CHAPTER 4

Evolution of the acute uveitic phase in Vogt-Koyanagi-Harada Syndrome

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ABSTRACT
Background: Vogt–Koyanagi–Harada (VKH) disease is an inflammatory and autoimmune condition characterized by panuveitis, serous retinal detachments and extraocular manifestations of the auditory, integumentary and central nervous systems. Patients with VKH can have good final outcomes if treated promptly with immunosuppressive agents and thus avoid complications. The purpose of this article is to report the evolution of the acute uveitis phase of VKH.
Case Report: We report a case of a 58 years old women who presented to ophthalmology emergency room with a 10-day history of a severe headache and progressive low visual acuity in the left eye (LE). After seven days she developed low visual acuity (VA) in other eye and important tinnitus in her left ear. Ophthalmological examination showed best-corrected visual acuity (BCVA) of 20/200 in both eyes; direct and consensual motor reflexes decreased bilaterally; anterior chamber reaction with 2+ cells in both eyes. At Fundoscopy: bilaterally hyperemic optic discs, serous retinal detachment, retinal folds in the papillomacular bundle and Dalen-Fuchs nodules. Angiofluoresceinography (AGF) showed bilateral macular hyperfluorescence (pinpoints). Optical Coherence Tomography (OCT) showed bilateral subretinal fluid and loss of foveal depression. Oral prednisone 1,2mg/kg/day and topical dexamethasone were introduced with progressive tapering for 9 months with complete improvement in VA of the right eye and partial improvement in VA of the left eye.
Conclusion: VKH can cause vision loss and blindness. Early diagnosis and adequate treatment with immuno-suppressive agents may halt disease progression and prevent recurrences and vision loss.

Keywords: panuveitis, corticosteroid, retinal detachment, tinnitus.

1 INTRODUCTION

Vogt-Koyanagi-Harada (VKH) disease is an immune-mediated disorder affecting both eyes. Although the exact cause of Vogt-Koyanagi-Harada disease is unclear, the most accepted mechanism involves a viral-triggered, T-cell-mediated autoimmune response against melanocyte-associated antigens in a genetically vulnerable individual. This typically manifests in several different stages during the disease course: the prodromal phase, acute uveitic phase, convalescent phase and chronic recurrent phase[1,2,3].

Several sets of diagnostic criteria have been proposed. The Revised Diagnostic Criteria (RDC) are widely used at present and are highly sensitive and specific. The RDC divide VKH disease into 3 categories— complete, incomplete, and probable — and do not allow a decision concerning a definite diagnosis for patients without any extraocular manifestation[4,5]. It is required bilateral ocular involvement, characteristic neurological/auditory and dermatological involvement without ocular trauma or surgery, and no other clinical or laboratory evidence to suggest an alternative diagnosis[1,6,7].

There have been trials of multiple different treatment regimens over the years, but the vest evidence for effectiveness in both short and long term is with early and aggressive high-dose systemic corticosteroids with a slow oral taper over at least 6 months. Rapidly stopping corticosteroids increases the chance of recurrence[1].

2 CASE REPORT

A 58 years old Caucasian women who presented to the ophthalmology emergency room of the Irmandade da Santa Casa de Misericórdia de São Paulo – ISCMSP with a 10-day history of a severe headache and progressive low visual acuity in the left eye. After seven days she developed low visual acuity (VA) in other eye and important tinnitus in her left ear. She denied use of tobacco smoking, alcohol and substance abuse. The patient denied ocular trauma, previous eye surgery, medication use, arthralgia, skin lesions and genitourinary symptoms.

The ophthalmological examination showed best-corrected visual acuity (BCVA) of 20/200 in both eyes. Direct and consensual motor reflexes decreased bilaterally. At anterior segment biomicroscopy: anterior chamber reaction with 2+ cells in both eyes. At Fundoscopy: bilateral hyperemic optic discs, diffuse serous retinal detachment, changes in the vitreoretinal interface, retinal folds in the papillomacular bundle and presence of Dalen-Fuchs nodules.

