International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

# Secondary Open-Angle Glaucoma in Sturge-Weber Syndrome - A Case Report

**Dr. Aishwarya Kumari[1], Dr Anuradha A[2], Dr Nishat Sultana Khayoom[3]**

[1]Junior Resident in MS Ophthalmology, Department of Ophthalmology, Minto Ophthalmic Hospital, Bangalore Medical College & Research Institute, India.
Email: *aishwarya.shcs[at]gmail.com*

[2]Assistant Professor, Department of Ophthalmology, Minto Ophthalmic Hospital, Bangalore Medical College & Research Institute, India.
Email: *anudanu[at]gmail.com*

[3]Professor, Department of Ophthalmology, Minto Ophthalmic Hospital, Bangalore Medical College & Research Institute, India.
Email: *nishatsultana29[at]gmail.com*

**Abstract:** ***Sturge-Weber Syndrome (SWS) is a rare neurocutaneous disorder characterized by facial capillary malformations, leptomeningeal angiomatosis, and ocular abnormalities, notably glaucoma. This case report describes a 16-year-old female who presented with progressive, painless vision loss in the left eye and a port-wine stain on the left side of her face. Ocular examination revealed episcleral vessel dilatation, elevated intraocular pressure (38 mm Hg), and glaucomatous optic nerve changes in the left eye, consistent with secondary open-angle glaucoma associated with SWS. Despite maximal medical therapy, intraocular pressure remained uncontrolled, and a trabeculectomy was performed, resulting in effective pressure reduction over a two-month follow-up period. This case underscores the importance of early recognition and multidisciplinary management of glaucoma in SWS to prevent irreversible vision loss and highlights the ocular clues that may point toward this neurocutaneous diagnosis.***

**Keywords:** Sturge-Weber Syndrome; neurocutaneous; port wine stain; glaucoma; trabeculectomy

## 1. Introduction

Sturge–Weber syndrome (SWS), a rare neurocutaneous phakomatosis, also known as encephalotrigeminal angiomatosis, is characterized by capillary-venous malformations affecting the brain and skin that may be linked to epilepsy and other forms of brain malformation. The classic presentation of Sturge–Weber syndrome includes hemangiomas of the choroid, congenital glaucoma, dural and leptomeningeal angiomatosis, which are more commonly found in the occipital and posterior parietal lobes, and unilateral facial nevus, which has a port-wine patch appearance (1).

Seizures, whether atonic, tonic, or myoclonic, are the most frequent neurologic manifestation of SWS and can occur at any age between birth and age 23. This syndrome is thought to affect 1 in 20,000 to 50,000 live births, while the exact incidence is unknown. There is no racial or gender preference in SWS (2-4).

Glaucoma is the most challenging ocular condition linked to SWS. Although late-onset glaucoma can sometimes develop in adulthood, glaucoma usually manifests unilaterally and is mainly identified during infancy (5,6). In this case report, we present a young adolescent girl with secondary open-angle glaucoma secondary to Sturge-Weber syndrome.

## 2. Methodology-Case Presentation

A 16-year-old female presented to the glaucoma clinic of a tertiary care hospital with complaints of gradual, painless, and progressive loss of vision in her left eye over the past four years. The patient had a history of persistent redness in the left eye since childhood and a pinkish discoloration (port-wine stain) on the left side of her face, specifically involving the forehead and temporozygomatic region, present since birth. She did not give any history of ocular pain, photophobia, watering, trauma, or previous eye surgeries. Additionally, she had a history of generalized tonic clonic epilepsy since the age of 10 years, for which she was being managed with tablet carbamazepine. There was no family history of similar ocular or dermatological complaints.

