JIMKI
JURNAL ILMIAH
MAHASISWA KEDOKTERAN
INDONESIA
e-ISSN : 2721-1924 p-ISSN : 2302-6391

Laporan Kasus

CRACKING THE CODE: BILATERAL OPTIC DISC SWELLING WITH SEROUS RETINAL DETACHMENT AS A CLUE OF VOGT-KOYANAGI-HARADA DISEASE

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Abstract

***Background**: Vogt–Koyanagi–Harada disease (VKH) is a rare autoimmune disorder that attacks melanocyte-containing tissues, particularly the eyes, and can involve the central nervous system, hearing, and skin. VKH is one of the leading causes of non-infectious panuveitis in pigmented populations, and delayed diagnosis can lead to permanent visual impairment. Therefore, early detection and appropriate corticosteroid therapy are crucial to prevent long-term visual impairment. The purpose of this case report is to describe the clinical manifestations, supporting examinations, and response to high-dose corticosteroid therapy in a patient with suspected VKH disease. **Case Illustration**: A 39-year-old male presented with one week of progressive bilateral visual loss accompanied by fever, headache, and tinnitus. Examination revealed anterior chamber cells and flare, optic disc swelling, and serous retinal detachment. Optical coherence tomography confirmed subretinal fluid with septa formation. The patient was treated with high-dose intravenous methylprednisolone followed by oral tapering, resulting in significant visual improvement and stable recovery over one year. **Discussion:** The combination of bilateral ocular inflammation, optic disc swelling, serous retinal detachment, and systemic prodromal symptoms was consistent with the acute uveitic phase of probable VKH disease. Multimodal imaging facilitated early diagnosis and treatment, contributing to favorable outcomes. **Conclusion:** Early diagnosis and prompt initiation of high-dose corticosteroid therapy are crucial in optimizing visual outcomes and preventing long-term complications in VKH disease.*

Keywords: Corticosteroid, Optic Disc Swelling, Retinal Detachment, Vogt-Koyanagi-Harada Disease.

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1. INTRODUCTION

Vogt-Koyanagi-Harada (VKH) disease is currently defined as an uncommon granulomatous inflammatory disorder that targets melanocyte-rich tissues, chiefly involving ocular, auditory, neurologic, and cutaneous systems. This disease is a rare multisystem autoimmune disorder that primarily affects individuals with dark skin pigmentation, such as Asian, Middle Eastern, and Native American populations. VKH accounts for approximately 7–8% of all uveitis cases in Asian populations. Although its prevalence varies between regions, VKH remains one of the leading causes of non-infectious panuveitis in areas with high melanin pigmentation.[1,2] The disease typically originates in the eye, where melanocytes residing in the choroidal stroma become the primary focus of immune-mediated inflammation. If left untreated in the early stage, the process may extend beyond the choroid to involve the optic disc and retina.[3,4]

This disease is classified as probable, incomplete, and complete. Probable VKH is limited to ocular manifestations without extraocular manifestation. Incomplete VKH shows typical ocular manifestations with only one systemic involvement (neurological or auditory or integumentary). Complete VKH is characterized by bilateral granulomatous uveitis accompanied by neurological/auditory and integumentary involvement. There are four clinical stages of VKH, namely the prodromal phase, acute uveitic phase, chronic convalescent phase, and chronic recurrent phase. This staging supports accurate diagnosis and management. This approach has been proposed to improve clinical recognition, therapeutic decision-making, and patient outcomes.[4,5,6] Management of VKH relies principally on systemic medical treatment, most often with high-dose corticosteroids and when necessary, adjunctive immunosuppressive agents. Early identification of VKH disease is crucial to prevent irreversible ocular damage and optimize therapeutic outcomes to improve prognosis.[1]

The aim of this case report is to present the clinical features, diagnostic evaluation, and management of a patient with probable VKH disease, emphasizing the importance of early recognition and prompt high-dose corticosteroid therapy to improve prognosis and preserve visual function.

2. CASE ILLUSTRATION

A 39-year-old male presented with complaints of progressively blurry vision in both eyes for one week

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prior to examination. The visual disturbance was accompanied by systemic symptoms including fever, headache, and tinnitus, as well as ocular discomfort characterized by glare and pain upon eye movement. The patient denied any history of trauma, previous ocular surgery, or systemic comorbidities, and no regular medication was reported.

