

## Case Report

### Early phase Vogt-Koyanagi-Harada Disease in Nepalese Elderly woman: A case report

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#### Abstract

**Introduction:** Vogt–Koyanagi–Harada (VKH) disease is defined as an autoimmune disorder characterized by bilateral granulomatous panuveitis with systemic manifestations, such as tinnitus, vertigo, and meningism caused by melanocyte antigen-reactive T-cells. Majority of VKH patients present at the age between 20 and 50 years. VKH is uncommon in elderly and challenging to manage. VKH is one of the important differential diagnosis of bilateral pan uveitis

**Case:** A 65 year/ female brought with chief complaint of sudden loss of vision in both eyes, headache and hearing problem for 1 month. She didn't give any history of other systemic illness, ocular surgery, ocular trauma, chronic use of medicament. Her visual acuity was hand movement with accurate projection of rays (HM) in both eyes. The intraocular pressure (IOP) was 12mmHg in both eye. Slit-lamp bio microscopy revealed features of Pan uveitis in both eye. Systemic work up revealed no any other abnormalities. A diagnosis of early phase VKH was made and treated with intravenous pulse steroid therapy followed by tapering dose of oral steroid along with immunomodulator resulting in a very good visual recovery.

**Conclusion:** VKH can present in elderly. immunomodulator should be considered in elderly to prevent side effect of steroid along with recurrence of inflammation.

**Key words:** Vogt-Kayanagi disease, Immunomodulator, Nepal, Elderly.

#### Introduction

Vogt–Koyanagi–Harada (VKH) disease is defined as an autoimmune disorder characterized by bilateral granulomatous panuveitis with systemic manifestations, such as tinnitus, vertigo, and meningism caused by melanocyte antigen-reactive T-cells (O'Keefe

and Rao, 2017). According to Kitamura (2005) VKH disease can occur in any period of life, from childhood to adulthood, but the majority of patients are between the age of 20 and 50. Yamamoto (2007) observed VKH on elderly Japanese although it is very uncommon in this age group. There is not any report of VKH reported in Nepalese elderly patient in literature.

Early phase VKH as defined by Yang et al (2018) has a better visual prognosis than late phase VKH. The mainstay of VKH disease treatment is early administration of Pulse corticosteroids therapy, but Yamamoto (2007) believes that it is more difficult to treat elderly

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patients than younger patients with high-dose corticosteroids because of the side effects. The visual prognosis of this disease lies in the early diagnosis and consequent use of corticosteroids and immunomodulators. The complications including cataract, glaucoma and choroidal neovascularization (CNV) and sub retinal fibrosis, together with its recurrence and chronicity, are the theft of sight in VKH disease as reported by Marcelo (2018).

### Case Report

A 65-year-old female, farmer residence of Rolpa District brought by her grandson with chief complaint of 'sudden loss of vision in both eyes 4 weeks back' following mild headache and hearing problem. It was associated with mild ocular pain and redness of eye. She didn't give any history of other systemic illness, ocular surgery, ocular trauma, chronic use of medicament. She was examined and treated as anterior uveitis with topical steroid and cycloplegics by an ophthalmic assistant at primary eye care centre 2 weeks back as per the out-patient department card with patients before presenting to us due to lack of improvement on vision. Clinical findings were not mentioned in that card.

On clinical examination, her visual acuity was Hand movement with accurate projection of rays (HM) in both eyes. The intraocular pressure (IOP) was 12mmHg in both eyes with Goldmann applanation tonometer (GAT).

Slit-lamp bio microscopy of right eye revealed diffuse conjunctival congestion, fine keratic precipitates along arlt's triangle 3+ cells and flare in anterior chamber as per Standardised Uveitis Nomenclature (SUN) grading system(SUN working group, 2005) , posterior synechiae involving 5 clock hour infero-nasally with pigment on anterior surface of clear lens. Left eye examination revealed diffuse conjunctival congestion, fine keratic precipitate along arlt's triangle, with 3+ cells and flare in

anterior chamber as per SUN grading system (Figure 1) (SUN working group, 2005). There were absence of iris nodules, iris atrophy and iris neovascularisation on both eyes.

