



Sturge Weber Syndrome with Secondary Glaucoma

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Abstract

We report a case of Sturge Weber syndrome (SWS) in a 6 month old child who presented with discolorations on the right half of the face, bigger size of right eye ball and seizures. Examination revealed port wine stain localized to the right half of face. Right eye was bigger in size; corneal diameter was 11.5 mm & 12.5mm vertical & horizontal respectively. Optic disc was small, vertically oval and cup to optic disc ratio was 0:4. Intra ocular pressure (IOP) was 22 mm Hg. Left eye was normal. CT scan revealed atrophic changes in hemi cortex with calcification in white and grey matter of all lobes of right cortex. Patient was put on topical anti glaucoma drops and was advised for trabeculectomy. At one year follow up the corneal diameter increased to 13 mm vertical and 14 mm horizontal. On optic disc evaluation, the cup to disc ratio was 0.6, Neuroretinal rim (NRR) thinning with inferior notching was noted. IOP was 18 mmHg. Parents did not give consent for glaucoma surgery. Patient was followed up to eight years of age.

Keywords: Sturge Weber Syndrome; Port Wine Stain; Glaucoma; Seizure; Hemiparesis

Introduction

Sturge Weber syndrome was first described by Sturge in 1879 and cerebral involvement was described by Kalischer in 1897 [1]. Sturge Weber syndrome (SWS), or Encephalo-trigeminal angiomatosis is a rare, congenital neurocutaneous syndrome characterized by unilateral facial cutaneous vascular malformation (nevus flammeus or port-wine stain) in association with ipsilateral leptomeningeal angiomatosis [2]. Angiomas of Sturge Weber Syndrome result due to failure of regression of a vascular plexus around cephalic portion of neural tube which is destined to become facial skin. This vascular plexus normally forms at 6th week of intrauterine life and regresses by 9th week. Failure of its regression results in residual vascular tissue which forms angiomas of leptomeninges, face and ipsilateral eye. These vascular plexus have abnormal blood flow pattern as vasomotor phenomenon, venous occlusion, thrombosis and "vascular steal phenomenon" resulting in ischemia, gliosis, atrophy and calcification of underlying cortical tissue [3]. The main ocular features are buphthalmos, haab's stria, and secondary glaucoma due to increased secretion of aqueous humor by Choroidal haemangioma or elevated episcleral venous pressure. The goal of treatment is to prevent optic nerve atrophy by controlling IOP by antiglaucoma drugs or by surgery. The diagnosis and management of Sturge Weber syndrome requires the combined skills of a radiologist, ophthalmologist, pediatrician and physiotherapist. Here we report a case of SWS with secondary glaucoma and seizures treated with topical antiglaucoma drugs and anticonvulsants. Patient was followed up to eight years.

Case Report

A 6 month old female child presented to eye OPD with chief complaints of discoloration on the right half of the face, intolerance to bright light, watering and bigger size of right eyeball. There was history of convulsions since 2 months, weakness in left half of body and delayed development. She was on anti convulsion therapy. Examination revealed port wine stain localized to the right half of the face along the ophthalmic and maxillary divisions of the trigeminal nerve. The right eye ball was bigger in size (Figure 1). Cornea was cloudy and she was intolerant to torch light. Left eye was normal. Examination of right eye under general anesthesia (EUA) revealed deep anterior chamber with haab's stria (Figure 2). Corneal diameter was 11.5 mm and 12.5mm vertical and horizontal respectively. Optic disc was small, vertically oval, and cup to disc ratio was 0.4:1. IOP in right eye was 22 and 14mmHg in left eye.

