ANGIOMATOUS CHOROIDAL AND ORBITAL LESIONS IN A PATIENT WITH STURGE WEBER SYNDROME

SÍNDROME DE STURGE WEBER: COMBINACIÓN DE LESIONES ANGIOMATOSAS COROIDEAS Y ORBITARIAS EN UN PACIENTE

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ABSTRACT

Case report: We present the case of a 34-year-old man with Sturge-Weber syndrome, who presented to the emergency room with left ocular pain and left chronic exophthalmos. He suffered an acute glaucoma secondary to pupillary block consequent upon an anterior lens subluxation. Orbital contrast Magnetic Resonance Imaging (MRI) was performed and he underwent intracapsular lens extraction.

Discussion: The MRI showed T2 enhanced lesions in the left meninges, choroid, and orbit, compatible with cavernous hemangiomas, as well as a dilated superior ophthalmic vein. Intraocular pressure after cataract surgery was 10 mm Hg, and visual acuity was less than 20/200 (Arch Soc Exp Oftalmol 2008; 83: 429-432).

Key words: Sturge-Weber syndrome, choroidal hemangioma, orbital hemangioma.

INTRODUCTION

Sturge-Weber syndrome is a rare, sporadic, and congenital disease. It belongs to the phacomatosis group, an association of skin and vascular injuries.

It is clinically characterized by ‘Port-wine’ stains on the trigeminal area, predominantly on the maxillary branch (88%), although the ophthalmic and/or mandibular branches may also be affected.

14% of occurrences are bilateral, with a higher prevalence of extrafacial involvement: leptomenyn-
geal angiomas on the parieto-occipital area, and ophthalmic alterations, mainly glaucoma (17%), diffuse choroidal angiomas, or localized choroidal angiomas with associated exudative retinal detachment, and more rarely orbital hemangiomas in extracranial locations.

Other less frequent clinical states include epilepsy (14%), and mental retardation (1).

**CASE REPORT**

34 year old male, diagnosed of Sturge-Weber syndrome during childhood. Acute glaucoma secondary to anterior lens luxation and pupillary block, so emergency surgery was performed.

Evidence was found under examination of a left hemifacial ‘Port-wine’ stain in the area of the first and second trigeminal branches, with severe exophthalmos and lower-temporal rejection of the eyeball, and a marked ‘Medusa head’ ingurgitation of lower episcleral vessels.

A mature cataract fully luxated to the anterior chamber was shown under examination with a slit lamp. The cataract was in contact with the corneal epithelium, with pupillary block and 60 mm Hg intraocular pressure (See Fig. 1 below). The ecography showed a retinal detachment in the posterior pole, together with an underlying hyperecogenic lesion.

Surgery was performed after an unsuccessful hypotensive medical treatment, with intracapsular removal of the lens. Ocular fundus confirmed suspicions of an upper nasal peripapilar choroidal injury, with an associated exudative retinal detachment.

The MRI study showed hyperintense T1 and T2 lesions in the leptomeninges of the left fronto-parietal region, with calcifications and atrophy of the frontal lobe. A choroidal lesion and two extra-conal orbital lesions were visible in the upper nasal quadrant (Fig. 2 below), and the upper temporal area (Fig. 3 below), both of them well delimited, and associated to dilation of the upper ophthalmic vein. All lesions were compatible with cavernous hemangiomas.

Intraocular pressure after surgery was maintained around 10 and 16 mm Hg, with no need for antihypertensive treatment, and final visual acuity under...
20/200 (bulk vision). Choroidal and orbital lesions are currently stable, so no treatment is necessary.

**DISCUSSION**

Ophthalmic manifestations are almost constant in Sturge-Weber syndrome. The most frequent ones are chronic glaucoma secondary to alteration in the venous return of the aqueous humour, but others have been described.

Choroidal hemangiomas are frequent amongst these, either localized or in a diffuse shape. The treatment of choice is laser therapy or external irradiation (2).

Orbital hemangiomas are rare. They are more frequently present as an extension to choroidal angiomas or to independent cavernous hemangiomas, as in our case. They may contribute to an increase of intraocular pressure, as ophthalmic veins are involved in their growth.

Other less frequent findings have been reported, such as posterior scleritis with uveal effusion leading to glaucoma from angular closure (3), juvenile ossifying orbit wall fibromas (4), etc.

The presence of an acute element in our case, with luxation of the lens into the anterior chamber is likely to be a consequence of the chronic inflammation process produced by the exudative retinal detachment due to zonular weaknesses, with no direct association with Sturge-Weber syndrome.

**REFERENCES**