Additional tests were requested: retinography of the right eye (Figures 1A and B) and left eye (Figures 2A and 2B) showed bilateral hyperemic optic discs, serous retinal detachment, retinal folds in the papillomacular bundle and Dalen-Fuchs nodules; angiofluoresceinography (AGF) of the right eye (Figure 3A) and left eye (Figure 3B) evidenced diffuse hyperfluorescence in the initial phase, macular hyperfluorescence points (pinpoints) with contrast leakage at a late stage and diffuse accumulation of contrast mainly in the left eye below the macula; optical coherence tomography (OCT) of the right eye (Figure 4) and left eye (Figure 5) showed loss of foveal depression and presence of subretinal fluid (serous retinal detachment); and serological tests with negative results.

Oral prednisone 1,2mg/kg/day (100mg/day) and 01 drop every 8 hours of topical dexamethasone 0,1% in both eyes were introduced. After 15 days of treatment BCVA of 20/50 in right eye and 20/70 in left eye with partial improvement of the optic disc hyperemia and serous retinal detachment (Figures 6 and 7). Slow tapered of dexamethasone dose to 01 drop every 08 hours and maintained oral prednisone dose.

At 30 days of treatment BCVA of 20/20 in the right eye and 20/30 in the left eye with complete improvement of the optic disc hyperemia of the right eye (Figure 8) and partial improvement of the optic disc hyperemia of the left eye (Figure 9), and complete resolution of the serous retinal detachment of both eyes (Figures 10 and 11). Slow tapered of dexamethasone dose to 01 drop every 12 hours and oral prednisone to 1,0mg/kg/day (80mg/day).

After 30 days of treatment, tapered 10mg of prednisone per month and suspended dexamethasone. Patient had BCVA improvement to 20/20 in the right eye and 20/25 in the left eye and sustained without corticosteroid after 9 months of treatment.

Figure 1A – Retinography of the right eye: hyperemic optic disc, serous retinal detachment and retinal folds in the papillomacular bundle

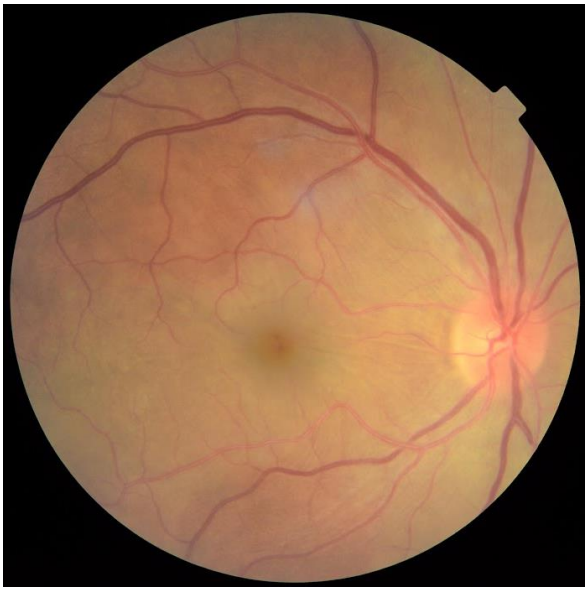


Figure 1B – Retinography of the right eye: Dalen-Fuchs nodules

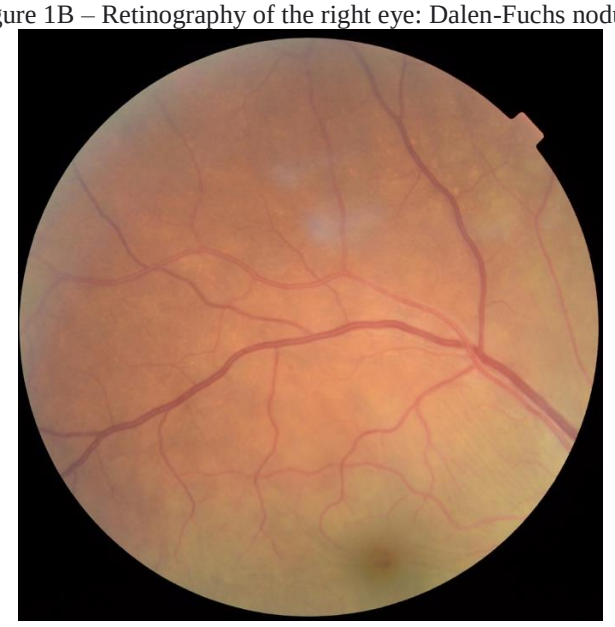


Figure 2A– Retinography of the left eye: hyperemic optic disc, serous retinal detachment and retinal folds in the papillomacular bundle.