On general examination, well-defined pinkish patches were observed on the forehead and temporozygomatic area of the left side of the face, consistent with facial capillary malformation. Ocular examination revealed a best corrected visual acuity (BCVA) of 6/6 in the right eye and 6/12 in the left eye. The anterior segment evaluation of both eyes showed normal lids, conjunctiva, and clear cornea, with normal anterior chamber depth and reactive pupils measuring 3 mm. While the right eye exhibited normal sclera, the left eye showed episcleral vessel dilatation. Intraocular pressure (IOP) measured by applanation tonometry was 18 mm Hg in the right eye and significantly elevated at 38 mm Hg in the left eye. Gonioscopy revealed open angles with ciliary body band (CBB) visible in all quadrants bilaterally.

Fundus examination of the right eye showed a cup-disc ratio (CDR) of 0.2 with a healthy neuroretinal rim (NRR), and an artery-to-vein ratio of 2:3 with a positive foveal reflex. In contrast, the left eye displayed a vertically oval optic disc with a CDR of 0.6 horizontally and 0.7 vertically, along with inferior rim thinning—features suggestive of glaucomatous optic nerve damage. Optical coherence tomography (OCT) showed disc asymmetry, and visual field testing was unreliable due to high fixation losses in both eyes.

## 3. Results

A diagnosis of secondary open-angle glaucoma in the left eye was established in the context of Sturge-Weber Syndrome. Given the uncontrolled IOP despite maximal tolerated antiglaucoma medications, a trabeculectomy was performed in the left eye. During the two-month follow-up period, the IOP in the operated eye remained within the low teens, indicating a favorable short-term surgical outcome.

## 4. Discussion

In both our case and the study by Sultana et al. (7) describe classic presentations of Sturge-Weber Syndrome (SWS), yet they reflect different clinical priorities, where our study focused primarily on ocular complications, while the other emphasizes cutaneous and oral manifestations.

In our case, the patient, a 16-year-old girl, presented primarily with ocular symptoms—specifically, a gradual, painless loss of vision in the left eye over four years. The ocular examination revealed severely elevated intraocular pressure (IOP) of 38 mm Hg, optic nerve changes including inferior rim thinning, and episcleral vessel dilation, all leading to a diagnosis of secondary open-angle glaucoma, while Sultana et al. (7) reported a 12-year-old Bangladeshi girl whose chief complaint was port-wine stain (PWS) on the left side of her face and oral mucosa, noticed since birth and intensifying with age. Unlike our case, the primary concern in their case was cosmetic and mucosal involvement rather than vision impairment. However, Raval et al. (4) described a 31-year-old woman with a single generalized tonic-clonic seizure (GTCS) episode two days prior to admission, and a history of epilepsy since infancy. Unlike our case, the primary presenting complaint in their study was neurological, not ophthalmologic, although the patient had extensive PWS involving the right side of the face, limbs, trunk, and thorax. According to Nema N et al. (8), around 45% of patients with bilateral SWS had bilateral glaucoma.

Comparatively, in the Chaudhary et al. (6) case report, the patient was a 4-year-old girl presenting with severe bilateral diminution of vision, which had progressively worsened over the past year. Unlike our patient, their child had no history of seizures or neurological symptoms, although CT imaging did reveal subtle subcortical calcifications, suggesting underlying cerebral involvement typical of SWS and showed bilaterality and severity of ocular involvement. Their child had significantly enlarged corneas in both eyes (13–14 mm), high IOP (30 mm Hg bilaterally), closed angles on gonioscopy, and a very high cup-disc ratio (RE: 0.9, LE: 0.8) with marked neuroretinal rim thinning.

Neurologically, both cases had a history of seizures, reinforcing the common association of epilepsy with SWS due to underlying leptomeningeal angiomatosis. In our case, the patient had epilepsy since the age of 10 years and was on tablet carbamazepine. In the Sultana et al. (7) study, seizures were described as febrile convulsions in early childhood, managed conservatively with oral diazepam and physical cooling methods, without evidence of ongoing seizure disorder. This difference in seizure severity and control may reflect the variability in cerebral involvement typical of SWS. At the same time, in the Raval et al. (4) study, their patient had infantile-onset epilepsy at three months of age, with multiple breakthrough seizures due to irregular treatment, highlighting the chronic nature and risk of complications in inadequately managed cases. However, in the Chaudhary et al. (6) study, the girl had no clinical neurological symptoms, illustrating that neurological signs in SWS may not always be apparent despite structural brain changes.

