NEOVASCULAR GLAUCOMA ASSOCIATED WITH STURGE WEBER SYNDROME

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Abstract: *We present the case of a 84 years old Woman dignosed with Sturge Weber Sindrome and Neovascular Glaucoma. There are described the steps followed for diagnosis and treatment.*

Rezumat: **:** *Se prezintă cazul unei paciente în vîrstă de 84 ani diagnosticată cu Glaucom neovascular OD □i Sindrom Sturge Weber. Se vor descrie metodele de diagnostic □i tratament aplicate.*

INTRODUCTION

The Sturge-Weber syndrome (SWS), also called encephalotrigeminal angiomatosis, is a neurocutaneous disorder with angiomas involving the leptomeninges (leptomeningeal angiomas) and skin of the face, typically in the ophthalmic (V1) and maxillary (V2) distributions of the trigeminal nerve. The cutaneous angioma is called a port-wine stain (PWS). The main ocular manifestations (buphthalmos, glaucoma) occur secondary to increased IOP with mechanical obstruction of the angle of the eye, elevated episcleral venous pressure, or increased secretion of aqueous fluid.

SWS is 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, as follows :

1. Type I - Both facial and leptomeningeal angiomas; may have glaucoma
2. Type II - Facial angioma alone (no CNS involvement); may have glaucoma
3. Type III - Isolated leptomeningeal angiomas; usually no glaucoma

The incidence of SWS is estimated at 1 per 50,000. No sex predilection has been identified. The inheritance is sporadic.

CASE PRESENTATION

Patient IM, 83 years old, retired,

Reason for admission:

- pain in the right eye
- headache in the right half of the head
- decreased vision in both eyes, RE> LE
- photophobia

Disease History: the symptoms started relatively insidious, about 4 years ago with pain and decreased vision in RE, headache, mainly in the right half of the head and photophobia. The symptoms worsen with the time and were accompanied by decreased vision in LE.

The patient presents at birth in the right half of the face (initial eyelid region and the right third of the frontal region) a tumor, purple color, slightly elevated, painless, which increased in size with age.

Personal physiological history: menarche- at 13 years, 3 births, menopause-at 52 years

Past medical history:

- Right facial hemangioma in childhood
- Ischemic heart disease
- Hypertension
- Gastric ulcer

General treatment: Enalapril, Aspacardin, Omeprazole.

Family history – denies

Clinical examination: In the right half of the face presents a slightly elevated tumor, dark purple, which includes right third of frontal region, eyelid and zygomatic region, nasal and the right half of upper lip. BP = 130/70 (antihypertensive therapy)

General examination: normal range for age

Ophthalmological examination: Visual acuity: RE = light perception, LE = 1/3 with optic correction (-4dsf), intraocular pressure: RE= 70mmHg, LE = 17mmHg, corneal diameter: 11mm in both eyes

Figure no. 1. Hemangioma in the right hemifetei

Biomicroscopy:

RE superficial and deep conjunctival hyperaemia, prominent, tortuous conjunctival and episcleral vascular plexuses, corneal edema, shallow anterior chamber; Irian stroma invaded by neoformation vessels, especially in the pupillary edge, pupil in mydriasis, nuclear and cortical lens opacifying, reddish brown colored.

LE - normal aspect, smooth glossy transparent cornea; Iris stroma-normal for age, pupil centered round reflective; lens with cortico-nuclear opacifying.

Fundus examination:

- RE- can not distinguish retinal details

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- LE -flat papilla, discolored temporal, arteries with increased parietal reflex macula with pale reflex.

Stage diagnosis

1. RE Neovascular Glaucoma
2. Sturge Weber Syndrome, oculo-cutaneous form
3. LE Miopia
4. Cortico-nuclear cataract RE>LE
5. Hypertension
6. Ischemic cardiac disease
7. Gastric Ulcer

Gonioscopy. RE: camerular angle elements not visible; neovascularisation present in the iris root and pigment collar. LE: open angle, neovascularisation absent.

Visual field:

- RE- can not see the spot
- LE – normal for age

Ultrasound RE : linear opacity, with low ecogenity, mobile with the eye mouvements, disposed transversely in the middle vitreous

Laboratory results- normal for age

Internal medicine consultation: Hypertension, Ischemic cardiac disease, Gastritis post NSAIDs

Neurological consultation: Sturge Weber Syndrome; Rec. Gabaran 0.3 mg/day

MRI Skull: Some areas gliotic demyelinating suggesting vascular ethiology located in central white mass paraventricular bilateral. Small dilatations Wirchow-Robin spaces capsulo- lenticular bilateral. Mild cortical atrophy supratentorial. Right etmoido-maxillary sinusitis.