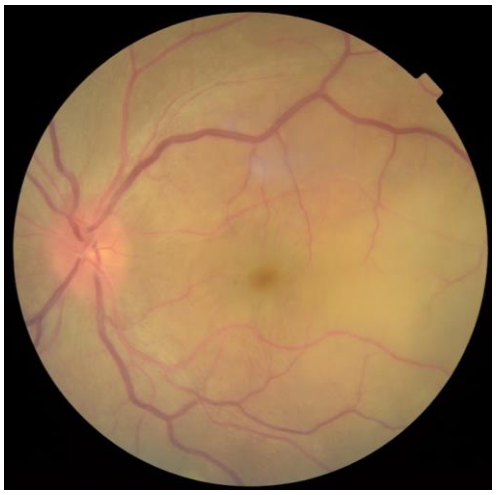


Figure 2B – Retinography of the left eye: Dalen-Fuchs nodules.

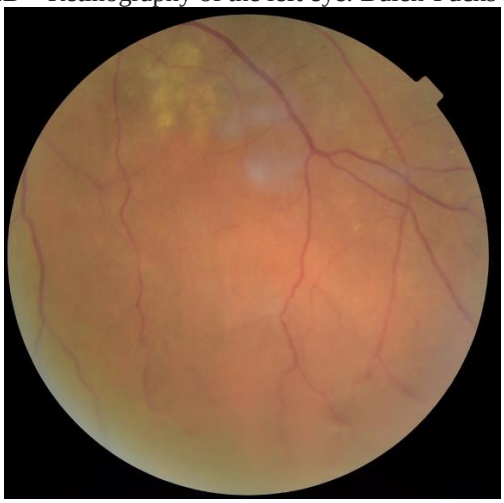


Figure 3A – Angiofluoresceinography of the right eye: diffuse hyperfluorescence in the initial phase, macular hyperfluorescence points (pinpoints) with contrast leakage at a late stage and diffuse accumulation of contrast below the macula.

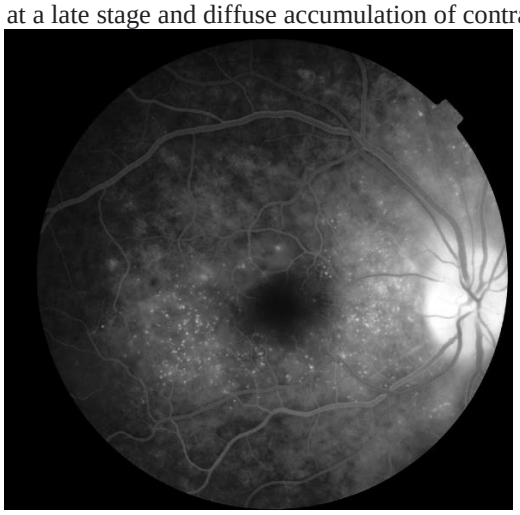


Figure 3B – Angiofluoresceinography of the left eye: diffuse hyperfluorescence in the initial phase, macular hyperfluorescence points (pinpoints) with contrast leakage at a late stage and diffuse accumulation of contrast below the macula.

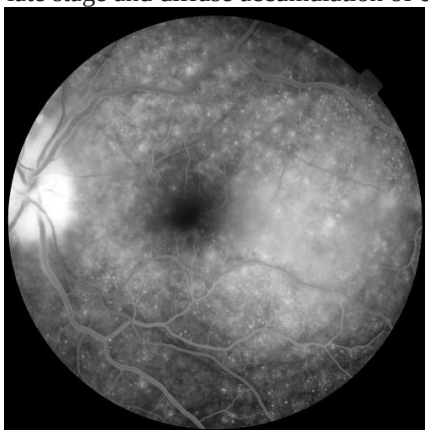


Figure 4 – Optical coherence tomography of the right eye: loss of foveal depression and presence of subretinal fluid (serous retinal detachment).