Cutaneous findings in our case was consistent with port-wine stains in the distribution of the trigeminal nerve. Our patient exhibited a pink, well-defined patch over the forehead and temporozygomatic region, which matched the ophthalmic and maxillary divisions of the trigeminal nerve, while in the Raval et al. (4) study, their case involved larger and more extensive areas, including hypertrophy of soft tissues on the face and limbs, consistent with advanced vascular malformations. The Sultana et al. (7) case demonstrated a more extensive pattern, involving all three divisions of the trigeminal nerve and extending to bilateral hands and legs, indicating a more generalized vascular involvement. Notably, in Sultana et al. (7) case, the pigmentation was described as haphazard and asymmetrical, and while it mostly respected the midline, some regions showed crossover—a finding often seen in extensive angiomatosis, while Chaudhary et al. (6) observed body involvement in addition to facial port wine stains. According to Troilius et al. (9), 87–90% of the port wine stains are restricted to the right side of the face and fifty percent of patients have lesion extension over the median, and thirty-three percent have both sides afflicted, whereas Waelchli et al. (10) claim that rather than following the trigeminal nerve, the distribution of port wine stains may resemble the distribution of the face's embryonic vasculature.

Radiologically, Sultana et al. (7) reported no abnormalities on skull X-rays or other systemic investigations, and the patient was followed up conservatively due to the absence of complications. On the other hand, our case involved advanced ophthalmic diagnostics, including optical coherence tomography (OCT) and visual field assessment, culminating in trabeculectomy to manage the refractory glaucoma. This demonstrates a more interventional approach due to sight-threatening complications.

The goal of glaucoma treatment is to lower intraocular pressure and prevent further damage to the optic nerve and visual field, where topical antiglaucoma drugs are the first line of treatment for the late-onset type of the condition, but they are more challenging to treat in the congenital form due to their rarity (11). In terms of management strategy, Sultana et al. (7) patient was observed without active intervention, given the lack of systemic or ocular complications. Our patient, however, revealed secondary open-angle glaucoma, confirmed through comprehensive ophthalmic evaluation including fundus exam, OCT, and IOP measurement and underwent surgical management (trabeculectomy) after medical therapy failed to control the IOP, and the patient showed a good early postoperative response with IOP reduction. This contrast underscores the importance of tailored intervention based on the organ system most affected. However, in contrary to our findings, Sharan et al. (12) reported that surgical failure, uncontrolled IOP, and poor vision outcomes have been commonly recorded, where SWS

has the lowest surgical success rate among secondary glaucoma.

However, in the Raval et al. (4) study, despite having facial port wine stains involving the ophthalmic division, had no reported ocular complaints or findings and their patient was medically managed, advised to continue antiepileptics and consider laser photocoagulation for the PWS for cosmetic purposes. The absence of diminished vision, normal CNS examination, and lack of specific ophthalmic data suggest either minimal ocular involvement or that it was not a focus of clinical evaluation.

On the other hand, in the Chaudhary et al. (6) study, the patient was initially managed with topical medications, including dorzolamide and prostaglandin analogues, with a notable IOP reduction to 20 mm Hg. However, due to the extent of optic nerve damage and poor visual acuity (only finger counting at 2 meters bilaterally), surgical management was anticipated and discussed with the family for future planning. This indicates a more conservative initial approach in a younger child, perhaps to delay surgery while monitoring therapeutic response and ocular development.

## 5. Conclusion

Glaucoma associated with Sturge-Weber Syndrome presents a complex clinical challenge requiring a nuanced and multidisciplinary approach. Its multifactorial etiology, involving both structural angle anomalies and elevated episcleral venous pressure, necessitates individualized treatment planning. Early diagnosis, prompt intervention, and vigilant long-term follow-up are essential to prevent irreversible vision loss and preserve the patient's visual function and quality of life. This case highlights the importance of recognizing the ocular clues of systemic conditions such as SWS, as timely intervention can significantly influence prognosis.