Consult ENT: Dg. Right maxillary rhinosinusitis. Cervical spondylosis with vertebro- basilar circulatory failure.

Positive diagnosis based on clinical and laboratory examinations:

- RE Neovascular Glaucoma
- Sturge Weber Syndrome oculo-cutaneous form
- Cortico-nuclear cataract OD>OS
- RE Posterior Vitreous Detachment
- LE Myopia
- Hypertension std. II cl. B
- Ischemic cardiac disease,
- Gastritis post NSAIDs
- Right maxillary rhinosinusitis
- Cervical spondylosis with vertebro-basilar circulatory failure

Positive diagnosis supporting

- OD increased intraocular pressure (70, 63 mmHg) -presence of neovascularisation in the iris stroma and pupil edge
- skin hemangioma wich involves V1 and V2 trigeminal nerve branches (ophthalmic and maxillary); hemangioma involves eyelid and bulbar conjunctiva OD.

Figure no. 2. The neoformation vessels iris

Differential Diagnosis:

Primary open angle glaucoma - excluded by clinical presentation, aspect of the LE fundus, LE visual field, gonioscopy.

Closed-angle glaucoma-primary attack / chronic – excluded by iris neovasculature, unilateral damage

Secondary glaucoma associated with: cataract, pigment dispersion syndrome, pseudoexfoliative glaucoma, glaucoma induced by steroid treatment.

Inflammatory glaucoma: Posner Schlossman Syndrome, Fuchs SDR.

Other secondary open-angle glaucomas: red cell glaucoma, ghost cell glaucoma, raised episcleral venous pressure, tumors. Schwarz Syndrome (anterior segment inflamation open angle, raised IOP arising from a rhegmatogenous retinal detachment).

Neovascular glaucoma of other causes:

- Ischemic CRVO or BRVO,
- Proliferative diabetic retinopathy,
- Sickle cell retinopathy,
- ACR obstruction,
- tumors of the retina or choroid.

Ocular ischemic syndrome in carotid arteries obstruction

Differential diagnosis of posterior vitreous detachment:

1. Retinal detachment
2. Choroidal detachment
3. Other vitreous opacities

Differential diagnosis of conjunctival hemangioma - with red-eye SDR of different etiologies (infectious, inflammatory)

Evolution. Without treatment – total and irreversible loss of vision in RE, due to NO atrophy

- eyeball perforation in case of very high IOP
- With treatment IOP stabilization and preserving visual function in case of response to treatment

Figure no. 3. Postoperative appearance

General Treatment: osmotic diuretics, carbonic anhydrase inhibitors, analgesics, NSAIDs, gastroprotective

Local treatment: beta blockers, corneal trophics, NSAIDs

Due to unfavorable evolution under conservative treatment is performed anti VEGF therapy in the anterior chamber followed by **surgical treatment**: trabeculectomy with peripheral iridectomy and use of antifibrotic (5FU)

There were no intraoperative complications

Postoperative evolution: increased IOP, sallow AC with uneven depth; bleb content- dark brown, high consistency (probably herniated iris, uvea blood clot)

Causes for high IOP and shallow AC post filtration surgery:

- Pupillary block
- Malignant Glaucoma
- filtration failure –obstruction of the sclerostomy and

scleral flap may be **internal** (incarceration of iris, ciliary processes or vitreous), **scleral** (fibrin, blood) or **external** (overly tight scleral flap suture)

- Suprachoroidal hemorrhage

Postoperator treatment

- mydriatic, anti-inflammatory steroids and drugs, bleb massage
- evolution was favorable, IOP decreased to 23-30 mmHg, corneal edema is reduced, remission of painful complains.

Prognostic

Good in case of bleb good function

Unfavorable in case of complications which will raise difficulties with diagnosing and treatment

Particularity of the case

- The rarity of this syndrome and its association with neovascular glaucoma
- Development of glaucoma in a very advanced age (most patients with Sturge Weber syndrome develop glaucoma in childhood)
- Early postoperative complication (obstruction of the sclerostomy) and favorable evolution after local treatment
- Difficulties raised by filtering surgery in cases of neovascular glaucoma

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