On ophthalmological examination of both eyes, the best-corrected visual acuity was counting fingers at 1 meter in the right eye and hand movement in the left eye. Slit-lamp biomicroscopy revealed cells and flare in the anterior chamber of both eyes, mid-dilatation with decreased pupillary light reflexes. The eyelids, conjunctiva, cornea, iris, and lenses were within normal limits. Posterior segment examination revealed vitreous cells. funduscopic evaluation revealed bilateral optic disc swelling, tortuous retinal veins, loss of macular reflex, and evidence of exudative retinal detachment (Figure 1). Optical coherence tomography (OCT) of the optic nerve head and retinal nerve fiber layer showed marked thickening in both eyes, while macular OCT demonstrated subretinal fluid with septa formation and choroidal folds, consistent with serous retinal detachment (Figure 2).

Figure 1. Funduscopic Evaluation

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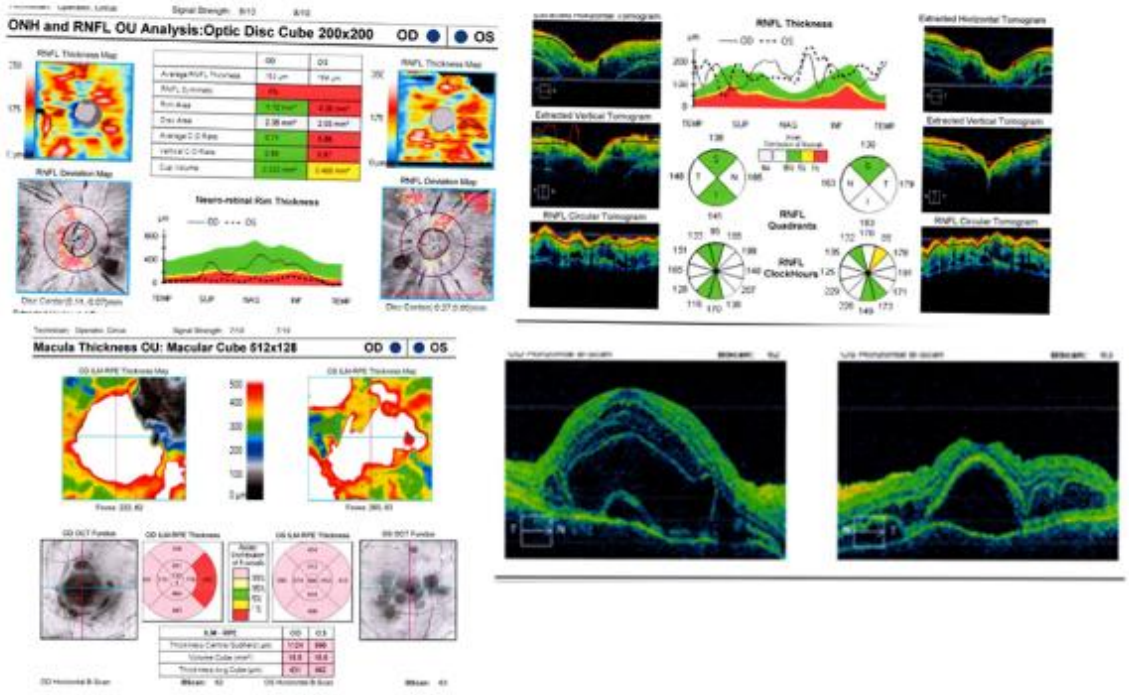

Figure 2. OCT Examination

Ancillary investigations included a broad laboratory panel to exclude infectious and autoimmune causes. Serology for cytomegalovirus, toxoplasma, syphilis (VDRL, TPHA), antinuclear antibody, rheumatoid factor, and anti-aquaporin-4 antibody were negative. Neuroimaging with magnetic resonance imaging (MRI) excluded optic neuritis and space-occupying lesions (Figure 3). Based on the constellation of ocular findings, systemic symptoms, and exclusion of other etiologies, the diagnosis of probable VKH disease was established. Bilateral optic disc swelling accompanied by serous retinal detachment is an important early clinical clue to VKH disease.