Fundus evaluation revealed bilateral disc edema with serous macular detachment with few nummular white lesions in the right eye suggesting Dalen Fuch's nodule (Figure 2). Vitritis was absent in both eyes. Optical coherence tomography (OCT) also revealed serous retinal detachment at the macular area with a retinal thickness of 409  $\mu$ m and 539  $\mu$ m in OD and OS respectively with irregular elevation of retinal pigment epithelium suggesting choroidal pathology (Figure 3).

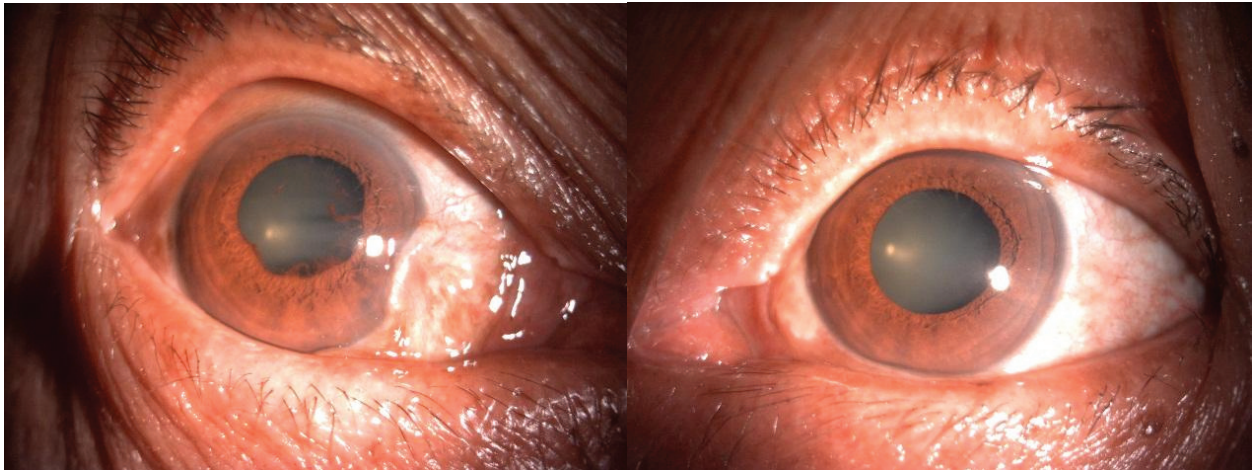
Markers of inflammation and autoimmunity including erythrocyte sedimentation rate, C-reactive protein , serum angiotensin converting enzyme level, total and differential leukocyte count were negative as were the serological testing for the diagnosis of infectious pathologies including HIV, HBsAg, HCV, TPHA and mantoux test. The review of systems revealed Headache and mild hearing problems. Pure tone audiometry revealed mid sensorineural hearing loss. Thus, this patient was diagnosed to have early phase VKH disease on the basis of diagnostic criteria of VKH disease given by Yang et al (2018).

High-dose pulse steroid therapy (intravenous methylprednisolone 1,000 mg/day for 3 days) was given along with topical steroid and cycloplegics. On the day of discharge, an increase of the VA to 6/60 (OD) and 6/36(OS) has been observed.

The patient was prescribed with oral prednisolone 50 mg once daily after breakfast for 2 weeks then tapered 10 mg/week till 7<sup>th</sup> week with pantoprazole 40 mg once a day. At the end of 1 week her visual acuity was 6/24 in both eyes, no conjunctival congestion and KP, 1 + cells and flare in anterior chamber and reduced macular retinal detachment. Three

weeks after the beginning of oral steroid therapy, the patient was prescribed with immunosuppressive medication Azathioprine 50mg once a day. At 4 weeks after presentation, Her BCVA improved to 6/9 both eyes, anterior segment inflammation subsides (figure 4),

serous macular detachment resolved (Figure 5 & 6) and disc edema was also resolving. There was splinter hemorrhage in the left disc which appeared on follow-up visit. Patient was symptom free on tapering dose of oral steroid and azathioprine.

