CLINICAL CASE**SERPIGINOUS CHOROIDITIS – A RARE CAUSE OF NEOVASCULAR GLAUCOMA****Lavinia Iftene¹, Alexandra Nicolae¹, Patricia Badila¹, R Nanu¹, Emil Ungureanu^{1,2}**¹The University of Medicine and Pharmacy “Carol Davila” Bucharest, Romania²The Clinic of Ophthalmology, The University Emergency Hospital of Bucharest, Romania

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Abstract

Neovascular glaucoma (NVG) is considered to be one of the rarest and most complex types of ophthalmologic diseases known up to the present moment, and is mainly caused by the obstruction of the iridocorneal angle through a neovascular membrane. We present the case of the patient MC, aged 46, who came to the Ophthalmologic emergency room of the University Emergency Hospital of Bucharest (SUUB), complaining of red, painful left eye (OS) as well as a severe decrease of the visual acuity (VA) in the left eye (OS). In the patient's medical record it is noted a surgical procedure for cataract which he underwent in another clinic 2 months prior to the current presentation. Systemically hypotensor treatment was instituted, topically an intravitreal Bevacizumab injection followed by scleral flap trabeculectomy in the left eye (OS). As a result, the evolution was favorable including a decrease in the intraocular pressure. The patient is discharged, with immunosuppressive therapy for serpiginous choroiditis according to the rheumatologic findings (Cyclosporine). Serpiginous choroiditis is an inflammatory lesion of unknown etiology part of the White Dot Syndromes (WDS), probably the most severe form of them. It is a very rare condition, and in the cases of patients with serpiginous choroiditis the frequency of the choroidal neovascularisation is reduced and the neovascular glaucoma is exceptional. The surgical treatment of the neovascular glaucoma has poorer long-term prognosis compared to other forms of glaucoma. The peculiarity of the case comes both from the rare cause of the neovascular glaucoma, specifically the serpiginous choroiditis, also a rare autoimmune disorder, and the need for clinical cooperation between the ophthalmologist and immunologist or a rheumatologist in order to establish the adequate treatment and follow-up.

Keywords: *serpiginous choroiditis, glaucoma***Introduction**

Neovascular glaucoma (NVG) is considered to be one of the rarest and most complex types of ophthalmologic diseases known up to the present moment, and is mainly caused by the obstruction of the iridocorneal angle through a neovascular membrane. The cause is a retinal ischemia which triggers the formation of VEGF

(vascular endothelial growth factor) leading to subsequent neovascularization in retina and iris, especially at the level of the iridocorneal angle. Thus, a neovascular membrane is formed which obstructs the angle and does not permit the filtration of the aqueous humour with subsequent increased intraocular pressure. This will eventually lead to the progressive destruction of the optic nerve.

The frequent causes leading to retinal ischemia are: diabetic retinopathy, especially in severe stages, retinal vascular occlusions, particularly the venous occlusions [1] and the ocular ischemic syndrome [2]. There are multiple rare causes leading to neovascular glaucoma, one of them being ischemia caused by chorioretinal inflammatory disorders, such as the serpiginous choroiditis in our case, an extremely rare, chronic, pathology recognized as pertaining to the family of White Dot Syndromes (WDS). Throughout this presentation we seek to familiarize the reader with the defining aspects of NVG, with its treatment and the one for the serpiginous choroiditis, so that at the end of the paper we could focus on the patient's peculiarities.

As a pathophysiological mechanism, the neovascular glaucoma includes the following phenomena: normally, the aqueous fluid is continuously secreted by the epithelium of the ciliary body into the posterior chamber of the eyeball, to escape through the pupil into the anterior chamber. From here, it is drained towards the iridocorneal angle, namely to the trabecular meshwork which continues with the Schlemm's canal and finally it continues with the episcleral venous system. In pathological conditions in general and in NVG in particular, this angle will be obstructed by the neovascularization processes that extend from the minor arterial circle of iris to the major arterial circle of iris, thus blocking the camerular angle and continuously increasing the IOP. The neovascularization is caused in our case by the serpiginous choroiditis, a choroidoretinopathy of unknown etiology, which affects the capillaries of the choroid, the retinal pigment epithelium and by adjacency the retina with the occurrence of the VA defects. This begins with subretinian infiltrations in the peripapillary area that irregularly proliferates, which later will lead to retinal ischemia, phenomenon equivalent to the occurrence of tissular hypoxia that in its turn will trigger the excessive synthesis of VEGF, the main cause of neovascularization.