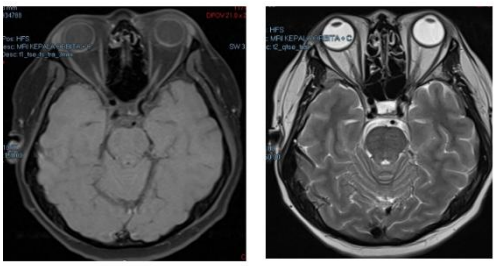

Figure 3. MRI Examination

The patient was initiated on high-dose intravenous (IV) corticosteroid therapy consisting of methylprednisolone 250 mg administered four times daily for three consecutive days. This was followed by a slow tapering regimen of oral methylprednisolone at 1 mg/kg/day, reduced weekly in accordance with clinical response.

On follow-up examination one week after initiation of methylprednisolone IV, the

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patient's visual acuity showed improvement. The right eye improved to 6/120, with pinhole (PH) correction reaching 6/15. The left eye improved to 6/21, with pinhole (PH) correction up to 6/12. Slit-lamp biomicroscopy revealed cells and flare in the anterior chamber of both eyes, mid-dilatation with decreased pupillary light reflexes. The eyelids, conjunctiva, cornea, iris, and lenses were within normal limits. Posterior segment examination revealed vitreous cells. Funduscopic examination revealed persistent optic disc hyperemia, tortuous retinal veins, and the exudative retinal detachment showed signs of resolution in both eyes. Color vision assessed with the Ishihara test showed 5/21 correct plates in the right eye and 8/21 in the left eye, and contrast sensitivity measured 0.60 in the right eye and 1.05 in the left eye, respectively.

Repeat fundus photography one week after methylprednisolone IV therapy demonstrated resolution of the exudative retinal detachment (Figure 4), while Humphrey visual field testing revealed only mild visual field defects (Figure 5). Follow-up OCT ONH-RNLF demonstrate increasing of the RNFL thickness, and OCT macula showing resolve of the exudative retinal detachment (Figure 6).