There was no evidence of choroidal haemangioma. CT scan revealed hyperdense echogenic areas on the right side of the cerebral hemisphere with calcification in white and grey matter of all lobes of right cortex (Figure 3). MRI brain revealed atrophic changes in right hemi cortex. Ipsilateral choroidal plexus was enlarged. Abnormal venous drainage with enlarged cortical veins due to shunting of blood was noted in left cortex (Figure 4). Patient was immediately put on topical Dorzolamide eye drops and advised for trabeculectomy. Patient came for follow up after one year. On examination of right eye- corneal diameter increased to 13mm and 14 mm vertical and horizontal respectively. Optic disc evaluation revealed cup to optic disc ratio of 0:6:1, thinning of neuroretinal rim with inferior notching was noted (Figure 5). IOP in RE was 18 mmHg. She was put on latanoprost 0.005% eye drop at night time in right eye. After 1 week IOP was 12 mmHg. The child was referred to pediatrician for management of seizures and to physiotherapist for hemiparesis. Patient was followed every six months. At every visit corneal diameter, IOP measurement and optic disc evaluation was done. Parents refused to give consent for surgery. Last follow up was at age of 8 years. Corneal diameter was constant. IOP was 14mmHg. Best corrected visual acuity in right eye was 6/18 and 6/6 in left eye.



Figure 1: Clinical photograph of the patient at 6 months, 1 year and 8 years of age

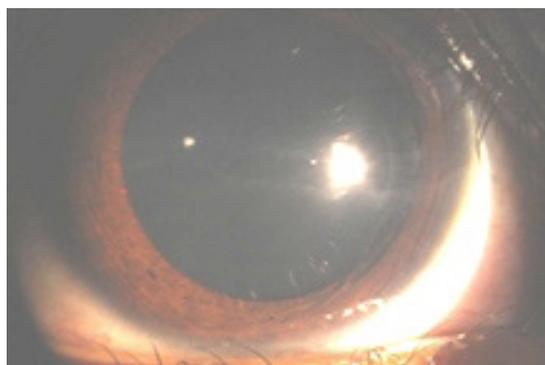


Figure 2: Photograph of corneal showing Haab's Stria

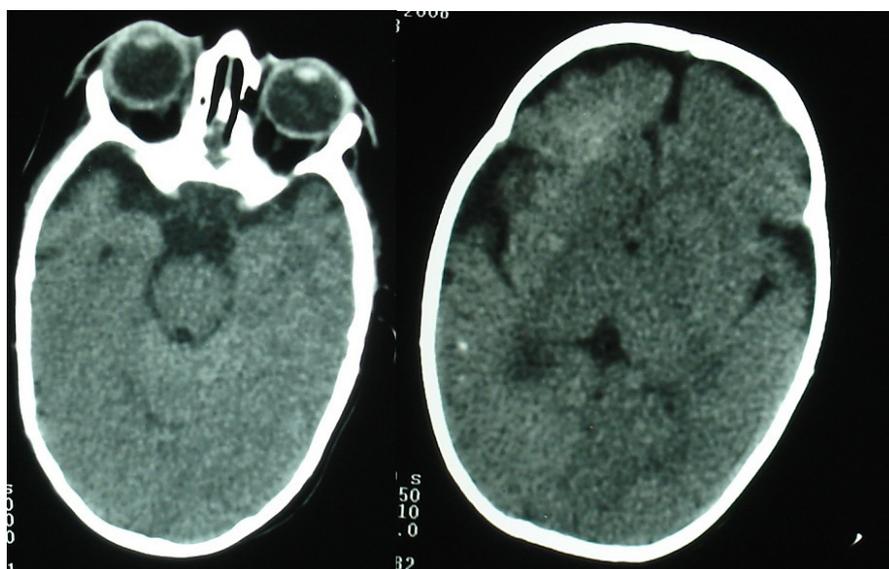


Figure 3: CT scan shows bigger size of right eye ball with calcification in right hemi cortex

