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CASE REPORT

Vogt-Koyanagi-Harada syndrome Case report

Nicula Cristina* **, Szabo Izabela*
***Ophthalmology Clinic, County Emergency Hospital Cluj-Napoca, Romania**
****"Iuliu Hatieganu" University of Medicine and Pharmacy, Cluj-Napoca, Romania**

Correspondence to: Cristina Nicula, MD,
Ophthalmology Clinic, County Emergency Hospital Cluj-Napoca, Romania
3-5 Clinicilor Street, Code 400006, Cluj-Napoca, Romania,
Mobile phone: +40722 849 575, E-mail: niculacristina65@yahoo.com

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Abstract

Vogt-Koyanagi-Harada syndrome is an uncommon multisystem inflammatory disorder characterized by panuveitis with serous retinal detachment and is often associated with neurologic and cutaneous manifestations including headache, hearing loss, vitiligo, and poliosis.

The case of a 62-year-old female with diabetes mellitus and a history of primary open angle glaucoma (POAG) in both eyes, operated on the left eye two weeks prior to the presentation and under topical antiglaucomatous drops, was reported. She presented at the ophthalmological service for decreased visual acuity (VA) in both eyes. The slit lamp examination revealed keratic precipitates and posterior iris synechiae in both eyes and an ExPress aqueous shunt in the left eye. Inferior retinal detachment was observed on ocular fundus examination on both eyes. Intraocular pressure value was in normal range under antiglaucomatous drops (dorzolamid + timolol).

The distinctiveness of this case was the association of the VKH syndrome with POAG and the inability to prolong the corticosteroid treatment, necessary in this case, due to the association of diabetes mellitus.

Keywords: Vogt-Koyanagy-Harada, retinal detachment, panuveitis

Introduction

Vogt-Koyanagi-Harada (VKH) syndrome is a multisystemic granulomatous autoimmune disease affecting organs with high melanocyte concentrations including the eye, central nervous system (CNS), inner ear, and skin [**1**]. The American Uveitis Society diagnostic criteria for VKH syndrome included no history of ocular trauma (including surgery) and at least one finding in three, out of the following four categories: 1) bilateral chronic iridocyclitis; 2) posterior uveitis, including exudative retinal detachment, disk hyperemia or edema and sunset glow fundus; 3) neurologic signs: tinnitus, meningismus; 4) cutaneous findings of alopecia, poliosis and/ or vitiligo [**2**].

The VKH syndrome can be divided into four clinical stages: 1) prodromal stage characterized by nonspecific symptoms such as fever, nausea, vertigo and the neurological symptoms like headaches, muscle weakness; 2) ophthalmologic stage, occurring a few days after the first stage - patients complaining of blurred vision, ocular pain, photophobia or central scotoma (bilateral in 80% of the cases); bilateral serous retinal

detachment often occurs, hearing disturbances may also be present; 3) convalescent stage, occurs within months from the onset and is characterized by poliosis involving the eyebrows and eyelashes, hair loss and vitiligo; 4) chronic recurrent stage - recurrent uveitis and ophthalmological complications [**3**].

Case report

In January 2014, the 62-year-old patient known with diabetes mellitus, alopecia, bilateral neurosensitive hypoacusis, and arthritis presented to the Eye Clinic with complaints of severe headaches associated with painful left eye, nausea and vomiting. Based on the symptoms, values of the intraocular pressure (IOP) (OD: 30 mmHg; OS: 40 mmHg), visual field and gonioscopy, which showed an open angle grade II/ III in both eyes, the patient was diagnosed with decompensated POAG in both eyes and immediately started a treatment with antiglaucomatous drops (fixed combination - bimatoprost + timolol). The IOPs in the left eye continued to be high, despite the medical treatment, so the patient underwent a surgical intervention for the implantation of an ExPress aqueous shunt. After the surgery, the IOP in the left eye was 19 mmHg.

Two weeks after the surgery, the patient complained of decreased visual acuity and ocular congestion in both eyes. The ophthalmological examination revealed signs of bilateral anterior and posterior uveitis associated with an inflammatory retinal detachment. The slit lamp examination showed keratic precipitates and posterior iris synechiae in both eyes and an ExPress aqueous shunt in place and the filtration bleb present in the left eye (**Fig. 1**).

