

Glaucoma Secondary to Sturge-Weber Syndrome

A discussion of the unique risks and the options for management.

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CASE PRESENTATION

A 33-year-old white female with Sturge-Weber syndrome affecting the upper and lower lids of her left eye was referred for consideration of filtration surgery. She had been diagnosed with secondary glaucoma in her left eye at the age of 7 and had since received constant treatment with topical IOP-lowering medications. She had not undergone previous glaucoma laser or filtration surgery. Her IOP over the last 3 years had increased to between 25 and 35 mm Hg, and another ophthalmologist had discussed with her the potential need for filtration surgery. At the time of presentation, the patient's treatment was limited to topical latanoprost q.h.s., because she was breast-feeding her 2-month-old infant. The IOP in her right eye had always been within normal limits.

Her BCVA measured 20/15 OD and 20/70 OS, with a dense, relative afferent pupillary defect. Slit-lamp biomicroscopy revealed dilated episcleral vessels in her left eye. Her IOP was 19 mm Hg OD and 27 mm Hg OS, with a central corneal thickness of 550 μ m OU. Gonioscopy revealed open anterior chamber angles (grade IV), with blood visible in the inferior 3 clock hours of Schlemm's canal in the patient's left eye. There was marked thinning of the neuroretinal rim of the left optic disc, with diffuse loss of the nerve fiber layer (Figure 1). This finding was confirmed by scanning laser polarimetry (GDx VCC; Carl Zeiss Meditec Inc., Dublin, CA). In addition, thickening of the posterior pole with orange-red discoloration suggested a diffuse choroidal hemangioma. The optic nerve and retina of her right eye appeared to be normal. There were superior and inferior arcuate scotomata in her left eye. The visual field in the patient's right eye was within normal limits (Figure 2).

Comments on Management

DSM: An interesting aspect of this case is that the patient's vision and optic nerve are reasonably well preserved for an adult with Sturge-Weber–related glaucoma. Although she does have moderate-to-advanced glaucoma, glaucoma secondary to Sturge-Weber syndrome is

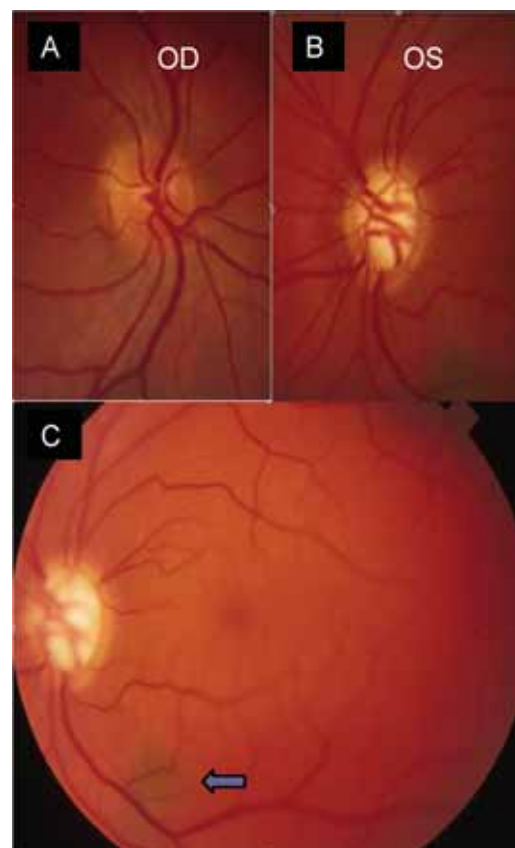


Figure 1. Optic disc photographs demonstrated a marked loss of the neuroretinal rim and a diffuse loss of the retinal nerve fiber layer in the patient's left eye. The optic disc of her right eye was normal (A and B). The left posterior pole appeared to have thickened and had an orange-red discoloration suggesting a diffuse choroidal hemangioma. A flat nevus was noticeable adjacent to the inferior arcade (arrow) (C).

