Journal of 21 September University for Medical and Applied Sciences
Volume (2), Issue (1): Jul 2023
P: 9-16
Journal homepage: http://21umas.edu.ye/masj

Case Series Study

Effect of Pulse Steroid Therapy on Visual Acuity and Fundus Picture in Cases of Vogt Koyanagi Harada Syndrome in Yemen: Case Series Study

Nabil Taresh*[1], Hisham Alakhlee[1], Basma Al-Refai[1], and Ahmad Labib[2]

1 Department of Ophthalmology, College of Medicine, 21 September University for Medical and Applied Sciences, Sana'a, Yemen.
2 Consultant Ophthalmologist, FRSE.

*Corresponding Author: Nabil Taresh, 21 September University for Medical and Applied Sciences, Sana'a, Yemen. Tel: +967777068997

Article History| Received: 02.04.2023 | Accepted: 04.06.2023 | Published: 28.07.2023

Abstract

Vogt-Koyanagi-Harada syndrome (VKHS) is a rare systemic disease with severe bilateral panuveitis associated with cutaneous, neurological, and auditory abnormalities. This study aims to evaluate the effect of pulse steroid therapy on the visual acuity and fundus picture of cases of VKHS presented at Maghrabi Eye Hospital in Yemen. Method: This retrospective case series was followed up for three years between (2007-2010) for three female patients who presented by bilateral hand motion and complained with headache, neck stiffness and difficulty in hearing. Results: After pulse steroid therapy, the best corrected visual was improved with resolution of optic disc swelling and serous retinal detachment detected by optical coherence tomography (OCT). One eye had complication as sunset glow and suprarational fibrosis. Conclusion: VKHS is uncommon disease in Yemen and affects mostly female. Urgent intervention with pulse steroid therapy and systemic corticosteroid and early diagnosis decrease the complications and improve the visual acuity.

Key words: Vogt Koyanagi Harada syndrome, pulse steroid therapy, visual outcome, Yemen.

Introduction

VKHS is a rare systemic disorder involving various melanocyte-containing organ systems, including the eyes, ears, skin, and meninges (1). Although VKH has been reported throughout the world, its appearance seems to be concentrated in certain racial and ethnic groups predominantly in Asians, Hispanics, and Native Americans (2). In the 12th century, a physician from the Arab world, Mohammad-al-Ghafiqi, described a disease with poliosis, neuralgias, and hearing changes (3). Alfred Vogt presented one instance in 1906 (4), Harada documented one case in 1923, and in 1926, a case of basically posterior uveitis with an

exudative retinal detachment connected to pleocytosis in the cerebrospinal fluid was described (CSF) (3). Six cases with bilateral nontraumatic chronic iridocyclitis linked to polio and vitiligo were described in great detail in 1929 (5). It was already well known that the symptoms of Vogt-Koyanagi syndrome and Harada's illness were very similar. The term Vogt-Koyanagi-Harada disease (syndrome) was first used in the majority of published publications in 1970, and by 2003, most authors had accepted it (6). VKHS involves the eye causing severe bilateral panuveitis associated with cutaneous, neurological, and auditory abnormalities. It initiates with a prodromal stage that lasts for a few days and is characterized by a viral-like illness that is frequently accompanied by headache, stiff neck, and confusion, followed by an acute uveitis stage which lasts for a few days and is characterized by the development of bilateral diffuse choroiditis, papillitis, and exudative retinal detachment (7). Most cases of VKHS were presented firstly to neurologists and symptoms that are related to eye were presented later (8). Thus, making the diagnosis of VKHS is very difficult. In the opposite, early medical treatment is mandatory to recover fast and to safe the vision (5). The etiologic and pathogenic factors in VKHS remain unclear (9). The major histopathologic feature of VKH is a diffuse granulomatous inflammation of the uveal tract with a preponderance of lymphocytes and epithiloid cells (10). The choriocapillaris are usually, but not always, involved. In the late stage, there are disappearance of choroidal melanocytes, chorioretinal scaring and occasionally choroidal neovascularization (11). When it is first diagnosed, VKHS usually responds to oral or intravenous high-dose corticosteroid therapy. Early treatment can benefit in preventing disease progression to later stages of its natural course (7). When corticosteroids are contraindicated due to a systemic underlying disease (such as diabetes), immunosuppressive therapy can be added to the regimen of treatment for VKH at various stages of the disease to increase the potency of corticosteroid therapy and decrease the overall dosage of corticosteroid therapy required to control the disease (12). Recently, several authors suggested using immunosuppressive medications in the very early stages of the disease (13) to lessen relapses and stop the development of sunset glow fundus, which may be linked to impaired visual function even when good visual acuity is still present (7).