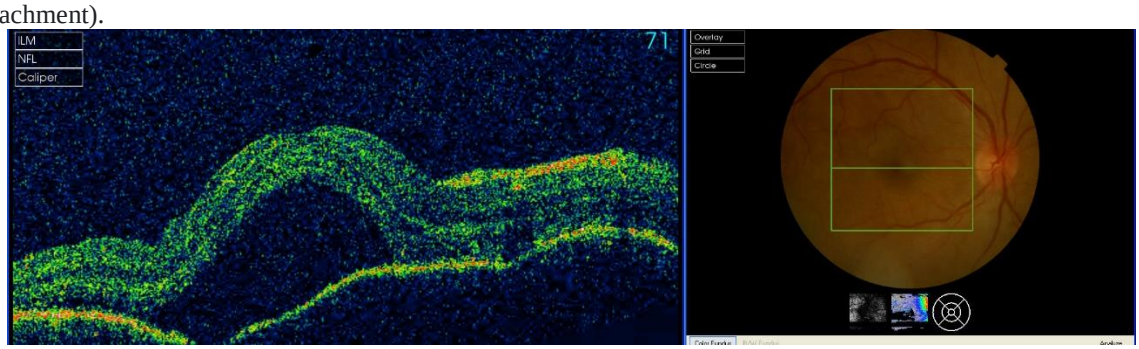


Figure 5 – Optical coherence tomography of the left eye: loss of foveal depression and presence of subretinal fluid (serous retinal detachment).

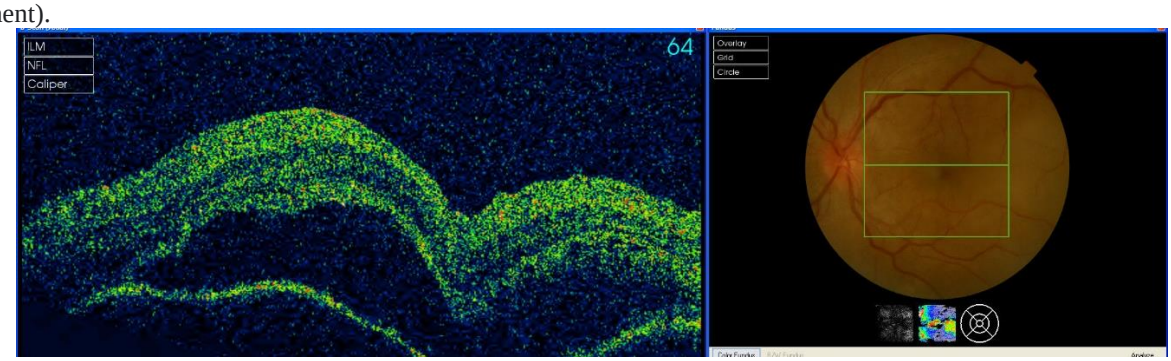


Figure 6 – Retinography of the right eye after 15 days of treatment: partial improvement of the optic disc hyperemia and serous retinal detachment

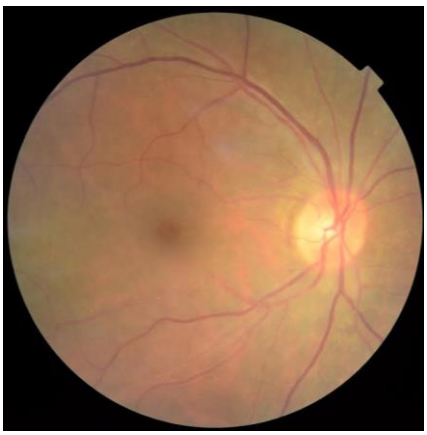


Figure 7 – Retinography of the left eye after 15 days of treatment: partial improvement of the optic disc hyperemia and serous retinal detachment.

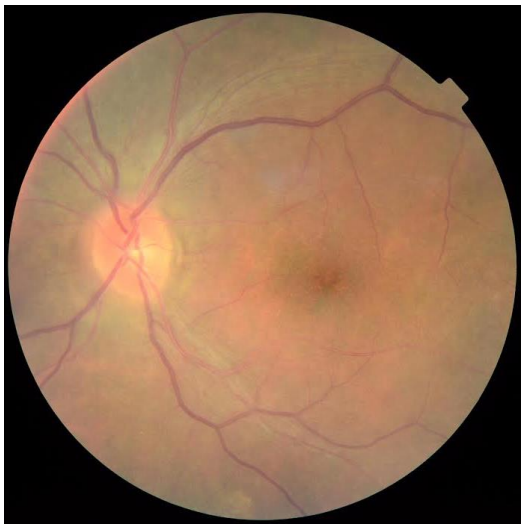


Figure 8 – Retinography of the right eye after 30 days of treatment: complete improvement of the optic disc hyperemia.