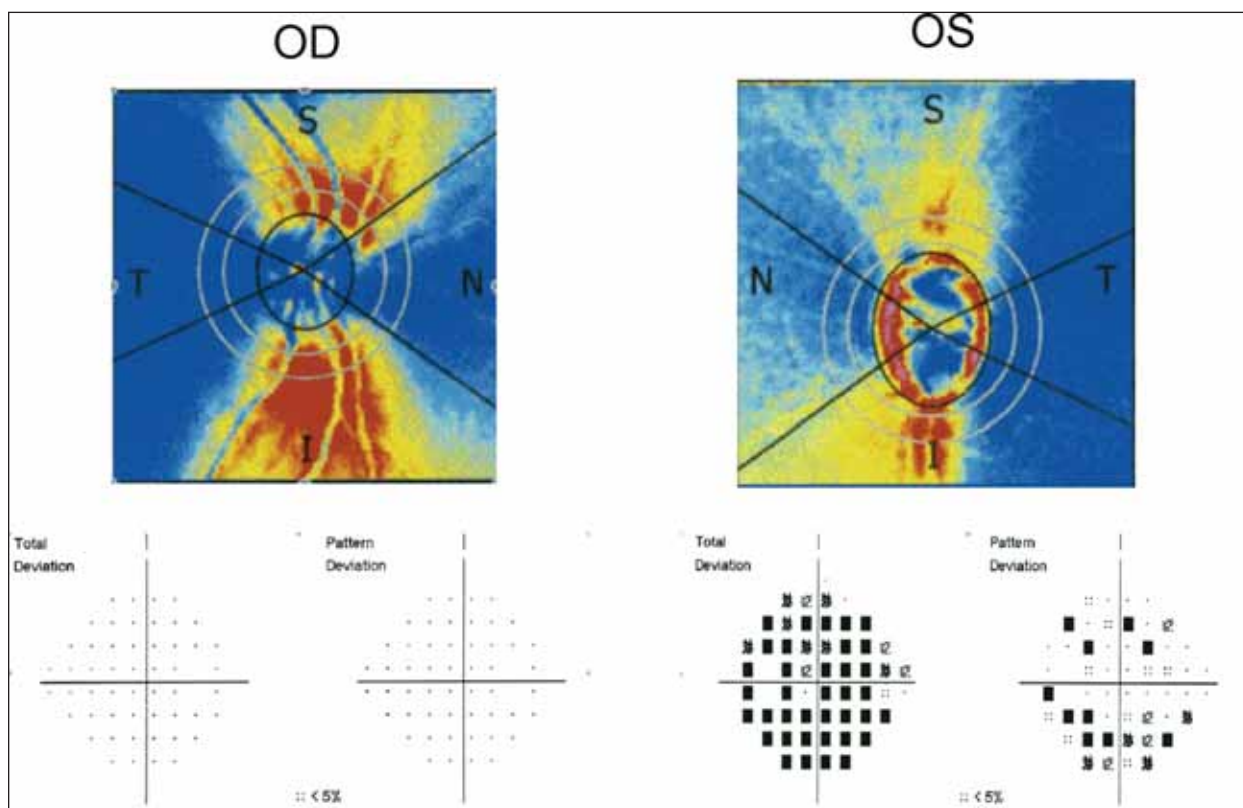


Figure 2. Thickness maps show marked, diffuse thinning of the retinal nerve fiber layer in the patient's left eye. This finding was associated with superior and inferior arcuate scotomata.

frequently at a more advanced stage in adults. Nonetheless, it is important to preserve her vision. With an IOP of around 30 mm Hg, there is a considerable risk of progressive optic nerve damage. The option of adding a topical medication at this stage is limited by the patient's desire to continue breast-feeding. Since laser trabeculoplasty is unlikely to have a favorable effect on IOP, that leaves the option of filtration surgery.

In addition to all of the normal risks associated with trabeculectomy and aqueous-shunting procedures using glaucoma drainage devices, there is the additional risk in Sturge-Weber eyes of choroidal effusions and/or suprachoroidal hemorrhage when a choroidal hemangioma is present.¹ This situation would influence the ophthalmologist's threshold for selecting surgery, and it may not be unreasonable to defer surgery until there is evidence of glaucoma progression.

RNW: In view of the high IOP in the patient's left eye, the advanced stage of her glaucomatous optic neuropathy, and her relatively young age, there is considerable risk of continued damage to her optic nerve. It seems appropriate, therefore, to consider further lowering her IOP.