Materials and methods

Patient MC, aged 46, came to the emergency room of the University Emergency Hospital of Bucharest (SUUB), for the following complaints: red, painful left eye (OS), decrease of the visual acuity (VA) in the left eye (OS).

In the patient's medical record we note a surgical procedure for cataract which he underwent in another clinic 2 months prior to the current presentation. Subsequently he stated that his sight gradually improved for about 10 days, after which he reported a sudden decrease of his visual acuity, when he came to the doctor – he was hospitalized for a few days following a treatment with Tarosin, Piracetam, Ederen 1tb/day, Cosopt. When discharged, the patient was advised to take Tobradex, Indocollyre, Piracetam, Cosopt. He was reassessed after a week, with an uncertain improvement of the symptoms. The patient was reassessed then after another 2 weeks, when he was referred to SUUB.

The physical examination showed the increase of intraocular pressure for the OS: VAOD = 2/3 ccp VAOS = 1/50 fc cd, TOD = 17 mm Hg, TOS = 30 mm Hg.

The slit-lamp examination reveals OD: a lens presenting predominant nuclear clouding and as for the OS: epithelial corneal and stromal edema; iris with rubeosis iridis, well-centered PC pseudophakia.

FOD presents specific changes for serpiginous choroiditis, as for the FOS it was established with difficulties due to the corneal edema, subsequently an aspect similar to that of the OD. The diagnosis was: OS Neovascular Glaucoma, OU serpiginous choroiditis, PC pseudophakia.

Figure 1 - Right-eye with the specific serpiginous choroiditis aspect (authors' personal collection)

Figure 2 - Left Eye serpiginous choroiditis – image taken subsequently to the initial assessment, after treatment (authors' personal collection)

Treatment

A general ocular hypotensor treatment consisting of carbonic anhydrase inhibitors - Acetazolamide 3 tb/day along with Aspacardin 3 tb/day (because Acetazolamide depletes potassium in the kidney), Mannitol 250 ml EVP x 2/day, topically a beta-blocker 1 drop x 2/day, carbonic anhydrase inhibitor 1 drop x 5/day, corticosteroid 1 drop x 3/day, mydriatic 1 drop x 3/day is instituted. The evolution is slow but favorable, with a decrease of the intraocular pressure OS (22 mm Hg) and partial reducing of the corneal edema. The gonioscopy highlights in the OS neovascular membrane in the iridocorneal angle. At the FO examination it is found out that the OS does not permit the laser retinal photocoagulation because of the remaining corneal edema and an intravitreal Bevacizumab injection is administered to OS. It appears that the rubeosis iridis disappeared and one week after the injection, for OS, scleral flap trabeculectomy is performed.

Figure 3 - Left Eye – Aspect after a week postoperatively. (authors' personal collection)

The evolution was favorable, with a decrease in the intraocular pressure (TOS = 14 mm Hg – under the strict combination of betablocker and carbonic anhydrase inhibitors 1 drop x 2/day). The patient is discharged with immunosuppressive treatment for serpiginous choroiditis according to the treatment prescribed by the rheumatologist (Cyclosporine). One month, postoperatively VAOS = 1/4 fc, TOS = 14 mm Hg.

