CASE REPORT

Sturge-Weber syndrome-associated glaucoma and intraocular osseous metaplasia: a unique complicated case

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SUMMARY

A 30-year-old European man was admitted to our centre complaining about severe pain of the right eye (OD) and right part of the face, redness and no vision of the OD. He had an 18-year history of secondary to Sturge-Weber syndrome glaucoma, 6-month history of red eye and 1-week history of pain in OD. The best-corrected visual acuity was no light perception OD and 20/20 OS. Intraocular pressure was 36 mm Hg OD. Examination revealed endophthalmitis, Sturge-Weber syndrome-associated glaucoma and complicated cataract of OD.

Unfortunately, no prescribed treatment helped this patient, so enucleation of OD was performed. During the enucleation, a subretinal 20 mm in width and 22 mm in length osseous tissue, partially vascularised, was removed surgically and was sent to the histological laboratory. Histopathologically, there were data of active inflammatory process, retinal detachment due to huge subretinal osseous metaplasia, gliosis and retinal pigment epithelial hyperplasia, and druses with ossification.

BACKGROUND

Sturge-Weber syndrome (SWS), also called encephalotrigeminal angiomas, is a neurocutaneous disorder with angiomas that involves the leptomeninges and the skin of the face, typically in the ophthalmic (V1) and maxillary (V2) distributions of the trigeminal nerve.¹ The incidence of SWS is ~1:50 000 infants.² According to the clinical manifestation, SWS is classified into four types: (1) presence of brain and facial angioma, with or without glaucoma; (2) port-wine stains (PWS) without brain involvement, with or without glaucoma; (3) isolated brain angioma, usually without glaucoma; and (4) type 1 associated with systemic manifestation such as tuberous sclerosis.³

Ocular findings are noticed in approximately 50% of patients with SWS.⁴ Ipsilateral to PWS glaucoma develops in 30%–70% of patients with SWS.^{5 6} The pathogenesis of glaucoma is complex and includes congenital malformation of the anterior chamber angle that increases resistance to aqueous humour outflow, increased episcleral venous pressure, increased aqueous humour production by either the ciliary body or the choroidal haemangioma and abnormal haemodynamics of the episclera and the anterior chamber

angle due to premature ageing of the trabecular meshwork–Schlemm's canal complex.⁴

CASE PRESENTATION

A 30-year-old European man was admitted to our centre complaining about severe pain of the right eye (OD) and right part of the face, redness and no vision of the OD. Family history is essentially negative. The beginning of the disease started with a gradual reduction of vision OD, which was noticed since he was 7 years old. When he was 10, he was examined in another ophthalmological clinic. At that time, the best-corrected visual acuity (BCVA) of the OD was 20/2000, and BCVA of the left eye (OS) was 20/20. Intraocular pressure (IOP) was OD 19 mm Hg and OS 18 mm Hg. Also, there was a visual field defect on periphery of OD. Ultrasonography revealed secondary exudative retinal detachment of the OD. The diagnosis was following 'secondary exudative retinal detachment, chorioretinitis, peripheral retinal degeneration of OD, SWS type 2'. The required therapy was prescribed. When the patient was 12, glaucoma of OD was diagnosed and antiglaucoma drops were prescribed. When he was 19, the examination revealed the BCVA was no light perception OD and 20/20 OS, IOP OD 48 mm Hg, OS 16 mm Hg. Antiglaucoma drops were prescribed, and almost annually, he visited his ophthalmologist.

On admission to our centre, the patient was complaining about severe pain of the OD and right part of the face, redness and no vision of the OD. He had a 6-month history of red eye and 1-week history of pain in OD. During this period, he visited his ophthalmologist and was taking the administered drugs. The BCVA was no light perception OD and 20/20 OS, IOP OD 36 mm Hg, OS 18 mm Hg. There was an angioma of the right half of the face, also expressed on the eyelids. There was significant pus, mixed injection and winding vessels of conjunctiva more especially in the upper arch. The cornea was dim and swollen; there was a 4–5 mm hypopyon. The lens was diffusely cloudy. The fundus reflex was missing. The left eye was normal except for peripheral retinal degeneration. The diagnosis was 'endophthalmitis, Sturge-Weber syndrome-associated glaucoma with decompensated IOP, complicated cataract OD'.

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Figure 1 A 30-year-old man with Sturge-Weber syndrome type 2 after evisceration of OD. There is a postoperative oedema of the eyelids and a cutaneous angioma on the right part of the face.

TREATMENT

The system treatment included ceftriaxone, metronidazole, dexamethasone, dexketoprofen and meloxicam. Paraocular injections with dexamethasone and gentamicin were performed. Topical treatment included levofloxacin, dexamethasone, diclofenac, latanoprost, timolol and brinzolamide.

Figure 2 After some time. The patient with ocular prosthesis inserted in OD.

Figure 3 A 20 mm in width and 22 mm in length osseous tissue, partially vascularised, that was removed during the evisceration.

OUTCOME AND FOLLOW-UP

After 4 days, due to the ineffectiveness of treatment, enlargement of hypopyon and impossibility to compensate for high IOP, evisceration of the right eye was performed (figure 1). After some time, ocular prosthesis was inserted (figure 2). During the evisceration, a subretinal 20 mm in width and 22 mm in length osseous tissue, partially vascularised, was removed surgically and was sent to the histological laboratory (figure 3). Histopathologically, there were data of active inflammatory process, retinal detachment due to huge subretinal osseous metaplasia, gliosis and retinal pigment epithelial hyperplasia, and druses with ossification.

DISCUSSION

According to the literature,⁷ an antiglaucoma surgery should have been performed after it became clear that IOP level was not controlled by medication. In adults with SWS-associated glaucoma, trabeculectomy, non-penetrating sclerectomy, valve drainage implants, ciliodesstructive procedures and Ahmed-type valves can be performed.⁴ Probably, if the antiglaucoma surgery was performed in time, BCVA was higher and the necessity of evisceration would be prevented afterwards. The histopathological finding was a testimony of the durable inflammatory process (endophthalmitis) leading to a subretinal osseous metaplasia.⁸ Intraocular osseous metaplasia is associated with durable chronic inflammation, trauma, phthisis bulbi and long-standing retinal detachment.⁸ In the previously described clinical case, a durable inflammatory process could mediate the pluripotential retinal pigment epithelial cells to transform into osteoblastic precursor cells.⁹ Unfortunately, we still do not know the reason why the severe uncontrolled inflammation occurred.

Take home message

- ▶ Glaucoma monitoring must be obligated in patients with Sturge-Weber syndrome (SWS). Usually, in order to control intraocular pressure, an antiglaucoma surgery along with antiglaucoma drops is recommended for SWS-associated glaucoma.
- ▶ We consider a durable severe intraocular infection with a subretinal osseous metaplasia by transformation of pluripotential retinal pigment epithelial cells into osteoblastic precursor cells.

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