**Conflicts of Interest**

The authors declare no conflicts of interest

## References

[1] Scott BK, Baranov D. Sturge-Weber syndrome. In: Roizen MF, Fleisher LA, editors. Anesthesia and Uncommon Diseases. 6th ed. Philadelphia: Elsevier; 2012. p. 251–295.
[2] Singh AK, Keenaghan M. Sturge-Weber Syndrome. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 [cited 2025 Jul 1]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK535405/
[3] Sujansky E, Conradi S. Sturge-Weber syndrome: age of onset of seizures and glaucoma and the prognosis for affected children. J Child Neurol. 1995;10(1):49–58.
[4] Raval DM, Rathod VM, Patel AB, Sharma B, Lukhi PD. Sturge-Weber Syndrome: A Rare Case Report. Cureus. 2022 Sep 5;14(9):e28786.
[5] Van der Hoeve J. The Doyne memorial lecture. Eye symptoms in phakomatoses. Trans Ophthalmol Soc UK. 1932;52:380–401.
[6] Chaudhary A, Garg S. Bilateral ocular manifestations of Sturge-Weber syndrome: a rare case report. Rom J Ophthalmol. 2024 Apr-Jun;68(2):173-176.
[7] Sultana W, Nasrin Rita S, Sadat SMA. Sturge-Weber syndrome – a rare congenital neuro-cutaneous disease. Updat Dent Coll J. 2015;5(2):47–51.
[8] Nema N, Jain J, Porwal V. A rare presentation of bilateral Sturge-Weber syndrome. Oman J Ophthalmol. 2014 Jan;7(1):46–48.
[9] Troilius A, Wrangsjö B, Ljunggren B. Potential psychological benefits from early treatment of port-wine stains in children. Br J Dermatol. 1998;139:59–65.
[10] Waelchli R, Aylett SE, Robinson K, Chong WK, Martinez AE, Kinsler VA. New vascular classification of port-wine stains: improving prediction of Sturge–Weber risk. Br J Dermatol. 2014;171(4):861–867.
[11] Basler L, Sowka J. Sturge-Weber syndrome and glaucoma. Optometry. 2011;82(5):306–309.
[12] Sharan S, Swamy B, Taranath DA, Jamieson R, Yu T, Wargon O, Grigg JR. Port-wine vascular malformations and glaucoma risk in Sturge-Weber syndrome. J Am Assoc Pediatr Ophthalmol Strabismus. 2009;13(4):374–378.

## Authors Profile

**Dr Aishwarya Kumari, MBBS,** Junior Resident in MS Ophthalmology, Department of Ophthalmology, Minto Ophthalmic Hospital, Regional Institute of Ophthalmology, Bangalore Medical College and Research Institute, Bengaluru, India. She completed her MBBS from AIIMS Patna with a gold medal in Microbiology. Her academic interests include glaucoma and neuro-ophthalmology, and she aims to contribute through research and government hospital service.

**Dr. Anuradha A, MBBS, MS (Ophthalmology),** Assistant Professor, Department of Ophthalmology, Minto Ophthalmic Hospital, Regional Institute of Ophthalmology, Bangalore Medical College and Research Institute (BMC&RI), Bengaluru, India. Her areas of interest include glaucoma and recent advances in ophthalmology, with a focus on integrating innovation into clinical practice and teaching. She has numerous national and international publications to her credit.

Professor Dr. Nishat Sultana Khayoom, MBBS, MS (Ophthalmology), F.GL (RGUHS), Professor, Head of Department, Department of Ophthalmology, Minto Ophthalmic Hospital, Regional Institute of Ophthalmology, Bangalore Medical College and Research Institute, Bengaluru, India. She is a senior academician and clinician with expertise in glaucoma and a strong commitment to teaching, clinical research, and comprehensive ophthalmic care. She is currently heading the Ophthalmology department in the prestigious Bangalore Medical College and Research Institute.