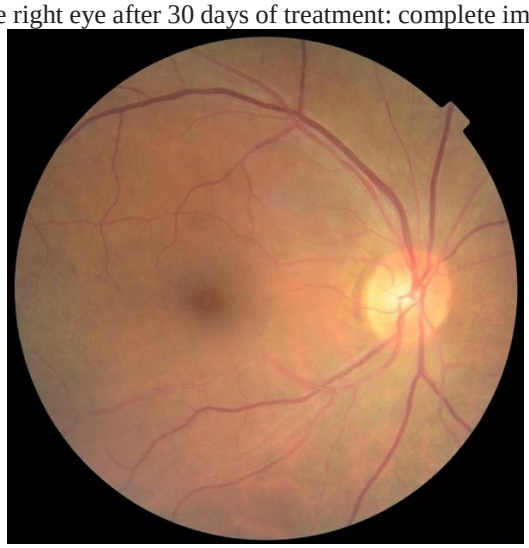


Figure 9 – Retinography of the left eye after 30 days of treatment: partial improvement of the optic disc hyperemia.

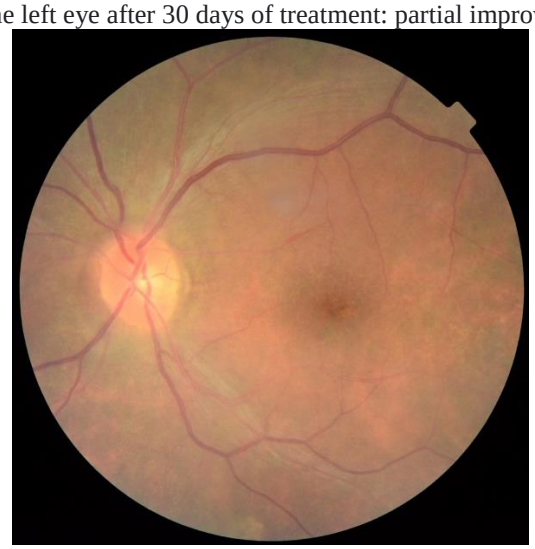


Figure 10 – Optical coherence tomography of the right eye: foveal depression and complete improvement of subretinal fluid (serous retinal detachment).

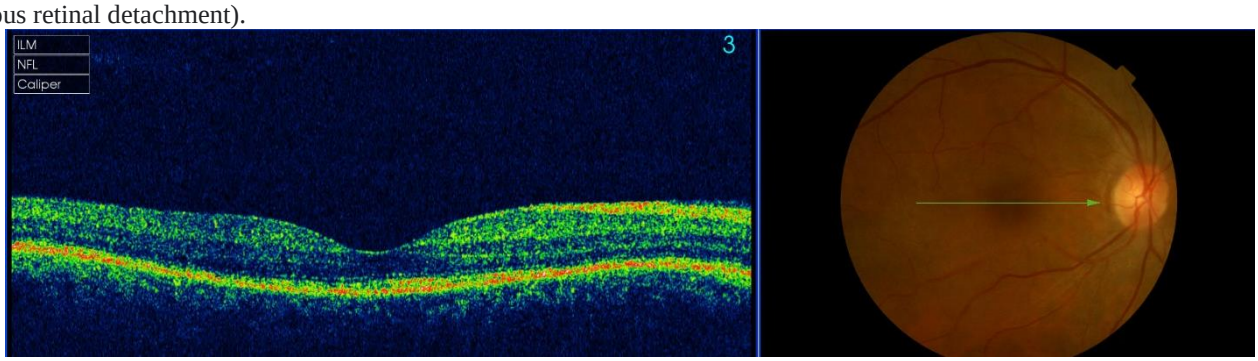
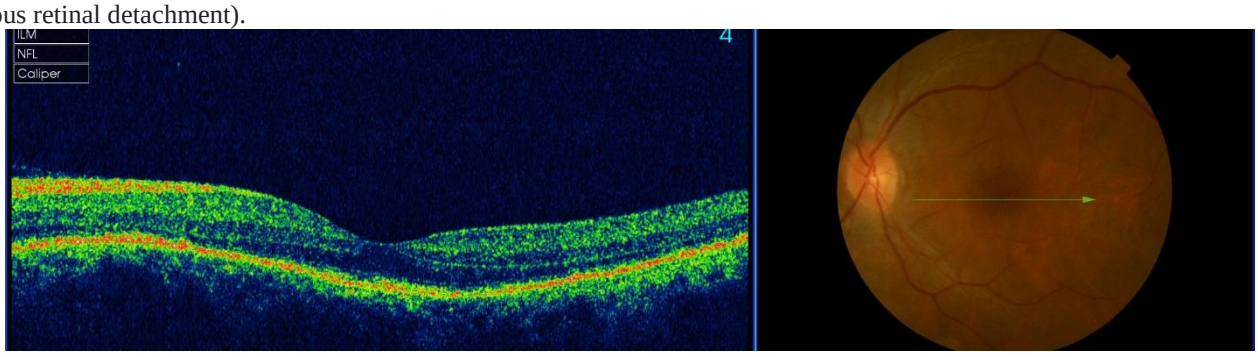