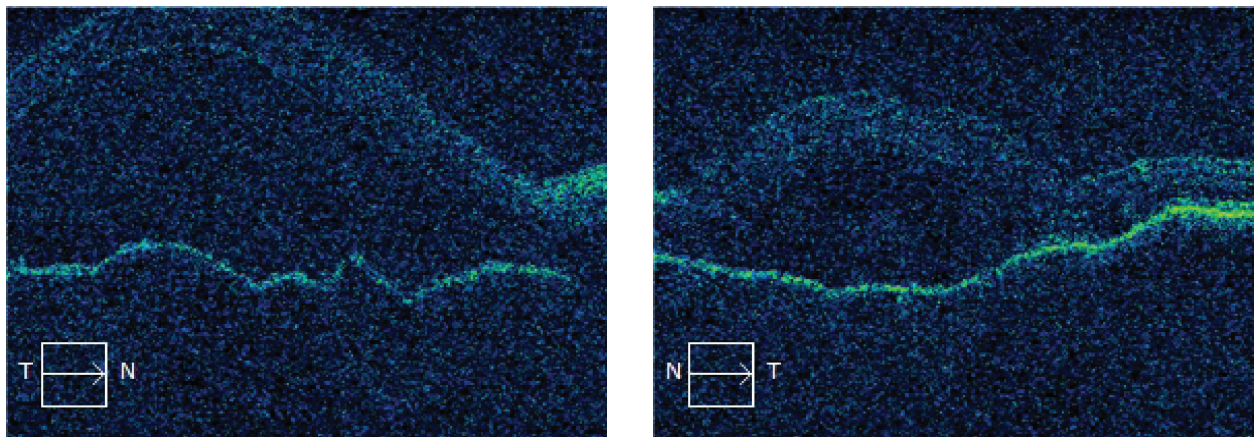


**Figure 1:** Anterior segment photo showing conjunctiva congestion in both eye and posterior synechiae in right eye

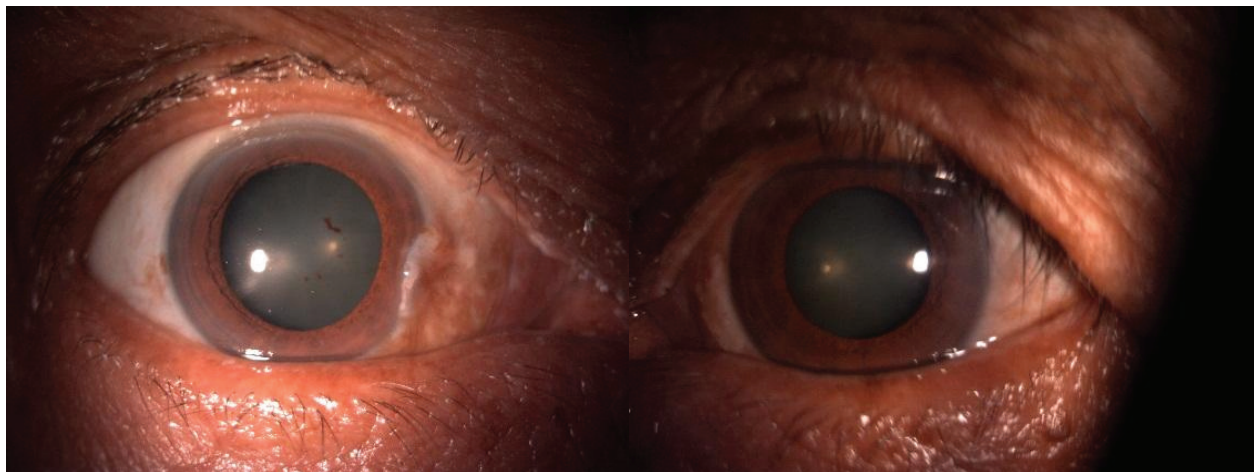


**Figure 2:** Fundus photo showing disc edema and