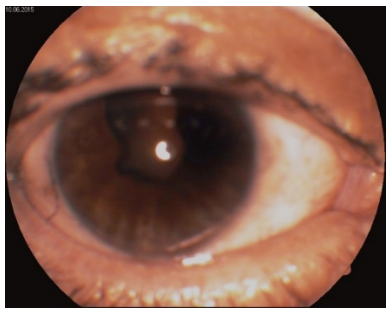

Fig. 1 Right eye with posterior iris synechiae

The ophthalmological examination revealed best corrected visual acuity of 20/ 400 wc in the right eye and 20/ 500 wc in the left eye.

The fundus exam revealed an inferior retinal detachment in both eyes with a C/ D ratio of 0.6-0.7 in the right eye (**Fig. 2**) and 0.8 in the left eye (**Fig. 3**). Intraocular pressure was 14 mmHg in the right eye and 15 mmHg in the left eye under fixed combination antiglaucomatous drops (bimatoprost + timolol).

The paraclinical examinations revealed:

-laboratory examinations: high inflammatory markers (VSH, CRP, fibrinogen), high glycemia, anemia.

-MRI examination revealed bilateral choroidal and retrobulbar contrast enhancement, while brain findings included white matter abnormalities on FLAIR and leptomeningeal enhancement.

-rheumatological examination showed the presence of vasculitis, arthritis.

-ENT examination revealed a bilateral neurosensitive hypoacusis.

-dermatological examination revealed alopecia and no signs of vitiligo or poliosis

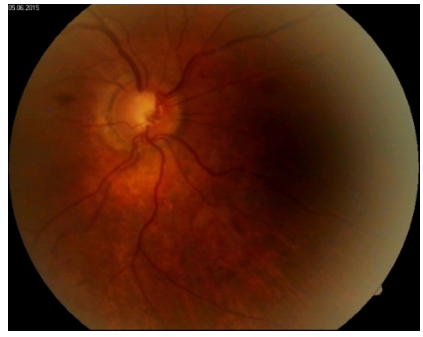

Fig. 2 Right eye fundus

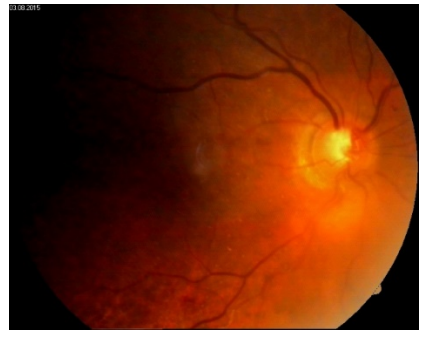

Fig. 3 Left eye fundus

The patient was treated with systemic hydrocortisone starting with 300 mg per day, reducing the value each third day; ceftriaxone 2 g per day and topical drops with a fixed combination of tobramycin - dexamethasone, mydriatic drops, each 5 drops per day and fixed combination antiglaucomatous drops dorzolamid - timolol twice per day in both eyes (BE).

Discussions

The patient was under the observation of a rheumatologist, being diagnosed with arthritis and underwent medical treatment with leflunomide pills for years before. Willingly she decided to stop taking treatment a few weeks prior to the ocular surgery.

The darker pigmented skin of our patient was also suggestive for VKH syndrome since studies have reported a predilection of the disease for female gender and darker pigmented races [**4,5**].

The treatment options in our case were limited due to the complications that corticosteroid medication had on the diabetic patient and mainly addressed the symptoms. In case of recurrence, the adding of steroid-sparing agents, such as methotrexate, azathioprine and cyclosporine might be taken into account.

Glaucoma may occur as a secondary ocular manifestation of VKH syndrome [**6**], as well as cataract.

Periodical ophthalmologic examination is recommended in order to monitor the IOP, the presence of ocular inflammation and consequently prevent a painful eye.

The distinctiveness of this case was the association of VKH syndrome with POAG and also the inability to prolong the corticosteroid treatment, necessary in this case, due to the association of diabetes mellitus.

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