In Yemen, there is no research about VKHS which is a rare disease and mostly affect women (14). Therefore, this study aimed to describe Effect of pulse steroid therapy on visual acuity and fundus picture in cases of Vogt Koyanagi Harada Syndrome presented to Maghrabi Eye Hospital, Sana'a, Yemen.

Case Presentation

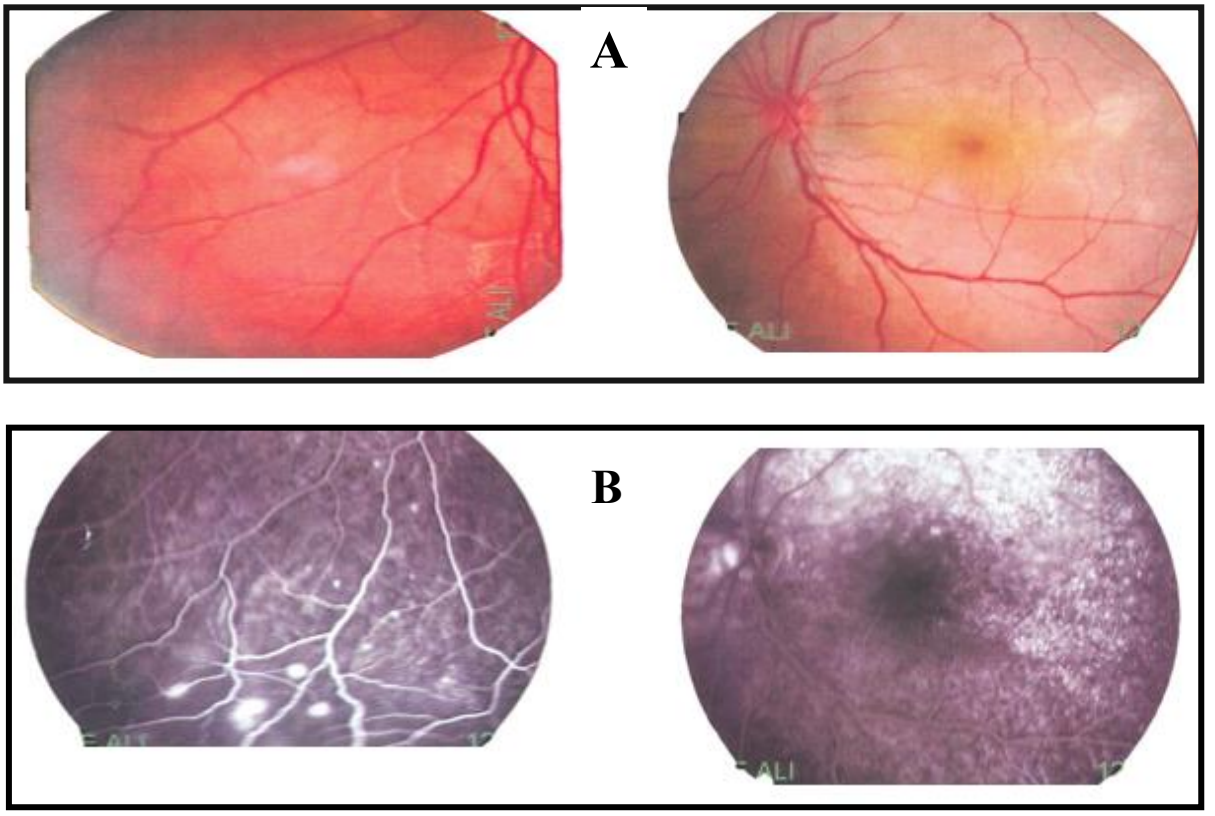
Method

Three female patients ranging between 20-50 years old were admitted to Maghrabi Eye Hospital in Yemen with bilateral poor vision, headache, neck stiffness and difficulty in hearing, denied comorbidities or other systemic symptoms. Ancillary exams for infectious diseases including serologies for syphilis (FTA-Abs and

VDRL), HIV, tuberculosis and toxoplasmosis, chest X ray and Mantoux test were ordered, as well as fundus imaging.