**Tables & Figures**

**Table 1:** Characteristics of Right and left eye of the patient

| | RIGHT EYE | LEFT EYE |
|---|---|---|
| BCVA | 6/6 | 6/12 |
| Lids | Normal | normal |
| Conjunctiva | Normal | normal |
| Sclera | Normal | Episcleral vessel dilatation |
| Cornea | clear | clear |
| Anterior chamber | Normal depth | Normal depth |
| Pupil | 3mm, RRR | 3mm, RRR |
| Lens | clear | clear |
| IOP( AT) | 18 mm Hg | 38 mm Hg |
| Gonioscopy | CBB CBB CBB CBB | CBB CBB CBB CBB |
| Fundus | CDR 0.2, NRR healthy, A;V=2:3, FR+ | CDR 0.6(H),0.7(V) Inferior rim thinning |

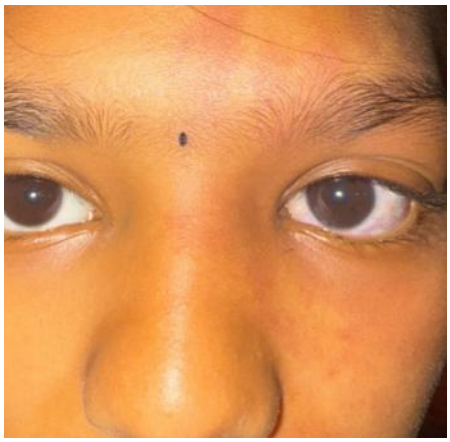
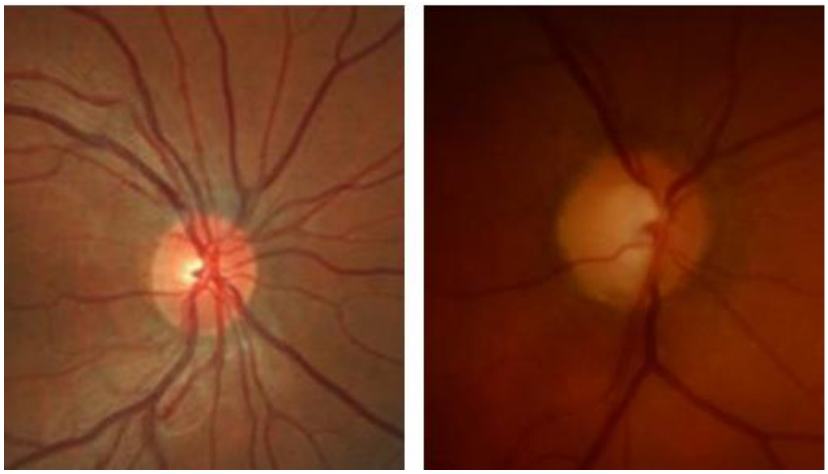


