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Bilateral Sturge-Weber Syndrome and glaucoma controlled with Ahmed valve implant

Síndrome de Sturge-Weber e glaucoma bilateral controlado com implante de válvula de Ahmed

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ABSTRACT

Sturge-Weber Syndrome is a rare neuro-oculocutaneous disorder. The authors describe the case of a 13 years old boy, presented with bilateral Sturge-Weber Syndrome and glaucoma. Surgical treatment with Ahmed valve implantation in both eyes was carried out achieving lower levels of intraocular pressure.

Keywords: *Glaucoma; Glaucoma drainage implants; Sturge-Weber Syndrome; Port-wine stain; Case reports*

RESUMO

A síndrome de Sturge-Weber trata-se de uma doença neuro-óculocutânea rara. Os autores relatam o caso de um paciente do sexo masculino, de 13 anos de idade, que se apresentou com Síndrome de Sturge-Weber bilateral e glaucoma. Foi realizado o tratamento cirúrgico com implante de válvula de Ahmed em ambos os olhos e alcançado a redução da pressão intraocular bilateral.

Descritores: Glaucoma; Implantes para drenagem de glaucoma; Síndrome de Sturge-Weber; Mancha vinho do porto; Relatos de casos

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Introduction

Sturge-Weber Syndrome (SWS) is one of the systemic hamartomatoses (phakomatoses). It is a rare neuro-oculocutaneous disorder[1]. Clinically, the neurological component manifests as epilepsy, mental retardation, and hemiplegia; the ocular component manifests as glaucoma and vascular malformations of the conjunctiva, episclera, choroid, and retina ; and the dermal as the familiar nevus flameus, or port-wine stain[2].

Glaucoma is more common in SWS than in any other systemic hamartosis, but its precise incidence is unknown[3]. It occurs as often as in 50% of the cases when the port-wine stain involves the maxilar and ophthalmic divisions of the trigeminal nerve[4]. The results of therapy for glaucoma associated with the Sturge-Weber syndrome are often disappointing[5].

In this case report we describe a 13 year-old boy presented with bilateral SWS and glaucoma controlled after surgery with Ahmed implants.

Figure 1: Bilateral facial angioma presents in patient with Sturge-Weber Syndrome

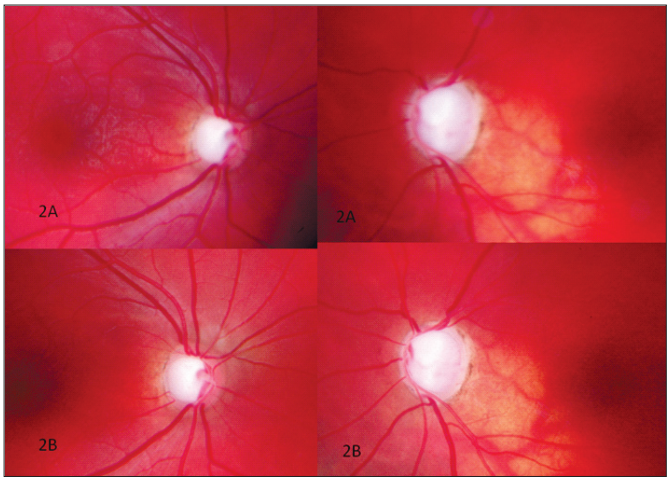

Figure 2: A – Cup to disk ratio of 0.6 X 0.6 in right eye and 0.9 X 0.9 in left eye in 2010; B – Cup to disk ratio in right and left eyes in 2012 without glaucomatous damage progression