serous macular retinal detachment in both eye

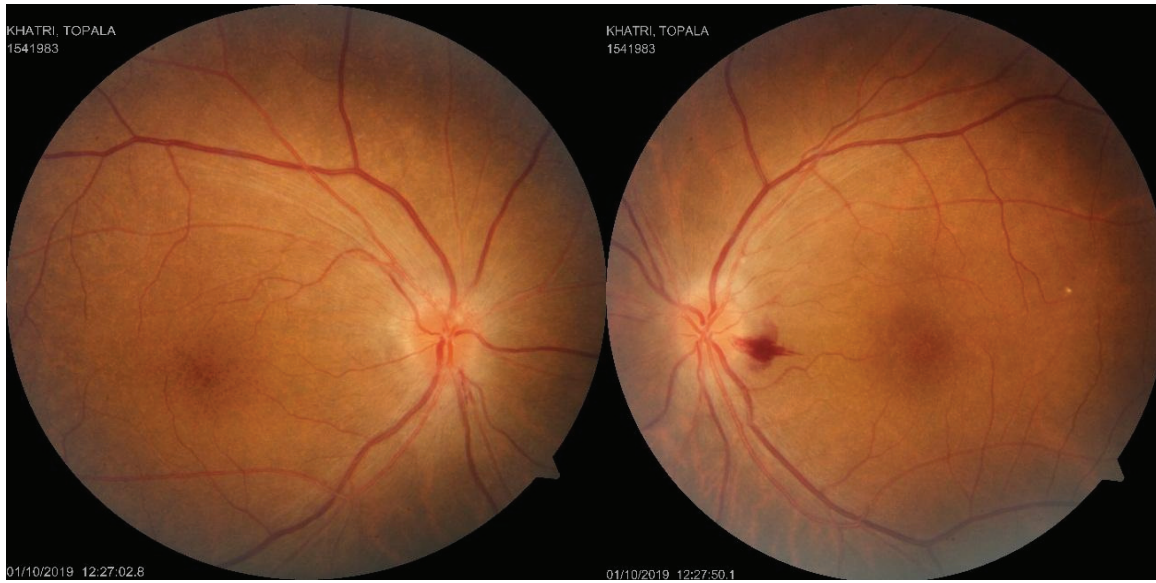




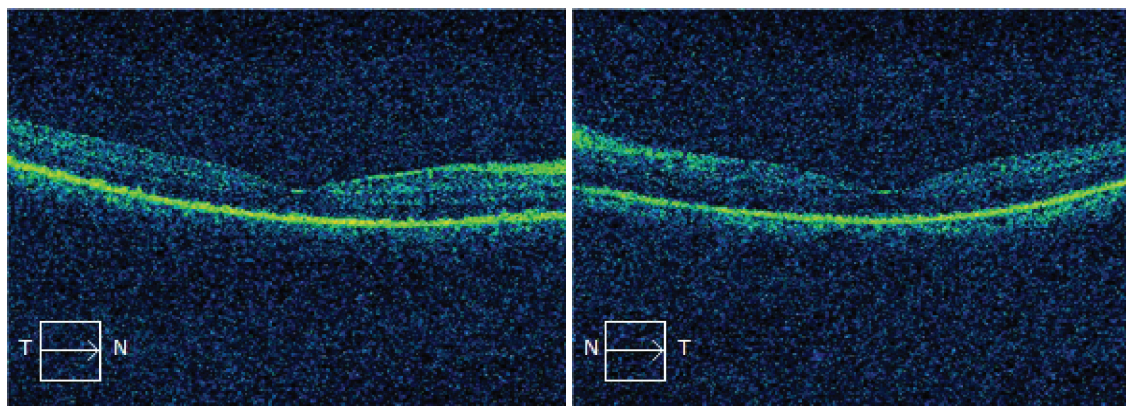
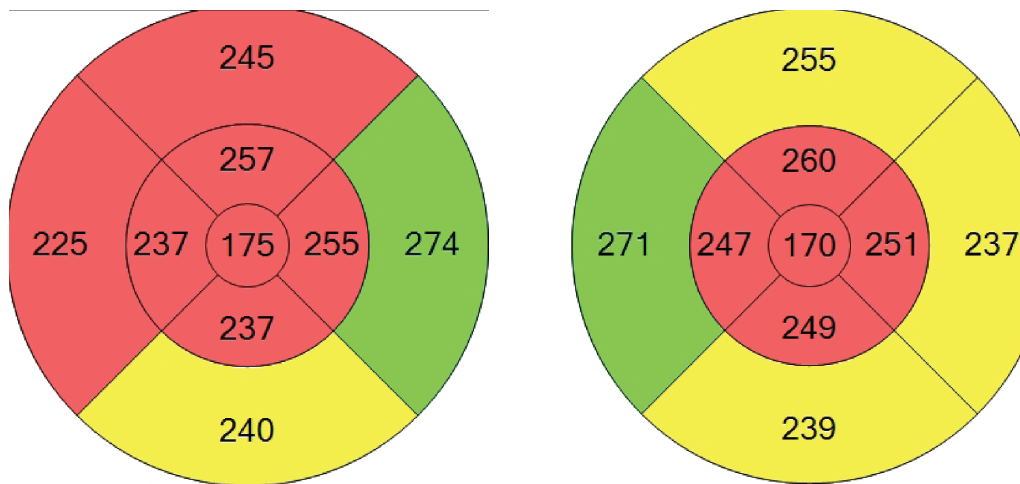
**Figure 3:** OCT showing serous sub retinal fluid with increased foveal thickness