Figure 4. Funduscopy Photograph One Week After IV Corticosteroid Therapy

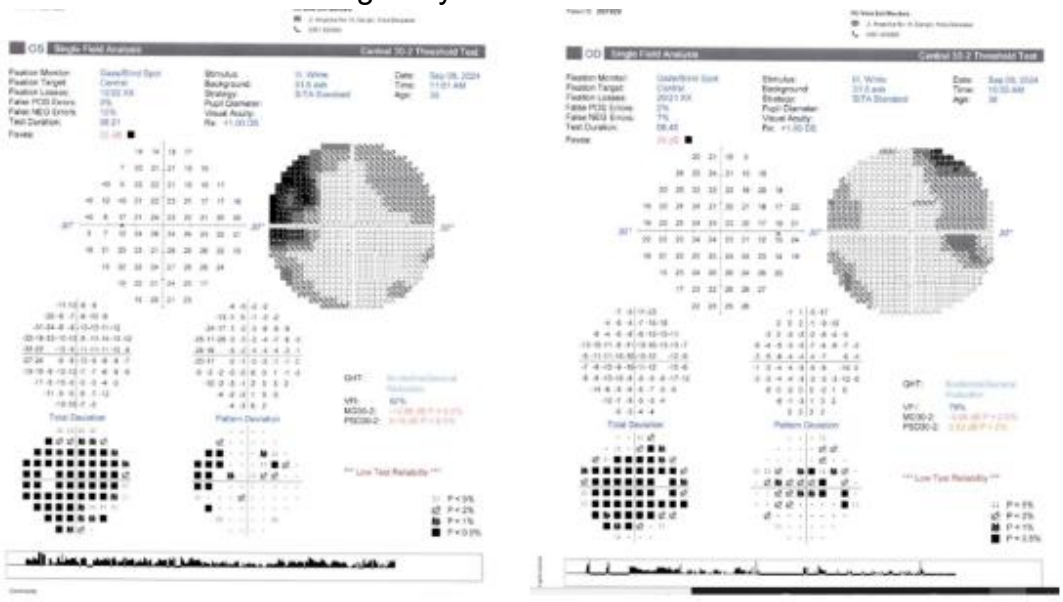

Figure 5. Humphrey Visual Field One Week After IV Corticosteroid Therapy

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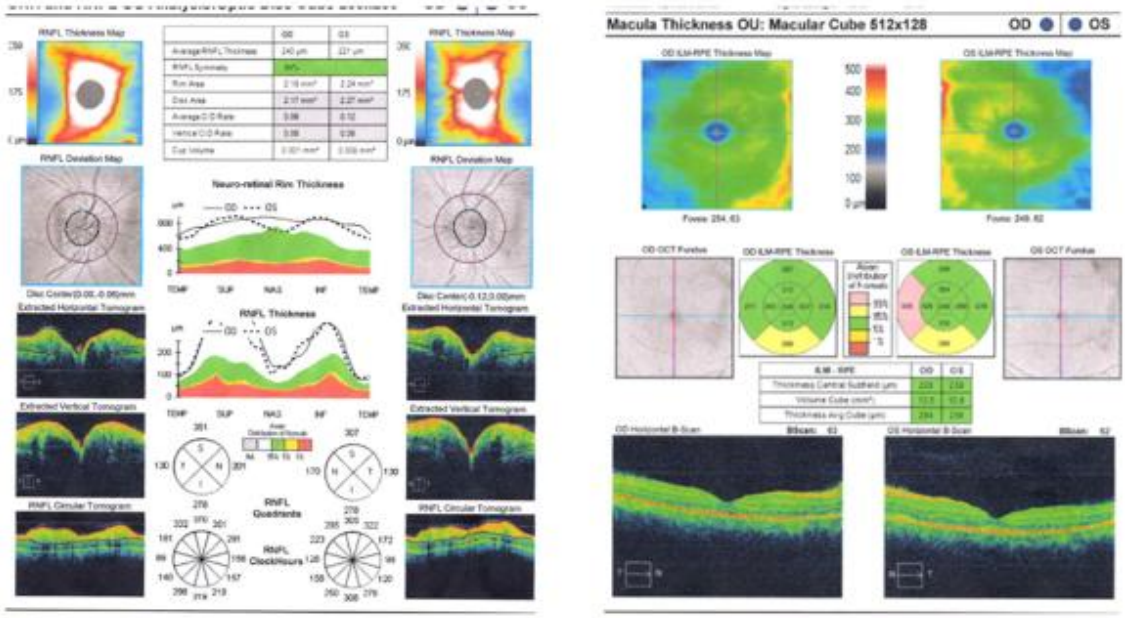

Figure 6. OCT, ONH-RNFL, and Macula One Week After IV Corticosteroid Therapy

At the one-month follow-up visit, the patient demonstrated further visual improvement. Best-corrected visual acuity reached 6/18 PH 6/7.5 in right eye and 6/12 PH 6/7.5 in left eye. Ishihara test improved to 15/21 plate in right eye 17/21 plate in left eye.

Contrast sensitivity test demonstrated 1.05 in right eye and 1.20 in left eye. By the second month of follow-up, the patient vision improve, achieving 6/15 PH 6/7.5 in the right eye and 6/12 PH 6/7.5 in the left eye. Ishihara test showed 19/21 plate in right eye 21/21 plate in left eye. Contrast sensitivity test demonstrated 1.35 in right eye and 1.50 in left eye.

At the three-month follow-up, the patient reported worsening blurry vision, with visual acuity reduced to 6/18 in the right eye and 6/7.5 in the left. Funduscopy revealed recurrent optic disc swelling and exudative retinal detachment, prompting an increase dose of methylprednisolone from 4 mg to 16 mg and initiation of azathioprine (Imuran) 120 mg/day after rheumatology department consultation. By the fourth month of observation (one month after combined methylprednisolone and azathioprine therapy), visual acuity improved to 6/12 in the right eye and 6/6 in the left. Funduscopy revealed optic disc swelling with resolution of the exudative retinal detachment. Ishihara testing showed 21/21 plates bilaterally, and contrast sensitivity improved to 1.35 in the right eye and 1.50 in the left eye, confirming substantial functional recovery. Serial Humphrey visual

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field (HVF) 30-2 testing demonstrated progressive functional recovery, with gradual improvement of visual field defect on both eyes (Figure 7). The reliability of the examinations also improved over time, reflecting consistent visual performance with treatment.

