

Sturge-Weber Syndrome and Glaucoma

BY LISA A. HERRYGERS, MD

CASE PRESENTATION

A 10-year-old white male with Sturge-Weber syndrome was referred for evaluation of glaucoma. The patient had developed elevated IOP in his left eye at 6 years of age, and therapy with topical glaucoma medications had maintained an IOP in the high teens. At the time of his presentation, the patient was taking Xalatan (Pfizer Inc., New York, NY) for his left eye.

On examination, the patient's UCVA was 20/20 OD and 20/25 OS. Goldmann applanation tonometry readings were 12 mm Hg OD and 24 mm Hg OS. The patient had a port wine stain on the left side of his face that involved the upper and lower lids and cheek, and he had a port wine stain on his right cheek that did not involve the upper right eyelid (Figure 1). Gonioscopy revealed open angles with angle structures that appeared normal, although blood was visible in Schlemm's canal. The remainder of the anterior segment examination was unremarkable. Dilated fundus examination revealed healthy-appearing optic nerves with a C/D of 0.3 OD and 0.8 OS. Fundus examination of the patient's right eye was normal but revealed tortuous vessels and a "tomato-catsup" appearance consistent with choroidal hemangioma in his left eye (Figure 2).

Over the course of the next 2 years, the IOP of the patient's left eye increased to 28 mm Hg. He began maximal topical glaucoma treatment, consisting of Lumigan (Allergan, Inc., Irvine, CA), Alphagan P (Allergan, Inc.), and Cosopt (Merck & Co., Inc., West Point, PA). Visual field testing remained within normal limits, but the patient developed progressive cupping of his left optic nerve.

HOW WOULD YOU PROCEED?

1. Would you advance medical treatment to Diamox (Wyeth Pharmaceuticals, Philadelphia, PA) or perform a surgical procedure?
2. If opting for surgery, would you perform a trabeculectomy with adjunctive mitomycin C (MMC) or place a seton?
3. What are the risks of performing filtering surgery in a patient with Sturge-Weber syndrome?

SURGICAL COURSE

The patient underwent an uneventful trabeculectomy with MMC in his left eye without the preplacement of posterior sclerotomies. No postoperative choroidal effusion or choroidal detachment occurred, and the patient developed a low, diffuse bleb. His IOP was in the low teens without topical medications. The optic nerve and visual fields have remained stable for 4 years, but I follow him closely for any increases in IOP.

DISCUSSION

Sturge-Weber syndrome is a phakomatosis that has no known hereditary pattern. Individuals with this congenital anomaly have angiomatous involvement of the meninges and brain (leptomeningeal angiodysplasia), which causes jacksonian seizures in 85% of patients, mental retardation in 60% of patients, and cerebrocortical atrophy.¹ Glaucoma has been reported in between 30% and 71%.²⁻⁴ Glaucoma is associated with a facial angioma (port wine stain) described as deep burgundy with various degrees of facial hypertrophy, usually in the distribution of the first and second divisions of the trigeminal nerve. Although the facial angioma is usually unilateral, between 10% and 30% are bilateral.⁵⁻⁷ Patients with upper-lid involvement are more likely to develop glaucoma.⁸ Vascular malformations of the conjunctiva, episclera, choroid, and retina are also found.



Figure 1. The port wine stain on the left side of the patient's face involved his upper and lower eyelids and cheek, whereas the port wine stain on his right cheek did not involve his upper right eyelid.

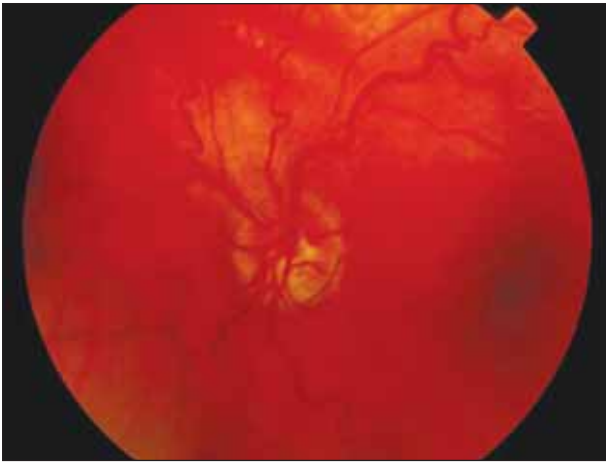


Figure 2. The patient's left eye demonstrated a "tomato-cat-sup" fundus consistent with choroidal hemangioma and an increased C/D.