JCC: If you elected to perform surgery, how would you

modify your technique to reduce the risk of choroidal effusion or hemorrhage?

DSM: I would perform a trabeculectomy using my standard technique with oblique sutures on the back of a trapezoidal flap. If cautery to the cleft edges of the flap can be avoided, one can usually get tight closure with just two sutures. One can then "burp" the wound by applying gentle pressure posterior to the flap during the early postoperative period if the IOP is elevated. I would probably also include a scleral window to act as an emergency drain should choroidal effusion or hemorrhage occur.

During conjunctival dissection, if I find that there are numerous grossly dilated episcleral vessels and the risk of traumatizing them is high, I will consider performing an aqueous-shunting procedure with a glaucoma drainage device. An advantage of a nonvalved glaucoma drainage device (eg, Molteno Implant [Molteno Ophthalmic Limited, Dunedin, New Zealand] or Baerveldt glaucoma shunt [Advanced Medical Optics, Inc., Santa Ana, CA]) is that one may implant it as part of a two-staged procedure, with several weeks between installation and opening. This staging may reduce the risk of postoperative hypotony. In my experience, however, the IOP after aqueous-shunting

procedures (in patients such as this one and others) is usually in the mid- to high teens and not as low as the IOPs that may be obtained after trabeculectomy.

RNW: Some of our colleagues preplace sutures that are positioned on the scleral flap prior to entering the anterior chamber. This technique may allow the rapid closure of the eye and may reduce the length of intraoperative hypotony.

DSM: Another option would be to place an anterior chamber infusion line to maintain positive pressure in the anterior chamber.

FM: The formation of posterior sclerectomies, at least in theory, reduces the risk of developing postoperative choroidal effusions. The expansion of tissue as a result of a choroidal effusion may lead to the rupture of the choroidal blood vessels. Scleral windows may avoid this complication by reducing the formation of choroidal effusions. The need for and benefit of this intervention, however, has not been conclusively demonstrated to date.²

JGC: In an eye like this patient's with 20/70 BCVA, would you consider cyclodestructive procedures?

DSM: Generally, cyclodestruction is the last option on my list. Although this procedure can be very effective at lowering IOP, my experience is in line with the literature, which suggests that this modality carries a significant risk of reducing central vision.³

The patient continued taking latanoprost for 2 more months. After she had discontinued breast-feeding her child, she also began using a topical beta-blocker, which had little effect on her IOP. The patient was reluctant to undergo filtration surgery but consented to argon laser trabeculoplasty with the understanding that the chance of its significantly lowering her IOP was small and that there was a slight possibility that the laser treatment could lead to an elevation of her IOP. Six weeks after argon laser trabeculoplasty, the IOP in her left eye remained elevated at 24 mm Hg. After a detailed discussion with the patient regarding further treatment options, she consented to undergo a trabeculectomy.

The procedure was performed under local anesthesia without complication. The surgeon applied intraoperative 5-fluorouracil (50mg/mL for 90 seconds) with a cellulose sponge. Despite tight closure of the scleral flap and meticulous conjunctival closure, her IOP on the first postoperative day was 14 mm Hg. It varied between 5 and 14 mm Hg during the early postoperative period. Small peripheral choroidal effusions were noted. The surgeon administered one subconjunctival injection of 5-fluorouracil 7 weeks postoperatively.

The patient's BCVA was 20/200 OS during the first postoperative week but improved to 20/70 with pinhole,

which was comparable to her preoperative visual acuity. In the fifth postoperative month, the IOP remained around 7 mm Hg, with a diffuse, quiet filtration bleb that had microcysts.

CONCLUSION

Young patients with moderate-to-advanced glaucoma who experience a persistent, marked elevation in IOP have a high lifetime risk of disease progression. For such an individual, surgery may be indicated in the absence of clear signs of progression. Sturge-Weber syndrome is associated with choroidal hemangiomas, which increase the risk of choroidal effusions and hemorrhage during and after intraocular surgery. It is advisable for surgeons to reduce intra- and postoperative hypotony as a means of potentially decreasing the risk of choroidal expansion. Performing intraoperative infusion of the anterior chamber, preplacing scleral sutures, and tightly closing the scleral flap and posterior sclerectomies may limit the occurrence of these complications. □

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