Evolution and prognosis

The prognosis of the intraocular tension is good on short and medium-term. On long-term the prognosis is poor due to the probability of fibrosis occurrence, the decrease in trabeculectomy efficiency and the subsequent increase of the intraocular pressure, all of them requiring further interventions. [3]

Discussions

Serpiginous choroiditis is an inflammatory lesion of unknown etiology part of the White Dot Syndromes (WDS), probably the most severe form of them. [4]. It is considered that these syndromes have in common the fact that the retinal pigmentary epithelium as well as the external retina present great endurance to a massive reduction of the vascular flow, more than the capillaries of the choroid, and the differences will be provided by expansion, level (arteriolar, precapillary), reversibility of the vasoocclusive processes, which will also determine the angiographic and clinical aspect of the disease. Thus, in the case of serpiginous choroiditis, medium - irreversible irregular lesions of the arterioles appear, with low responsiveness to treatment. [5]

The major causes of neovascular glaucoma, according to frequency are: diabetic retinopathy in diabetes mellitus, especially unbalanced and retinal vascular occlusions, especially the venous occlusions. The cause is the retinal ischemia, which determines the VEGF synthesis with the occurrence of subsequent neovascularization. Serpiginous choroiditis is a rare disease, and in the case of the patients suffering from serpiginous choroiditis, the frequency of choroidal neovascularization is reduced (6-12 %) [6], and the neovascular glaucoma is exceptional.

The surgical treatment of the neovascular glaucoma has poorer long-term prognosis compared to other forms of glaucoma. The most popular options considered are scleral flap trabeculectomy with 5-FU or mitomycin and respectively Ahmed Valve Implant. Both procedures are commonly performed after intravitreal injection with VEGF inhibitors which provides a temporary regression of the neovascularization and thus provides a therapeutic range needed for the surgical procedure to be free of as many adverse effects as possible - especially hemorrhage and rapid fibrosis with closing of the filtration flap. [7].

The surgical procedure consists of the excision of the deep part of the sclerocorneal junction, including the Schlemm canal, followed by iridectomy, in order to free the iridocorneal angle.

For teaching purposes, the surgical procedure can be divided into the following steps:

1. Topical (peribulbar / subtenonian) anesthesia. This can be performed by setting the eyeball with the help of a thread at the superior rectus muscle or at the level of the sclerocorneal limbus.

2. Conjunctiva detachment. This has been performed through an incision at the sclerocorneal limbus/fornix. Simultaneously the maintenance of moderate hemostasis by electrocautery is sought.

3. Detachment of the scleral flap (which may be triangular or trapezoidal according to the surgeon's preferences). This consists of the detachment of a flap of 5x5, 1/3 or 1/2 from the scleral thickness.

4. Trabeculectomy. Meanwhile concurrently, anticoagulants are also administered, such as Mitomycin C, 5-FU – an antimetabolite used for the prevention of fibrosis.

5. Iridectomy. Its role is not to allow the trabeculectomy blockage by subsequent iris synechiae and to allow the decifluz aqueous humor to the subconjunctival filtration flap.

6. The scleral flap suturing may be performed with one or two separate threads (10-0).

7. The conjunctiva suturing is performed where the incision and conjunctiva detachment were made, either in the fornix or in the sclerocorneal limbus. It's usually a Surjet suture (simple continuous suture), the thread used being removed one month postoperatively.

Conclusions

- Serpiginous choroiditis is an inflammatory lesion of unknown etiology part of the White Dot Syndromes (WDS), probably the most severe form of them.

- Serpiginous choroiditis is a very rare condition, and in the cases of patients with this pathology, the frequency of the choroidal neovascularisation is reduced.

- The neovascular glaucoma is exceptional. Its surgical treatment has a poorer long-term prognosis compared to other forms of glaucoma. The most popular options considered are scleral flap trabeculectomy with 5-FU or mitomycin and respectively Ahmed Valve Implant. Both procedures must be performed after intravitreal injection with a VEGF inhibitor which provides a temporary regression of the neovascularization and thus provides the therapeutic range needed for the surgical procedure to be free of as many adverse effects as possible

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The particularity of the case comes both from the rare cause of the neovascular glaucoma, specifically serpiginous choroiditis, also a rare autoimmune disorder, and the need for clinical cooperation between the ophthalmologist and the immunologist or a rheumatologist in order to establish the adequate treatment and follow-up.

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