Visual acuity in all cases was hand motion. Ocular examination revealed normal anterior segment and intra ocular pressure. All cases underwent fundoscopy, fluorescence angiography, B-scan ultrasonography, and ocular coherent tomography (OCT). The patients were followed up for three years between (2007-2010).

Case1- A 20-years old patient: fundus examination showed optic disc hyperemia and exudative retinal detachment (Figure 1A), fluorescence angiography showed punctuate pin point hyperfluorescence, dye polling in the fovea and inferiorly (Figure 1B), O.C.T showed hypo-reflective between neurosensore retina and retinal pigment epithelium indicating the presence of serous fluid (Figure 1C), and ultrasonography showed serous retinal detachment located at the posterior pole and inferiorly, diffuse, low to medium reflective choroidal thickening, (Figure 1D).



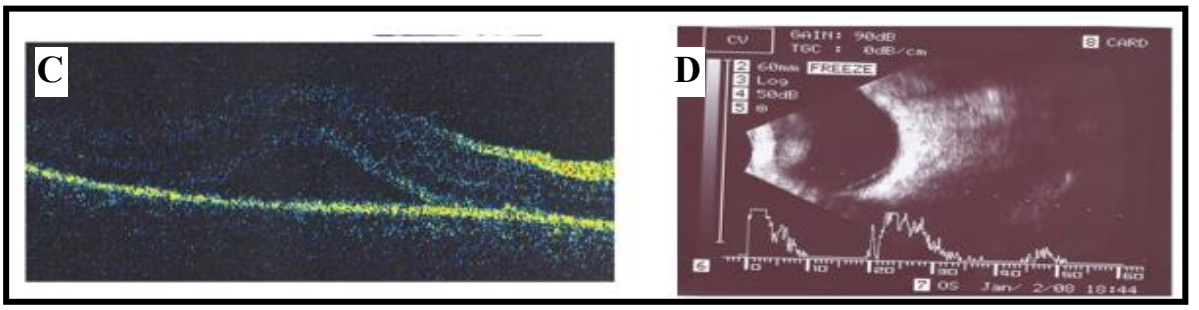


Figure 1: Initial examination of the left eye in case one (A) Color fundus images, (B) Fluorescence angiography, (C) O.C.T, and (D) ultrasonography.

Case 2- A 35-years old patient, (Figure 2). (A) Color fundus images of the left eye showed optic disc edema and exudative retinal detachment in the posterior pole. (B) Fluorescence angiography showed hyperflourasceinnse and late leakage at the disc, punctuate pin point hyperflourescence, dye polling in the fovea. (C) O.C.T showed hypo-reflective between neurosensore retina and retinal pigment epithelium indicating the presence of serous fluid. (D) Ultrasonography showed serous retinal detachment located at the posterior pole and inferiorly, diffuse, low to medium reflective choroidal thickening.

