Case report

Management of childhood glaucoma in Sturge Weber Syndrome: A challenge

Poonam Lavaju, Pawan Mahat
Department of Ophthalmology
BP Koiral Institue of Health Sciences, Dharan, Nepal

Abstract

Background: Sturge-Weber syndrome (SWS) is a rare congenital neuro- oculo-cutaneous disorder. Glaucoma is one of the ocular abnormalities associated with it. Management of glaucoma in SWS is challenging. **Objective**: To report surgical and visual outcome of trabeculectomy in bilateral glaucoma in Sturge-Weber syndrome and the challenges encountered in its management. **Case description**: A six-year-old girl presented with gradually progressive increase in size of both the eyes associated with watering, photophobia and diminution of vision since birth. On examination she was diagnosed to have bilateral SWS with glaucoma, refractive to antiglaucoma medications. She underwent trabeculectomy with intraoperative use of 5- FU. The post operative period was complicated with choroidal effusion in both the eyes with shallow anterior chamber. In the second post operative day, the right eye anterior chamber re-formation was done with ringer lactate solution. There was spontaneous resolution of choroidal effusion with normalization of intraocular pressure with no adverse effect on visual acuity within one week. **Conclusion**: Treatment of glaucoma associated with Sturge-Weber syndrome is challenging. When planning surgical intervention an increased risk of choroidal effusion associated with it, should be kept in mind.

Keywords: Sturge-Weber syndrome, glaucoma, choroidal effusion, trabeculectomy

Introduction

Bilateral Sturge-Weber syndrome (SWS) is a rare congenital disorder. It is characterized by a triad of cutaneous facial angioma, leptomeningeal angioma and ocular manifestations. Glaucoma is common in SWS, reported in 30-70% cases (Kristin et al 2004). The treatment of glaucoma in a patient with SWS is always difficult and the best method to manage is still controversial. It carries high risk of severe intraoperative and postoperative complications such as massive choroidal effusion or hemorrhage and serous retinal detachment.

This article describes a case of bilateral SWS with glaucoma in a six-year- old child managed with trabeculectomy with intraoperative use of 5-FU and its complications.

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Address for correspondence
Dr Poonam Lavaju
Additional Professor
Department of Ophthalmology
BP Koirala Institute of Health sciences, Dharan, Nepal
Mobile no: 00977-9852047026
E-mail: drpoonamlavaju@yahoo.com

Case report

A six-year- old girl was brought by her parents with complaints of gradually increase in size of both the eyes, diminution of vision, photophobia and watering since birth. There

was no history of ocular trauma or significant systemic complaint including no seizure disorder.

She was normally delivered at full term. Family history was not significant. She was treated elsewhere with eye drops 0.5% timolol b.d. and 2% dorzolamide three times a day (t.i.d.) in both the eyes for six months.

Erythematous well defined irregular shaped plaque was present on both the sides of the face involving right ear lobule and right side of upper trunk with mild right sided facial hypertrophy, suggestive of bilateral SWS (Figure 1).

Ocular examination revealed, best corrected visual acuity in the right eye, 2/60 and that in the left eye was perception of light with accurate projection of rays in all the quadrants. The eyes were aligned. Extraocular movements were full in all positions of gaze. Both the eyes were buphthalmic. Tortous conjunctival vessels were present in the right eye in the superotemporal region (Figure 2). Cornea showed presence of Haab's striae and the anterior chamber was deep with loss of iris pattern in both the eyes. The diameter of cornea was 14x14mm and 14x14.5mm in the right and the left eyes respectively. Sclera showed diffuse pigmentation and dilated tortuous episcleral vessels, which were more marked in the right eye. Pupillary reaction showed relative afferent pupillary defect in both the eyes. Lens was clear. Axial length of the right and the left eyes were 27.13 mm and 28.12mm respectively. Intraocular pressure was 26mm Hg in the right eye and 17.3mmHg in the left under halothane, 0.5% timolol and 2% dorzolamide eye drops. Funduscopy examination revealed pale disc with total cupping with dilated tortuous vessels in both the eyes. The left eye had choroidal hemangioma of two disc diameter present superotemporally.

