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Short Communication

Sympathetic Ophthalmia is a Non-Disappearing Disease in the XXI Century

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Abstract

New technologies in ophthalmomicrosurgery in developed countries have led to a change in the epidemiology of sympathetic ophthalmia (SO) and vitreoretinal surgery on damaged eyes is becoming a risk factor. At the present stage of the 21st century, SO is not disappearing disease, its frequency remains at the level of the end of the twentieth century, amounting to 0.3 - 0.9% in adults and 0.24% in children. The authors draw attention to the modern understanding of SO as a multifactorial autoimmune disease. The clinical features of the frequent occurrence of sympathetic panuveitis, the difficulties of diagnosis and the need to use multimodal imaging - OCT, A-OCT, FFA and B-scan ultrasonography.

Keywords: *Sympathetic Ophthalmia; Vitreoretinal Surgery; Multifactorial Autoimmune Disease*

Sympathetic ophthalmia (SO) is a bilateral autoimmune granulomatous panuveitis (uveitis) that occurs after a complicated penetrating injury or intraocular surgery of one, rarely both eyes.

The unusual nature of this disease, when the second eye is involved in the process only because the first one is damaged and inflamed, the complexity and lack of clarity of its etiopathogenesis, the tragedy of vision loss in one or both eyes, forces scientists to actively study this problem, starting from the XIX century, when the Scottish ophthalmic surgeon W. Mackenzie in 1840 described in detail this disease, known since the time of Hippocrates, suggested the term "ophthalmia sympathica", which has since become firmly established in ophthalmology.

The etiopathogenesis of SO is unclear. According to modern concepts, sympathetic ophthalmia is a multifactorial autoimmune disease of both eyes that occurs as a result of a T-cell immune response to autoantigens of the uveal tissue, pigmented epithelium and retina [1-8].

Sympathetic ophthalmia occurs as a result of "disruption" of the immune privilege of the eyes in cases of blinded penetrating trauma (injury, surgery), a combination of clinical risk factors and immunoregulatory disorders in genetically predisposed individuals.

Sympathetic ophthalmia is a rare condition. By the beginning of the twentieth century, the incidence of SO was 2 - 3% after penetrating wounds and 0.02 - 0.2% after intraocular surgery. Analysis of the incidence of SO in different periods of the twentieth century showed that the improvement of surgical and medical treatment of penetrating trauma led to a decrease in the frequency of SO by 8 - 10 times and by the end of the twentieth century it was 0.2 - 0.4% after penetrating wounds and 0.01 - 0.05% after surgery [1].

At the turn of the century and at the beginning of the XXI century, the epidemiology of SO is changing in developed countries. New technologies in ophthalmomicrosurgery have led to almost the disappearance of SO after classical intraocular operations, and some ophthalmic surgeons began to say that SO - disappearing disease. This contributed to a decrease in alertness to the possibility of occurrence and to the late diagnosis of SO.

On the other hand, new technologies and increased surgical activity of ophthalmic surgeons have led to an increase in SO after repeated, multiple operations, more often after vitreoretinal surgery on damaged eyes, performed both during primary surgical treatment and during surgical rehabilitation of damaged eyes [9-13].

Based on a survey of 876 ophthalmologists in England and 85 ophthalmologists in Scotland, D. Kilmartin., *et al*. [10] believe that retinal re - surgery is a risk factor for SO with a frequency of 1 case per 799 operations. Among the newly identified 18 cases of SO that occurred in the period from 1997 to 1998, 11 (61.1%) developed after vitreoretinal surgery and 9 of them after multiple vitreoretinal surgery.

In a retrospective analysis of 10 cases of SO observed from 1993 to 2003 at the National Eye Center of Singapore, 9 patients with SO developed after repeated operations on previously operated [7] or injured eyes [2], including 6 after vitreoretinal surgery [12].

We believe that from an immunological point of view, repeated operations lead to repeated sequestration of uveoretinal autoantigens and the most immunogenic retinal S-antigen, a decrease in immunosuppressive protective factors in the anterior chamber, vitreous body and subretinal fluid, and activation of a secondary immune response in the presence of T cells of immunological memory in the body.

In general, in the XXI century, the frequency of SO does not tend to decrease and is at the level of the end of the XX century. It is 0.3 - 0.9% in adults after penetrating wounds [13] and 0.24% in children [14].