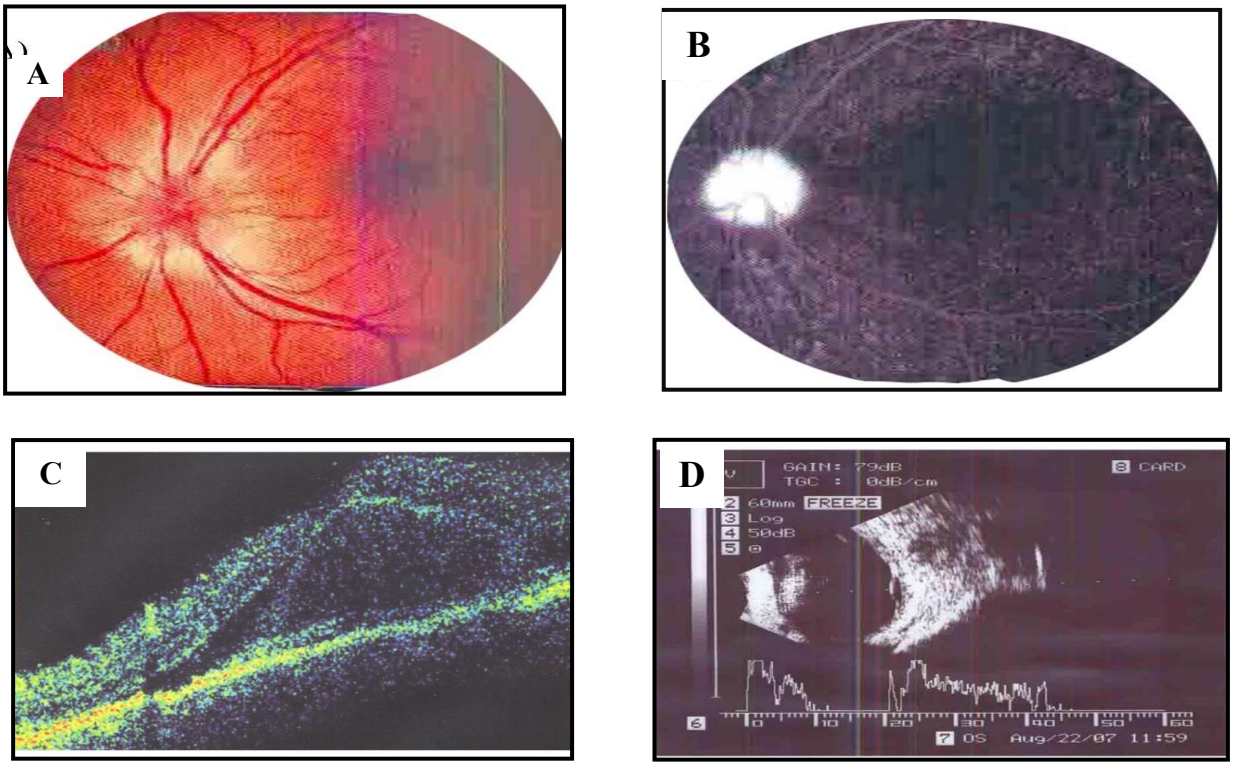


Figure 2: Initial examination of the left eye in case two (A) Color fundus images, (B) Fluorescence angiography, (C) O.C.T, and (D) ultrasonography

Case 3- A 50 years old patient, (Figure 3). (A) Color fundus images of the right eye showed optic disc edema and multiple pockets exudative retinal detachment in the posterior pole and in the periphery. (B) Fluorescence angiography showed hyperflourasceinnse and late leakage at the disc, punctuate pin point hyperflourescence, dye polling in the fovea. (C) O.C.T showed hypo-reflective between neurosensore retina and retinal pigment epithelium indicating multiple serous detachments. (D) Ultrasonography showed serous retinal detachment located at the posterior pole and inferiorly, diffuse, low to medium reflective choroidal thickening.