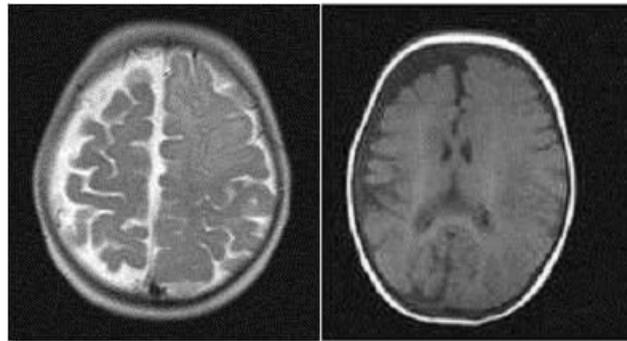


Figure 4: MRI brain shows atrophy of right hemi cortex

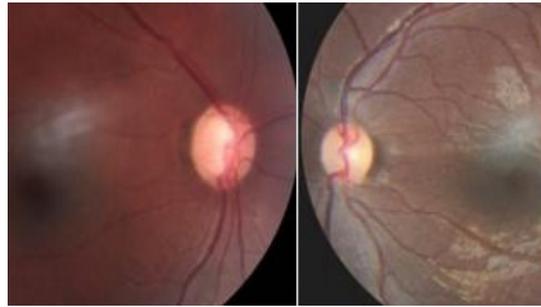


Figure 5: Optic disc photograph showing thinning of neuroretinal rim with inferior notching right eye and normal disc left eye

Discussion

SWS are a rare, sporadic condition involving vasculature in the facial skin, CNS, and eye. It belongs to a group of disorders known as phakomatoses. The incidence of this condition is approximately 1 per 50,000 live births. The chief ocular manifestation is glaucoma, in which developmental anomaly of the anterior chamber angle and elevated episcleral venous pressure are underlying mechanisms.

SWS are characterized by classical triad of Port Wine stain in the face, leptomeningeal capillary-venous malformation (angiomas) and ocular abnormalities. SWS are referred to as complete when both CNS and facial angiomas are present and incomplete when only one area is affected without the other. The Roach scale is used for classification [4].

Type I: Both facial and leptomeningeal angiomas; may have glaucoma

Type II: Facial angiomas alone; may have glaucoma

Type III: Isolated leptomeningeal angiomas; usually no glaucoma

In view of the presence of port wine stain in right side of face, glaucoma, seizures, hemiparesis, mental retardation and CT scan findings of calcification in cortex; this child was diagnosed as complete SWS / type 1 SWS. Treatment and prognosis depends upon the nature and severity of clinical features. Glaucoma occurs in 30 to 70 percent of individuals with SWS [5]. The clinical features resemble that of congenital buphthalmos. The incidence of glaucoma increases with port wine stain involvement of the ipsilateral eyelid. Weiss proposed that elevated episcleral venous pressure is the main mechanism underlying late-onset glaucoma [6]. Glaucoma associated with SWS is generally more difficult to manage than other forms of glaucoma, with a lower success rate and an increased risk of surgical complications. If topical medication does not produce an adequate response, glaucoma filtration surgery remains the procedure of choice. However, the success of this procedure has been variable in children. Trabeculectomy is associated with a much higher risk of choroidal effusions or suprachoroidal hemorrhage in eyes with SWS. Awad et al studied 15 SWS-glaucoma patients. Control of glaucoma was successfully achieved with medical treatment in 7 of 22 eyes. This was their initial treatment of choice. Of the 15 eyes that required surgery, 3 eyes developed persistent postoperative hypotony as late postoperative complications and lost vision [7].

Mandal recommended combined trabeculotomy-trabeculectomy in patients with early-onset glaucoma to address both of the underlying mechanisms and reported IOP of 16 mmHg or less without medications over a mean follow-up of 28 months in 10 eyes [8]. Glaucoma drainage implants have been shown to be effective in eyes with SWS. Budenz and colleagues reported successful outcomes using a two-stage Baerveldt glaucoma implant in 10 eyes with SWS. All eyes had IOP of 21 mmHg or less over a mean follow-up of 35 months without additional glaucoma surgery [5].

Seizure is a common feature which often occurs during the first year of life due to hypoxia, ischemia and gliosis of cortex [9]. Mental retardation is always associated with seizures. Presence of port wine stain can cause psychological trauma to patient and development of personality is affected in almost all patients. These can be treated by dermabrasion, flash lamp pulsed dye lasers and tattooing [10].

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