In the work of EA Drozdova., *et al*. [13] showed that in the period from 1994 to 2014, among 4751 patients with open trauma, SO occurred in 22, which was 0.46%. Moreover, the authors note that in 2012 it increased to 1.86% (3 cases per 161), and in 2013 to 1.99% (3 cases per 151) after a domestic criminal injury with subconjunctival ruptures untreated for several weeks.

We studied the clinical and epidemiological features of SO in 25 patients observed in the period from 2000 to 2021. Of these patients 16 (64%) had SO after repeated intraocular surgery, mainly after repeated vitreoretinal surgery [14] performed on injured [11] or operated [3] eyes. The remaining 9 patients (36%) developed SO after penetrating wounds. We did not observe any cases of SO after single operations, including cataract extraction and anti-glaucomatous operations.

The terms of SO occurrence after injury were different - from 2 - 3 weeks to a year -21 people (84%). It should be noted that SO began to occur more often not in the first 2 - 3 months, as previously, but later-in 4 - 6 months-13 people (52%). This can be explained by the more frequent implementation of repeated vitreoretinal surgery on injured or operated eyes during these periods. We also observed 4 patients (16%) who developed SO at a late stage (from 1.5 to 18 years after repeated vitreoretinal surgery).

EA Drozdova., *et al*. [13] note a more significant incidence of late SO (63.6%) when it occurs from 1 to 45 years after injury, which the authors explain by performing repeated surgery a later time during surgical rehabilitation of damaged eyes.

Clinically, most patients were diagnosed with panuveitis-21 people. (84%), rarely isolated anterior uveitis - 3 people (12%) and posterior uveitis - 1 person (4%). The number of clinical features of SO that occurs after repeated vitreoretinal surgery are noted. In these patients, the greatest changes in the initial stage occurred in the posterior part - diffuse choroiditis, papillitis and serous retinal detachment.

In the early stages of SO on OCT, thickening and folds of the choroid, edema of the optic nerve disc, macular edema, detachment of the neuroepithelium and areas of formation of inflammatory granulomas can be detected. Ophthalmoscopically, these changes may not be visible, so it is very important to perform an OCT scan in patients with anterior uveitis for early diagnosis of its transition to panuveitis.

At the same time, the first complaint of patients was not a decrease in visual acuity and signs of uveal inflammation in the anterior part, but a deterioration in vision in the dark, which patients initially did not pay attention to and turned to the ophthalmologist late, which led to a late diagnosis of SO.

Traditionally, the diagnosis of SO is mainly based on the history of injury and clinical features of both eyes. Frequent involvement of the posterior parts of the uveal tract, retina, and optic nerve in the process necessitates a complex of early and differential diagnostics with multimodal imaging [16-18].

At the present stage of multimodal visualization of the paired eye in vocational training helps in the early preclinical diagnosis of CO, allows you to track the progression of the disease, detect relapses and monitor the response to therapy. New OCT and A-OCT devices have become useful non-invasive tools for assessing the microstructure of the retinochoroid and microvessels in patients with SO, and their use is becoming increasingly important. It is important to use these techniques as early as possible in patients with suspected SO.

Thus, the data of recent years do not allow us to attribute SO to disappearing diseases, as some ophthalmic surgeons believe.

While eye injuries occur with damage to antigen-containing uveal tissues, lens, and retina, normally isolated from the body's immune system, and ophthalmic surgeons will perform multiple surgical interventions on one or both eyes that violate the protective immune mechanisms of the eye that protect it from immune inflammation, at the present stage, not fully deciphered immunopathogenesis, SO cannot become an disappearing disease.

The rarity of SO can be explained by the immunological features of the eye as an immunoprivileged organ and the multifactorial nature of its etiopathogenesis, which is based on the "failure" of the eye's immune privilege in complicated penetrating trauma in combination with clinical, immunological and genetic risk factors, as well as significant frequencies of prophylactic nucleation of eyes with post-traumatic uveitis.

A complex mechanism of protection of the eye from immune inflammation was formed in the course of long-term evolutionary development of anatomical and physiological features of the eye, due to which it is often subject to open trauma with damage to internal autoantigenic structures. Many factors are needed to shake the evolutionarily complex mechanism of protecting the internal structures of the eyes from immune inflammation.

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