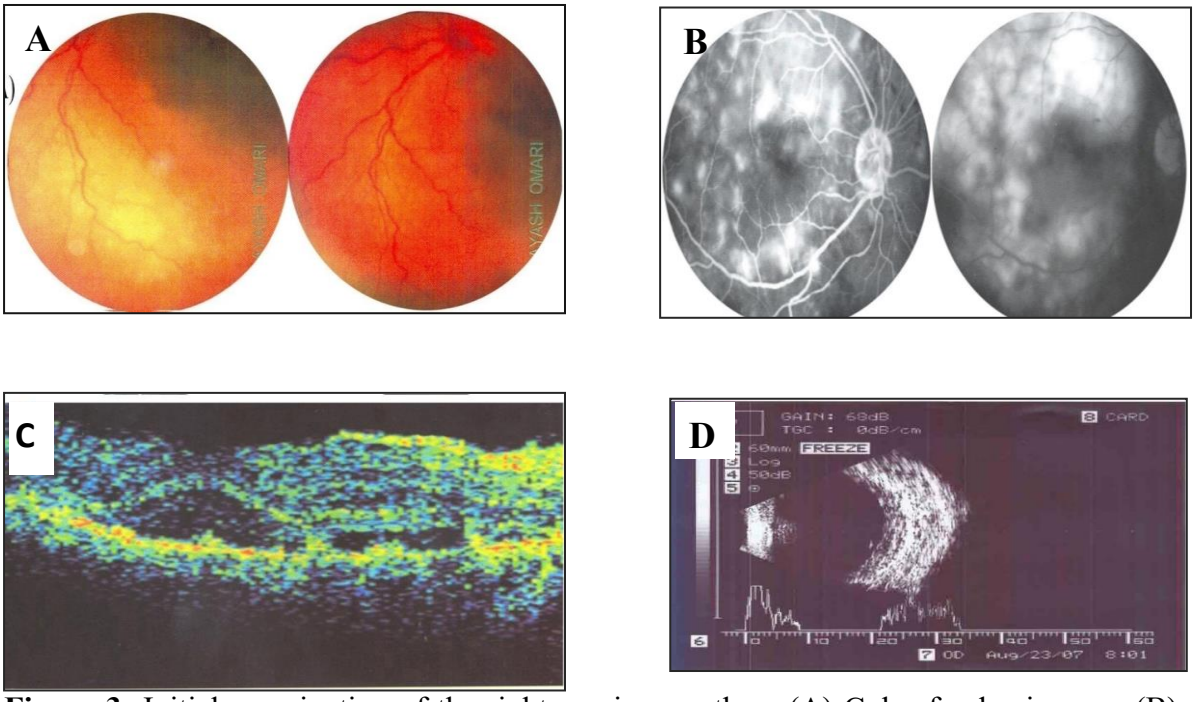


Figure 3: Initial examination of the right eye in case three (A) Color fundus images, (B) Fluorescence angiography, (C) O.C.T, and (D) ultrasonography

All cases were treated by pulse steroid therapy with intravenous Methylprednisolone 1G daily for three consecutive days followed by oral steroid (1mg per kg) with long term tapring.

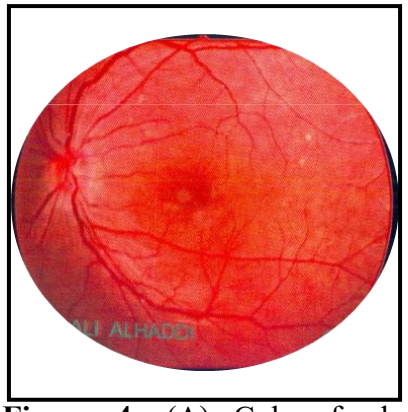
Results

Table (1) and Figures (4-6) show that after steroid therapy, best corrected visual acuity was improved, and resolution of the serous detachment was detected by color fundus and OCT. Convalescent phase showed retinal pigment epithelium (RPE) changes in all cases. Figure 4 shows color fundus image of the left eye and OCT shows RPE depigmentation (sunset glow) in case1, Figure 5 shows color fundus images of the left eye and OCT shows RPE scare in case 2, and Figure 6 shows color fundus images of the left eye and OCT shows atrophy, subretinal submacular fibrosis in case 3.

Table 1. Best corrected visual acuity

Cases	Eye	Vision
Case 1	OS	20/40
Case 2	OS	20/22
Case 3	OD	20/160

A



B

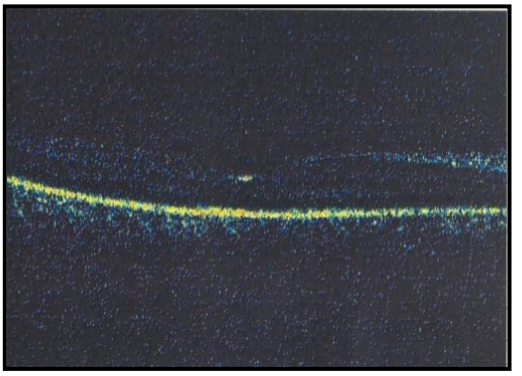
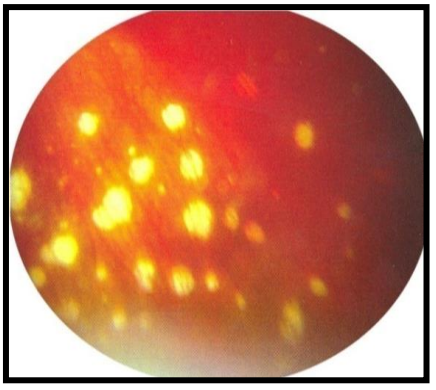