**Figure 4:** Anterior segment Photo after treatment showing resolved sign of inflammation with pigment on anterior lens surface



**Figure 5:** Fundus photo after treatment showing resolved disc edema and serous retinal detachment. Left eye showed disc hemorrhage



**Figure 6:** OCT showing normal foveal contour with reduced foveal thickness after treatment



## Discussion

Early recognition of VKH and immediate institution of intravenous pulse steroid therapy is the key to successful visual recovery in this disease. Delay in the diagnosis is a known problem in Nepal due to difficult geography, lack of transportation facility, poverty, advanced age of patient, lack of skilled eye health care provider especially general ophthalmologist and uveitis expert and equipped eye hospital in rural Nepal.

Presenting complaint in our case was sudden loss of vision as shown by most of elderly cases in the case series of Yamamoto et al (2007) and Rodrigues-Barros et al (2018). We could observe only fine KP in our case whereas Rodrigues-Barros et al (2018) reported mutton fat KP in all cases. This finding in our case might be as a result of treatment for anterior uveitis which had already been started 2 weeks before presenting to us. Anterior uveitis was severe in this case as reported by other authors (Yamamoto, 2007; Rodrigues-Barros, 2018). Serous RD was also observed in all cases by Rodrigues-Barros et al (2018) as in our case.

Management of VKH in elderly is challenging due to higher incidence of steroid related side effects and associated morbidities (Yamamoto et al, 2007; Rodrigues-Barros et al, 2018). Our case received pulse steroid intravenously like in the case of other studies. The immunomodulator used in our case was azathioprine as used in Japan and Portugal in some cases but cyclosporine was used in most cases in Portugal (Yamamoto et al, 2007; Rodrigues-Barros et al, 2018). Final Visual acuity was 6/9 in both eye in our case similar to two of case in Portugal who were detected early but one of the cases in there series had no visual improvement due to proliferative vitreo-retinopathy as well as delayed treatment (Rodrigues-Barros et al, 2018). We detected weight gain as the only side effect of steroid therapy whereas other authors reported

systemic arterial hypertension, hyperglycemia, weight gain, osteoporosis and Hallucination too (Yamamoto et al, 2007; Rodrigues-Barros et al, 2018).

This case highlights that VKH is one of the differential diagnoses of bilateral pan uveitis even in elderly patient in Nepal. Steroid sparing agents should be considered to prevent relapse of VKH. This case also showed that, If VKH can be detected at an early phase it has very rewarding visual recovery.

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