**Figure 1:** Well defined pinkish patches seen on forehead and temporozygomatic part of left side of face



**Right eye Fundus** **Left eye Fundus**

**Figure 2:** Fundus

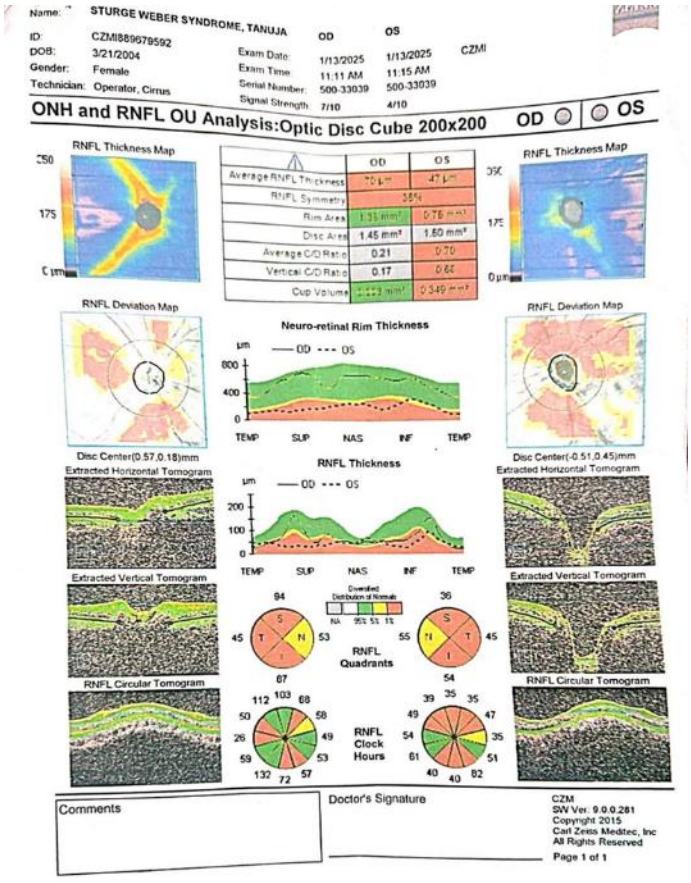



**Figure 3:** RNFL-OCT showing disc asymmetry

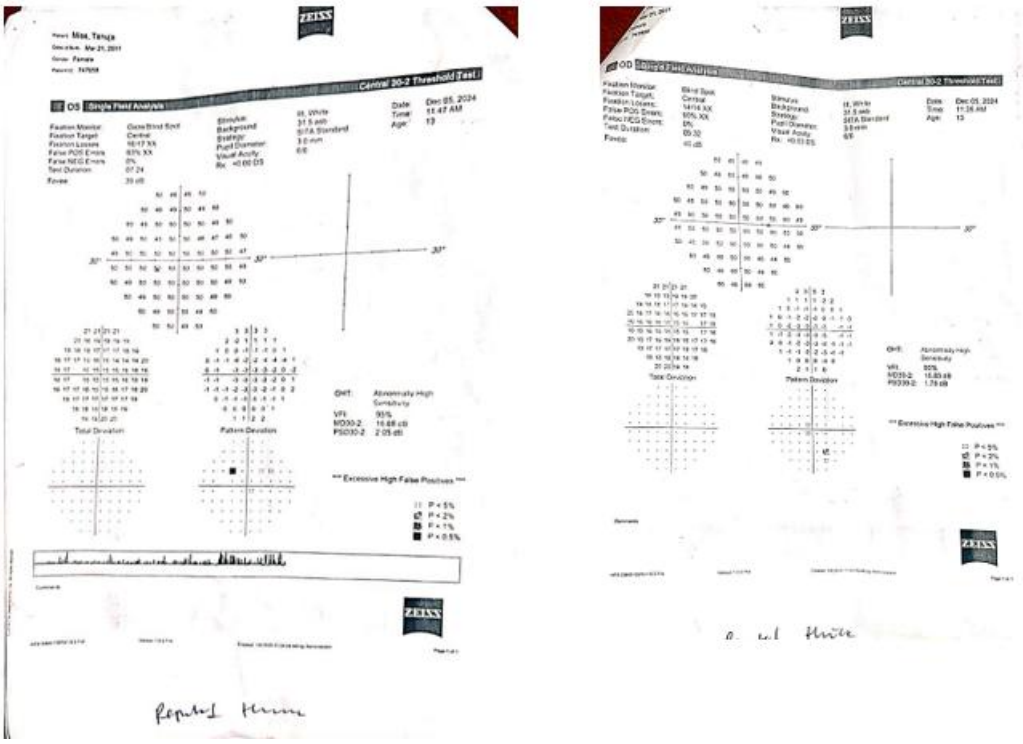


**Figure 4:** Unreliable visual fields due to high fixation losses in both eyes

**Before Surgery** **Post op day 1**

**Figure 5:** Anterior segment of LE before surgery and on post-op day 1