Figure 4: (A) Color fundus images after treatment, and (B) O.C T RPE depigmentation after treatment

A



B

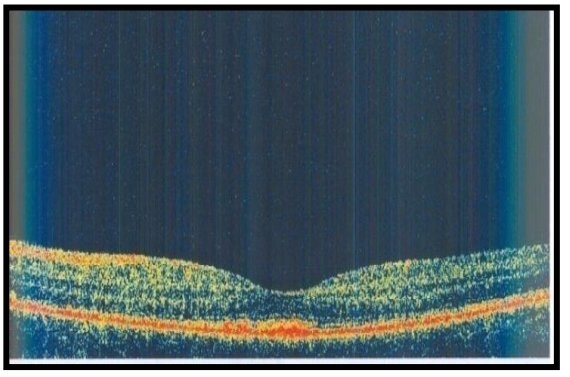


Figure 4: (A) Color fundus images after treatment, and (B) O.C T RPE scare after treatmen

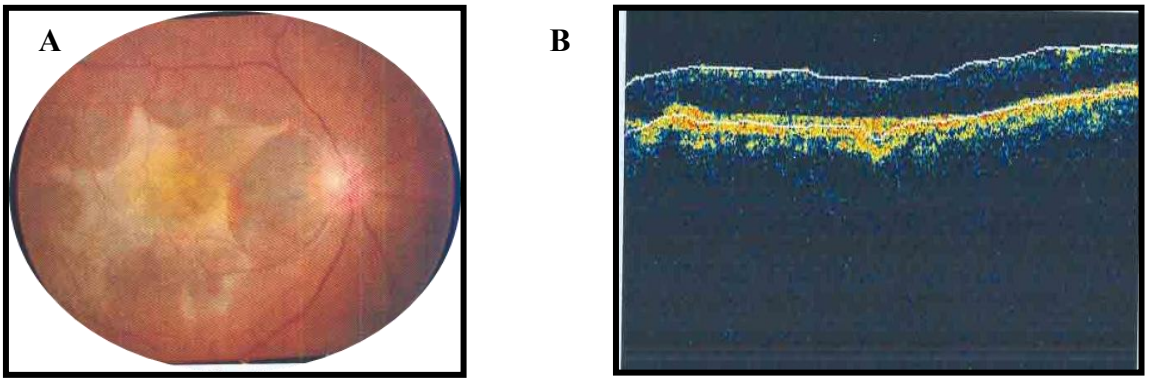


Figure 4: (A) Color fundus images after treatment, and (B) O.C T RPE atrophy, supretinal,supmacular fibrosis after treatment

Discussion

VKHS, a multisystemic inflammatory condition, needs prompt initial treatment to minimize secondary complications and vision loss (16). The treatment of VKH most commonly contains corticosteroids in the acute phase of the disease. When corticosteroids alone are not enough to control a condition or when a patient's response to corticosteroids is insufficient, immunomodulatory therapy may be started. Pulse intravenous corticosteroids, consisting of methylprednisolone 1000 mg/day for 3 days, have been routinely utilized (12). According to our research, intravenous methylprednisolone therapy may cause individuals with acute VKH disease to experience a rapid reduction in inflammation followed by a rapid improvement in visual. This is consistent with a few smaller case studies that show early recovery of visual acuity and quick reduction of subretinal fluid (12, 17-19).

Conclusion

The aim of this study was to investigate the effect of pulse steroid therapy on visual acuity and fundus picture in 3 cases of VKHS in Yemen. VKHS is a rare disease and more common in females worldwide. This is the first study in Yemen. The cases were treated with pulse steroid therapy. The results of this study show that pulse steroid therapy led to rapid resolution of inflammation and subsequently induce rapid recovery of visual acuity in patients with VKHS on the visual acuity and fundus picture of cases. This study suggests that pulse steroid should be used as first line for treatment of VKHS. The most important practical implications are early diagnosis and proper treatment to decrease the complication and to improve the visual acuity.

Financial disclosure: There are no conflicts of interest.

References

.1 Johal B, Johal H, Lukaris AJUBCMJ. The clinical presentation and diagnosis of Vogt-Koyanagi-Harada syndrome. 2015;6:20-2.