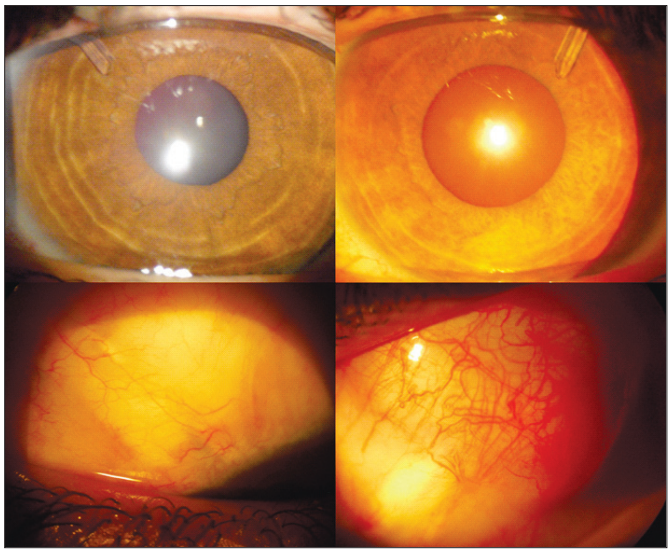

Figure 3: Biomicroscopy pictures showing Ahmed valve positioned over the temporal superior sclera in both eyes

Case report

A 13 year-old boy was refereed from the neurological department in 2010 for evaluation of Sturge-Weber Syndrome and low visual acuity. By this time he was already in use of Travoprost, Dorzolamide and Timolol Maleate ophthalmic solutions; and carbamazepine, clobazam and lamotrigine for the neurologic symptoms. He carried a bilateral facial angioma since birth (figure 1). Visual acuity was 1.0 in the right eye (OR) (refraction: -0.25 -0.50 X 15º) and 0.2 in the left eye (OS) (refraction: -3.50 -0.75 X 130º). Intraocular pressure was 22/24mmHg in OR and OS respectively at 09:30am. At gonioscopy, blood was observed in Schlemm's canal in OS. Scleral spur was seen in both eyes. Central corneal thickness was 612/600µm in OR and OS respectively. Fundoscopy revealed a cup to disk ratio of 0.6 X 0.6 in OR and 0.9 X 0.9 in OS (figure 2A).

Patient was submitted to surgical Ahmed valve implantation in both eyes. One month separated both procedures. Two years after the surgery, the patient presents intraocular pressure of 15mmHg at 10:00am in both eyes under fixed combination of dorzolamide and timolol maleate bid. Gonioscopy examination did not reveal blood in Schlemm's canal any longer. Visual acuity was still 1.0 in OR but dropped to count fingers at 1 meter in OS. There was no apparent progression between 2010 and 2012 of the cup to disk ratio as observed in retinographies (figure 2B). The Ahmed valves were implanted over the temporal superior sclera in a one stage procedure without any further complications (figure 3).

Discussion

Glaucoma associated with SWS is a rare and usually unilateral condition[4]. Here we describe a case of a bilateral disease with asymmetric optic disk damage.

The glaucoma's pathophysiology in SWS has been investigated for many years. Two mechanisms for glaucoma are theorized. In cases with buphthalmos and congenital glaucoma, the chamber angle is often anomalous, as in other types of congenital glaucoma. In later onset juvenile cases, the chamber angle more often appears normal. A premature aging of the

trabecular meshwork and the Schlemm's canal complex, as shown by Cibis et al. histopathologically, is a primary cause of juvenile glaucoma[6]. It is suggested that both mechanisms relate to the abnormal hemodynamics of episclera and chamber angle, due to persistence of Streeter's primordial vascular plexus[6]. More recent studies support the hypothesis that elevated episcleral venous pressure plays an important role in eyes with SWS glaucoma[7]. Frequently, patients with SWS and glaucoma have choroidal hemangiomas and/or episcleral or conjunctival hemangiomas[4], however this patient did not present any of these.

Clinical therapy for glaucoma in SWS if often ineffective and most patients require glaucoma surgery for a better intraocular pressure control. Trabeculectomy is associated with a high number of post-operative complications as choroidal effusion[8]. Other options for treatment described are goniotomy and trabeculotomy in early onset cases[4]. Van Emelen et al. had good results with cryocoagulation of the ciliary body combined with topical medication[9]. Some authors described less complications with glaucoma implants[1,10,11], which is why we have chosen to perform this type of procedure primarily.

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