Figure 11 – Optical coherence tomography of the left eye: foveal depression and complete improvement of subretinal fluid (serous retinal detachment).



3 DISCUSSION

There are no laboratory tests that diagnose VKH; rather, the diagnosis is clinical[8]. As such, several sets of diagnostic criteria have been proposed[4,5,9,10]. The Revised Diagnostic Criteria for VKH Disease classify patients as having complete VKH, incomplete VKH, and probable VKH[4]. Criteria 1 to 4 were filled up as the patient developed a severe headache, low visual acuity in both eyes, important tinnitus in her left ear and the ophthalmological examination showed hyperemic optic discs, serous retinal detachment, Dalen-Fuchs nodules and hyperfluorescence points (pinpoints).

VKH disease classically has four clinical phases including prodromal; acute uveitic; chronic convalescent; and chronic recurrent stage. The prodromal phase usually lasts few days and patients typically complain of neurological and auditory symptoms. Optic disc swelling is rare in this early prodromal phase. However, during the acute uveitic phase, optic disc hyperemia is a frequent find- ing. In this stage impairment of vision occurs due to diffuse choroiditis and multiple serous retinal detachments. Several weeks later, the disease progresses to the convalescent phase, which is characterized by depigmentation of the choroid ‘‘sunset glow fundus’’. In this phase, the optic disc may appear pale[3,11]. We report a case of women in acute uveitis phase: headache, hyperemic optic discs, serous retinal detachment and tinnitus. She did not develop integumentary findings (alopecia, vitiligo or poliosis) because these symptoms occur in the convalescence phase[12]. Therefore, the patient would probably fulfill all the criteria being classified in complete VKH[4] when in the convalescence phase.

The goal of treatment in VKHD is to suppress active ocular inflammation, prevent disease relapse and avoid sight- threatening complications. As such, early diagnosis and rapid commencement of treatment are important in preserving vision in these young patients[13]. Because VKHD can involve multiple organs, the mainstay of treatment is based on high-dose systemic corticosteroids[14,15].

Oral prednisone at a dose of 1–2 mg/kg/day started early in the course of the disease followed by slow tapering to avoid recurrences is the generally accepted regimen. Slow tapering of the corticosteroid dose, with frequent follow-up examinations, is warranted in order to avoid recurrence of posterior segment inflammation. Therefore, the patient used 1,2mg/kg/day of oral prednisone at the diagnosis and slow tapered in the following 9 months.

4 CONCLUSION

It is very important to recognize Vogt-Koyanagi-Harada disease early and to start appropriate treatment—most commonly high-dose of corticosteroids— since preventing long-term visual loss and other potential complications depends on starting treatment early. The prognosis is generally good with continued immunosuppression. Follow-up is important to identify and act on any complications**.**

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