.2 Herbort CP, Jr., Tugal-Tutkun I, Abu-El-Asrar A, Gupta A, Takeuchi M, Fardeau C, et al. Precise, simplified diagnostic criteria and optimised management of initial-onset Vogt-Koyanagi-Harada disease: an updated review. Eye (Lond). 2022;36(1):29-43.

.3 Sen HNJW, Fundamentals NsU, Practice C. Vogt-Koyanagi-Harada Syndrome. 2021:296.

.4 Herbort CP, Mochizuki MJIO. Vogt–Koyanagi–Harada disease. Springer; 2007. p. 63-5.

.5 Baltmr A, Lightman S, Tomkins-Netzer OJCO. Vogt–Koyanagi–Harada syndrome–current perspectives. 2016:2345-61.

.6 Urzua CA, Herbort CP, Jr., Takeuchi M, Schlaen A, Concha-Del-Rio LE, Usui Y, et al. Vogt-Koyanagi-Harada disease: the step-by-step approach to a better understanding of clinicopathology, immunopathology, diagnosis, and management: a brief review. J Ophthalmic Inflamm Infect. 2022;12(1):17.

.7 Accorinti M, Saturno MC, Iannetti L, Manni P, Mastromarino D, Pirraglia MP. Treatment and Prognosis of Vogt-Koyanagi-Harada Disease: Real-Life Experience in Long-Term Follow-Up. J Clin Med. 2022;11.(13)

.8 Lavezzo MM, Sakata VM, Morita C, Rodriguez EEC, Abdallah SF, da Silva FT, et al. Vogt-Koyanagi-Harada disease: review of a rare autoimmune disease targeting antigens of melanocytes. 2016;11:1-21.

.9 Zhang Q, Fan X, Tian M, Han HJBn. A case presentation of an IgA nephropathy patient with Vogt-Koyanagi-Harada syndrome. 2020;21(1):1-5.

.10 Rao NA, Moorthy RS, Inomata HJIoc. Vogt-Koyanagi-Harada syndrome. 1995;35(2):69-86.

.11 Chan C-C, Palestine AG, Kuwabara T, Nussenblatt RBJAjoo. Immunopathologic study of Vogt-Koyanagi-Harada syndrome. 1988;105(6):607-11.

.12 Nakayama M, Keino H, Watanabe T, Okada AA. Clinical features and visual outcomes of 111 patients with new-onset acute Vogt-Koyanagi-Harada disease treated with pulse intravenous corticosteroids. Br J Ophthalmol. 2019;103(2):274-8.

.13 Herbort CP, Jr., Abu El Asrar AM, Takeuchi M, Pavesio CE, Couto C, Hedayatfar A, et al. Catching the therapeutic window of opportunity in early initial-onset Vogt-Koyanagi-Harada uveitis can cure the disease. Int Ophthalmol. 2019;39(6):1419-25.

.14 Chang JH-M, Wakefield DJOi, inflammation. Uveitis: a global perspective. 2002;10.79-263:(4)

.15 Hosseini SM, Dourandish M, Mazouchi M. Unilateral Presentation of Vogt-Koyanagi-Harada Syndrome. Journal of ophthalmic & vision research. 2020;15(1):99-103.

.16 Dias NdC, Ventura CV. Vogt-Koyanagi-Harada syndrome: a discussion about resistance to corticotherapy. Revista Brasileira de Oftalmologia. 2021;80.(3)

.17 Yamanaka E, Ohguro N, Yamamoto S, Nakagawa Y, Imoto Y, Tano Y. Evaluation of pulse corticosteroid therapy for vogt-koyanagi-harada disease assessed by optical coherence tomography.

American journal of ophthalmology. 2002;134(3):454-6.

.18 Badsha H, Edwards CJ. Intravenous pulses of methylprednisolone for systemic lupus erythematosus. Seminars in Arthritis and Rheumatism. 2003;32(6):370-7.

.19 Miyanaga M, Kawaguchi T, Shimizu K, Miyata K, Mochizuki M. Influence of early cerebrospinal fluid-guided diagnosis and early high-dose corticosteroid therapy on ocular outcomes of Vogt–Koyanagi–Harada disease. International Ophthalmology. 2007;27(2):183-8.