She underwent trabeculectomy with intra-operative use of 5- FU on the right eye followed by the left one week apart. In the postoperative period, she was commenced on eye drops of 1% prednisolone acetate 2 hourly, 5% moxifloxacin four times a day and 1% tropicamide t.i.d.

Postoperatively, the status of visual acuity was the same in both the eyes. On the first postoperative day, the right eye developed massive kissing choroidal effusion with shallow anterior chamber (van Herrick grade III). On the second postoperative day, anterior chamber was reformed with ringer lactate solution. Similarly, in the left eye, on the second postoperative day, there was hypotony with shallow anterior chamber (van Herrick grade I). Funduscopy examination revealed minimal choroidal effusion in the inferior nasal quadrant sparing the macula.

She was started on oral prednisolone 1mg/kg body weight in tapering dose. Within one week, there was spontaneous resolution of choroidal effusion with well formed anterior chamber and normalization of IOP in both the eyes.

At three months follow up, the best corrected visual acuity in the right eye was 4/60 and that in the left 2/60 with accurate projection of rays in all the quadrants. Anterior chambers were well formed with functioning bleb and the IOP was 10mmHg with flat retina in both the eyes.

Discussion

The management of glaucoma with SWS, refractive to medical therapy is challenging and controversial. Several modalities of management had been suggested in the literature. Many surgeons prefer goinotomy (Shaffer et al, 1970; Iwach et al, 1990) or trabeculotomy (Rothkoff et al, 1979; Iwach et al, 1990) as the operation of choice but the success rate is less than that for primary congenital glaucoma (Shaffer et al 1970; Rothkoff et al 1979). The relatively low success rates and high risk of choroidal effusion with goniotomy and trabeculotomy has led to the opinion that an additional filtering surgery to bypass the episcleral veins such as

trabeculectomy or aqueous shunting procedure is current approach in patients refractive to medical therapy (Bellows et al 1979, Ali et al 1990; Iwach et al 1990; Hamush et al 1999; Budenz et al 2000).

Mandal (1999) performed combined trabeculotomy - trabeculectomy in ten eyes of nine patients with SWS with glaucoma and concluded that the procedure was safe, effective and sufficiently predictable to be considered as the first option of surgical management. One eye had choroidal detachment with spontaneous resolution.

The debate on the need for posterior sclerotomy at the time of filtering surgery to prevent choroidal effusion and haemorrhage remains unresolved. Mandal (1999) showed no need of prophylactic posterior sclerotomy while Bellows et al (1979) have advocated it. In our patient, we had not performed prophylactic sclerotomy. Spontaneous resolution of effusion has been reported in the literature (Mandal, 1999; Bellows et al, 1979; Shihab et al, 1983).

Our patient developed hypotony and choroidal effusion in the post operative period, which resolved spontaneously with no visual deterioration, though the right eye required anterior chamber re-formation with ringer lactate solution.

Conclusion

Management of glaucoma associated with SWS is challenging. While planning surgical intervention an increased risk of choroidal effusion should be kept in mind and managed meticulously.

Figure 1: Erythematous well defined irregular shaped plaque present on both the sides of the face , right ear lobule and upper trunk, suggestive of bilateral Sturge-Weber syndrome

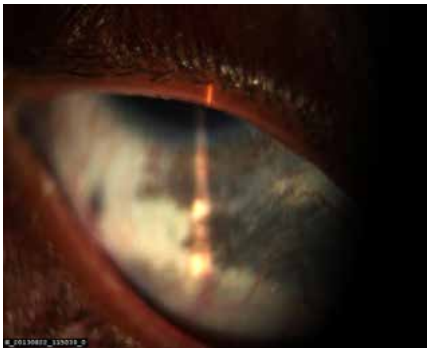

Figure 2: Sclera showed diffuse pigmentation with dilated tortuous episcleral vessels

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