intraoperative hemorrhage and suprachoroidal effusion. Another is the presence of dilated episcleral vessels which may lead to scar tissue in the trabeculectomy area due to a high rate of fibrin extravasation. This process would cause failure in the long term. Complications such as intraoperative suprachoroidal effusion and expulsive hemorrhage are observed less in closed system surgery. For these reasons, we used both an anterior chamber maintainer and valve implantation. We saw no problems during the intraoperative and postoperative period. IOP was within the desired limits after surgery.

The choroidal hemangioma is a benign vascular tumor of the choroid. It occurs in two distinct clinical forms: a circumscribed form that is almost always isolated and nonsyndromic, and a diffuse form, usually part of SWS (3). Most circumscribed choroidal hemangiomas are noted first when they produce visual

symptoms caused by accumulation of serous subretinal fluid, degenerative changes in the macular retina, or both (3). Diffuse choroidal hemangiomas are usually detected at baseline ophthalmic evaluation of patients who have a facial nevus flammeus, before the onset of symptoms. Visual impairment in eyes that have either diffuse or circumscribed choroidal hemangioma ranges from none to total blindness (3).

The association of SWS with nevus of Ota is infrequently reported (14). One of our cases had the association. During follow-up this patient had no complaints.

It is possible to see rare ocular findings such as choroidal hemangioma and nevus of Ota in patients with SWS, besides the common ophthalmic features. When surgery is needed in cases with glaucoma, drainage valve implantation supported by an anterior chamber maintainer appears to be a good choice for treatment. This method may reduce the risk of intraoperative suprachoroidal effusion and expulsive hemorrhage by stabilizing IOP during surgery.

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