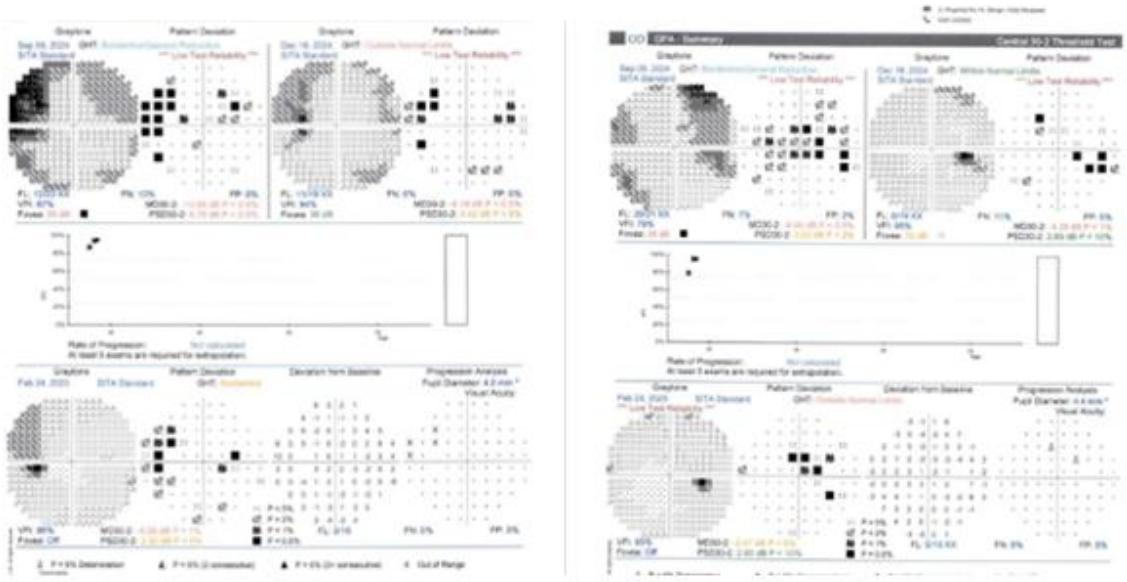

Figure 7. Serial HVF 30-2 report

Longitudinal monitoring over one year showed sustained recovery. At the one-year follow-up (final follow-up), best-corrected visual acuity was 6/6 in both eyes. Anterior segment examination was normal, and funduscopy demonstrated slight hyperemia of the optic disc with resolution of the exudative retinal detachment (Figure 8).

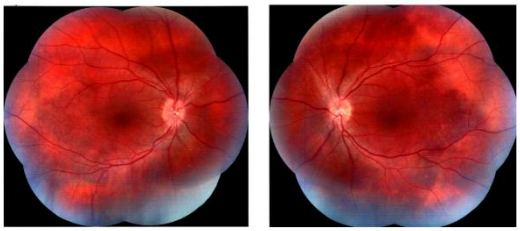

Figure 8. Funduscopy Photograph After One Year Follow-Up

3. DISCUSSION

The clinical spectrum of VKH encompasses ocular, neurologic, cutaneous, and auditory manifestations.[7,8] Early ocular signs include multifocal serous retinal detachment, iridocyclitis, and optic disc swelling.[9] Patient in this case report presented with bilateral optic disc swelling, serous retinal detachment, and signs of active uveitis, mirroring the early ocular spectrum of VKH, and classified as probable VKH because the patient fulfilled only the essential criteria and bilateral ocular involvement without extraocular manifestations.

The precise cause of VKH disease has not yet been fully elucidated,

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although prevailing hypotheses suggest an autoimmune mechanism in which T lymphocytes mount a response against melanocytes, often following exposure to or recovery from viral infection. Among the viruses investigated, cytomegalovirus and Epstein-Barr virus (EBV) have been considered potential contributing factors, though a conclusive association has not been established. The fundamental process is believed to involve a breakdown of immunological tolerance toward melanocytes, resulting in a non-necrotizing granulomatous inflammatory reaction affecting ocular tissues, as well as the inner ear, skin, and hair.[10,11]

Epidemiologically, VKH most commonly presents in individuals during their third and fourth decades of life, although the reported age range extends from as young as three years to late adulthood.[12] Pediatric-onset disease is often characterized by a more severe and protracted course, with a greater likelihood of complications such as subretinal fibrosis. In contrast, elderly patients, particularly those over 65 years of age, tend to present with optic disc hyperemia and choroidal detachment more frequently. A sex predilection has also been noted, with women affected more often than men.[13,14] The patient in this case is a 39-year-old male, fitting within the most common age bracket for VKH onset but representing the less frequent male demographic.