Of the patients who develop glaucoma, approximately 60% have congenital glaucoma with buphthalmos, anisometropia/amblyopia, and advanced optic nerve cupping. Forty percent of patients develop glaucoma later in childhood or in adulthood.^{2,3,9} Congenital glaucoma in Sturge-Weber syndrome is often associated with immature angle development, similar to the angle development seen in primary congenital glaucoma. Histopathologic studies from trabeculectomy specimens have ranged from a poorly developed scleral spur, a thickened uveal meshwork, and an anteriorly displaced iris root to specimens with little structural abnormality. Juvenile or later-onset glaucoma patients often have normal-appearing angles. Histopathologic studies have shown little structural abnormality, but examples of an anteriorly displaced ciliary muscle and the juxtacanalicular region replaced by vascular structures and connective tissue have also been observed.

In 1973, Weiss¹⁰ proposed two mechanisms for the development of glaucoma in Sturge-Weber syndrome. In congenital glaucoma, he hypothesized that increased episcleral venous pressure from episcleral angiomas combined with abnormal angle development contributes to the development of glaucoma. In later onset cases with normal-appearing angles, he thought that glaucoma occurred due to elevated episcleral venous pressure, which has been found in patients with Sturge-Weber syndrome.¹⁰⁻¹²

Regardless of the mechanism of the development of glaucoma, controlling IOP depends on the patient's presentation. In congenital glaucoma cases with buphthalmos, surgical management with goniotomy and trabeculectomy should be attempted initially. Because these procedures

are often ineffective, the patient may require filtering surgery to control his IOP. In juvenile or later-onset glaucoma, topical glaucoma medications are often the first-line treatment, but they frequently fail eventually to control the IOP. Limited success with laser trabeculectomy has been reported.³ Trabeculectomy with MMC or aqueous drainage devices have been successful in controlling IOP.

Filtering surgery on patients with Sturge-Weber syndrome carries the risk of massive intraoperative choroidal effusion or expulsive hemorrhage. The choroidal effusions are thought to be secondary to the rapid movement of fluid from the intravascular to the extravascular spaces in the setting of sudden hypotony and increased venous pressure. It has been proposed that this devastating complication can be minimized by the creation of a posterior sclerotomy prior to entering the anterior chamber.¹² Eibschitz-Tsimhoni et al¹³ questioned the necessity of prophylactic posterior sclerotomy, however, in a series of 17 patients with Sturge-Weber syndrome or Klippel-Trenaunay-Weber syndrome. They compared patients who received prophylactic posterior sclerotomy to those who did not and found no significant incidence of suprachoroidal hemorrhage, choroidal effusion, or choroidal detachment in either group.

The management of glaucoma in patients with Sturge-Weber syndrome can be difficult. As with all glaucomas, early detection and treatment are important for maintaining the patient's useful vision. □

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1. Beck RW, Hanno R. The phakomatoses. *Int Ophthalmol Clin*. 1985;25:97.
2. Alexander GL, Norman RM. *The Sturge-Weber Syndrome*. Bristol, England; John Wright & Sons Ltd., 1960.
3. Iwach AG, Hoskins HD, Hetherington J, et al. Analysis of surgical and medical management of glaucoma in Sturge-Weber syndrome. *Ophthalmology*. 1990;97:904-909.
4. Sullivan TJ, Clarke MP, Morin JD. The ocular manifestations of the Sturge-Weber syndrome. *J Pediatr Ophthalmol Strabismus*. 1992;29:349-356.
5. Duke-Elder S, Jay B. *System of Ophthalmology: Diseases of the Lens and Vitreous; Glaucoma and Hypotony*. St. Louis; CV Mosby, 1969.
6. Shaffer RN, Weiss DI. *Congenital and Pediatric Glaucomas*. St. Louis; CV Mosby, 1970.
7. Peterman AF, Hayles AB, Dockerty MB, et al. Encephalotrigeminal angiomas: clinical study of 35 cases. *JAMA*. 1958;167:2169-2176.
8. Font RL, Ferry AP. The phakomatoses. *Int Ophthalmol Clin*. 1972;12:1.
9. Mattox C. Sturge-Weber syndrome. In: Epstein DL, Allingham RR, Schuman JS, eds. *Chandler and Grant's Glaucoma*. 4th ed. Baltimore, MD; Lippincott Williams & Wilkins; 1997: 433-436.
10. Weiss DI. Dual origin of glaucoma in encephalotrigeminal haemangiomas. *Trans Ophthalmol Soc UK*. 1973;93:477-493.
11. Phelps CD. The pathogenesis of glaucoma in Sturge-Weber syndrome. *Ophthalmology*. 1978;85:276.
12. Bellows AR, Chylack LT, Epstein DL, et al. Choroidal effusion during glaucoma surgery in patients with prominent episcleral vessels. *Arch Ophthalmol*. 1979;97:493-497.
13. Eibschitz-Tsimhoni M, Lichter PR, Del Monte MA, et al. Assessing the need for posterior sclerotomy at the time of filtering surgery in patients with Sturge-Weber syndrome. *Ophthalmology*. 2003;110:1361-1363.