This disease typically progresses through four distinct clinical phases. The prodromal phase often resembles a flu-like syndrome. Within one to two days, patients frequently develop visual disturbances including blurred vision, photophobia, ocular discomfort, and conjunctival hyperemia. Some individuals also report hypersensitivity of the skin, scalp, or hair to light touch during this period. The subsequent uveitic phase is characterized by the appearance of inflammatory cells within the anterior chamber, vitreous cavity, or both. Choroidal thickening becomes evident, accompanied by optic disc edema and hyperemia. Ophthalmoscopic examination may reveal multiple serous retinal detachments or subtle macular folds; in more severe cases, bullous detachment can occur.[15,16] Over several months in the chronic convalescent phase, progressive depigmentation of the fundus may emerge. This phase characterized by the disappearance of active inflammation with residual manifestations such as depigmentation of the ciliary body (sunset glow fundus) and retinal atrophy. Meanwhile, the chronic recurrent phase is characterized by the recurrence of inflammation, particularly chronic

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granulomatous anterior uveitis, which can cause progressive ocular complications.[8] The constellation of systemic manifestations experienced by the patient, including fever, headache, and tinnitus, together with ocular complaints such as blurry vision, glare and pain on eye movement, is consistent with the stage of VKH disease. The patient in this report exhibited many of these acute manifestations, including ocular pain, glare, bilateral optic disc swelling, and exudative retinal detachment, consistent with VKH's acute uveitic phase.

The diagnosis of VKH disease is primarily clinical, requiring the recognition of a characteristic constellation of ocular, neurological, auditory, and integumentary manifestations, which can be further substantiated by laboratory and imaging studies. On examination, slit-lamp evaluation may reveal features of inflammation.[17,18]

Among available imaging modalities, indocyanine green angiography (ICGA) is considered the most valuable technique for both diagnosis and longitudinal monitoring of VKH. ICGA provides superior visualization of the choroid and retinal pigment epithelium, which are principal sites of pathology in early disease. Due to its longer wavelengths, ICGA offering a sensitivity approaching 90–100% for VKH detection. ICGA has limitations because it is invasive, requires intravenous injection of contrast agents, and not always available in health facilities. [17,19,20] As an alternative, OCT while less invasive, plays a complementary role in the multimodal diagnostic workup of VKH. OCT can identify characteristic features such as multilayered subretinal fluid, septations, and dynamic changes in choroidal thickness and volume.[21,22] In this reported case, multimodal ocular imaging was critical and OCT is useful in establishing a diagnosis.

Several important differential diagnoses were considered, including optic neuritis, papilledema, and neuroretinitis. However, these were excluded based on the presence of bilateral uveitis, serous retinal detachment and optic disc swelling, with favorable response to corticosteroid therapy, which are more consistent with VKH disease.[23]

Therapy for VKH disease has been high-dose IV corticosteroid pulse therapy, used in acute VKH to rapidly suppress severe inflammation, followed by oral corticosteroids with gradual tapering to maintain remission. Dose adjustment is necessary to prevent relapse and preserve visual function. The onset of treatment initiation was directly

related to the likelihood of recurrence.[23,24] Importantly, because VKH is a multisystem disease, local ocular treatment alone does not influence extraocular manifestations. Consequently, the use of systemic steroid are best reserved for patients with systemic features.[25,26]

Immunomodulatory therapy has become an essential adjunct in VKH management, particularly in patients with recurrent or chronic disease. A wide range of agents has demonstrated efficacy.[27,28] Adalimumab has recently emerged as a promising option for refractory disease, particularly in patients with corticosteroid-related complications or in difficult-to-treat pediatric cases.[29] Surgical intervention in VKH is generally limited to addressing complications secondary to chronic inflammation or corticosteroid exposure. Common procedures include cataract extraction, glaucoma surgery for IOP control.[30,31] The administration of high-dose IV corticosteroids with vigilant tapering in this patient is in accordance with the recommended first-line therapy for acute VKH disease.

4. CONCLUSION

This case highlights the importance of early recognition of VKH disease in patients presenting with bilateral optic disc swelling and serous retinal detachment. Prompt initiation of high-dose systemic corticosteroids in the acute phase can significantly improve visual prognosis. Adjunctive use of OCT and visual field testing proved valuable in monitoring therapeutic response, while MRI was crucial in excluding alternative neuro-ophthalmic conditions.

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