Case Report

An Unusual Presentation of Uveo-meningeoencephalitic Syndrome

Prachi Abhishek Dave[1], Neha Bharti[2], Bhupesh Singh[2], Sudhank Bharti[2]
[1]Centre for Sight, Delhi, India
[2]Bharti Eye Hospital and Institute, Delhi, India

ABSTRACT

Introduction: Unilateral uveo-meningeoenchephalitic syndrome is a relatively rare entity. Our case highlights the importance of keeping this entity in mind when a unilateral picture suggestive of Vogt Koyanagi Harada syndrome crops up.

Case: A 34-year-old male came with chief complaints of blurring of vision in the right eye since two days with a prodrome of intense headache and redness in the right eye. On examination, the vision in the right eye was counting fingers close to face and 20/20 in the left eye. Clinical examination suggested unilateral uveo-meningeoenchephalitic syndrome which was confirmed on multimodal imaging.

Observations: This case highlights the fact that though uveo-meningeoenchephalitic syndrome is bilateral, by definition; the initial presentation may still be unilateral and a prompt diagnosis and treatment can prevent the involvement of the other eye.

Conclusion: Any case presenting with signs of symptoms suggestive of Vogt Koyanagi Harada should be treated as Vogt Koyanagi Harada even though the initial presentation may be unilateral. A prompt diagnosis and early treatment will ensure that the other eye does not get involved. Early Vogt Koyanagi Harada may just present with choroidal hyperpermeability and multiple septate pockets of SRF without any vitreous cells or anterior segment inflammation.

Key words: Choroid, Posterior uveitis, VKH syndrome.

Financial Interest : Nil
Conflict of Interest : Nil

Corresponding Author
Dr. Prachi Abhishek Dave
Consultant - Vitreoretina Services,
Centre for Sight Group of Hospitals,
Delhi, India.
E-mail: drpdjhala@gmail.com

Received : 20.12.2020
Accepted : 19.06.2022

Access this article online
Website: www.nepjol.info/index.php/NEPJOPH
DOI: https://doi.org/10.3126/nepjoph.v14i2.33643

ISSN: 2072-6805, **E-ISSN:** 2091-0320

INTRODUCTION

Uveo-meningeoenchephalitic syndrome or Vogt Koyanagi Harada (VKH) is an autoimmune multisystem disorder with bilateral granulomatous panuveitis and ENT, neurological and dermatological manifestations (Burkholder, 2015). VKH is seen in adults, commonly women (Mota and Santos, 2010). It has 4 phases: prodromal phase featuring neurologic and auditory manifestations such as headache, tinnitus, neck stiffness; acute uveitic phase showing diffuse choroiditis, disc swelling and exudative retinal detachment; chronic recurrent phase showing granulomatous anterior uveitis and the convalescent phase characterised by sunset glow fundus (Burkholder, 2015).

Rao et al (Rao et al, 2010) conducted a study on ethnically and geographically diverse groups of patients and concluded that two clinical signs are specific to VKH-exudative RD and sunset glow fundus. Though VKH is a bilateral disease, involvement of the other eye can be sequential (Beniz et al, 1991). VKH may have variable anterior and posterior segment inflammation but it primarily begins in the choroid so a prompt diagnosis and treatment may completely prevent an anterior and vitreous reaction (Lodhi, Reddy and Peram, 2017). Our case is unique since it has signs and symptoms typical of VKH but the involvement is unilateral whereas VKH is conventionally described as a bilateral disease. The sequential involvement of the other eye was prevented in our case due to a prompt diagnosis and treatment.

CASE REPORT

A 34-year-old male came with complaints of blurring of vision in the right eye (OD) since 2 days. He gave a history of intense headache and redness of the right eye 2 days prior to loss of vision. He had no significant past or medical history. The best corrected visual acuity (BCVA) in OD was counting fingers close to face and 20/20 in the left eye (OS). On examination, the anterior segment was unremarkable in both eyes. A dilated fundus examination of OD revealed a clear vitreous with no vitreous inflammation, disc edema with hyperemia and multiple serous detachments all over the fundus (Figure 1A). OS was within normal limits (Figure 1B). The patient was advised a fundus fluorescein angiography (FFA), Indocyanine green angiography (ICGA) and optical coherence tomography (OCT). Patient did not consent to an ICGA. Multimodal imaging (Figure 2A, 2B and 2C) of OD was suggestive of unilateral VKH syndrome. After ruling out an infectious pathology the patient was diagnosed to have incomplete unilateral VKH and was started on intravenous methylprednisolone 1 gm daily for 3 days followed by oral prednisolone (1mg/kg/day) with a slow taper. His systemic parameters were periodically evaluated. He improved clinically (Figure 3A) and showed a small residual pocket of SRF on OCT at the end of 1 month (Figure 3B) which completely resolved at the end of 2 months (Figure 3C) with a final visual acuity of 20/40 in OD. He was continued on a slow taper of oral steroids for 6 months.

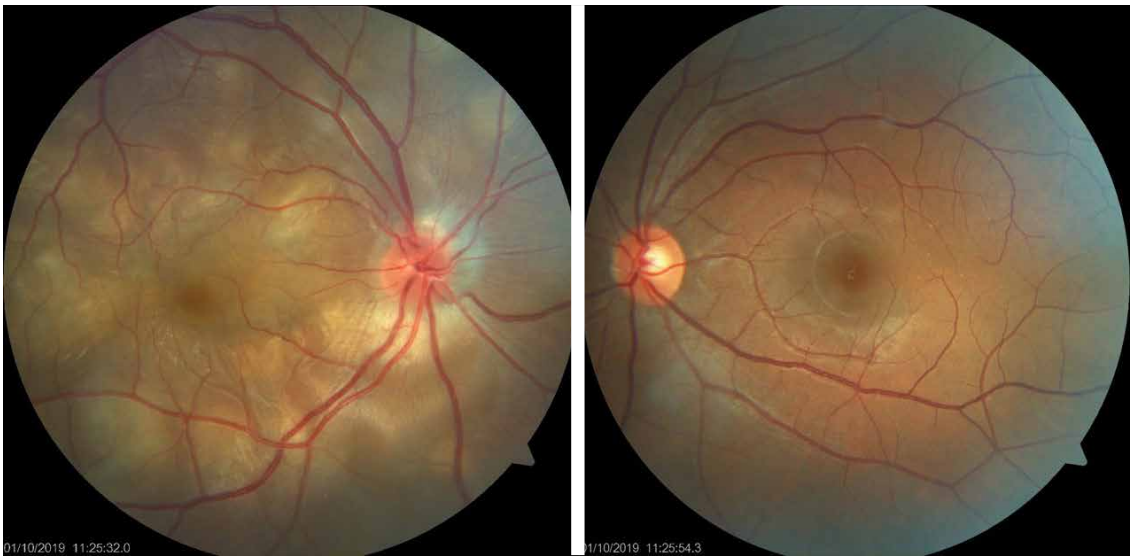

Figure 1A: Colour fundus photograph of the right eye showing disc hyperemia with edema (white arrow) and multiple pockets of serous detachments of the retina (black arrows) suggesting VKH syndrome.

Figure 1 B: Colour fundus photograph of the left eye showing a normal fundus.

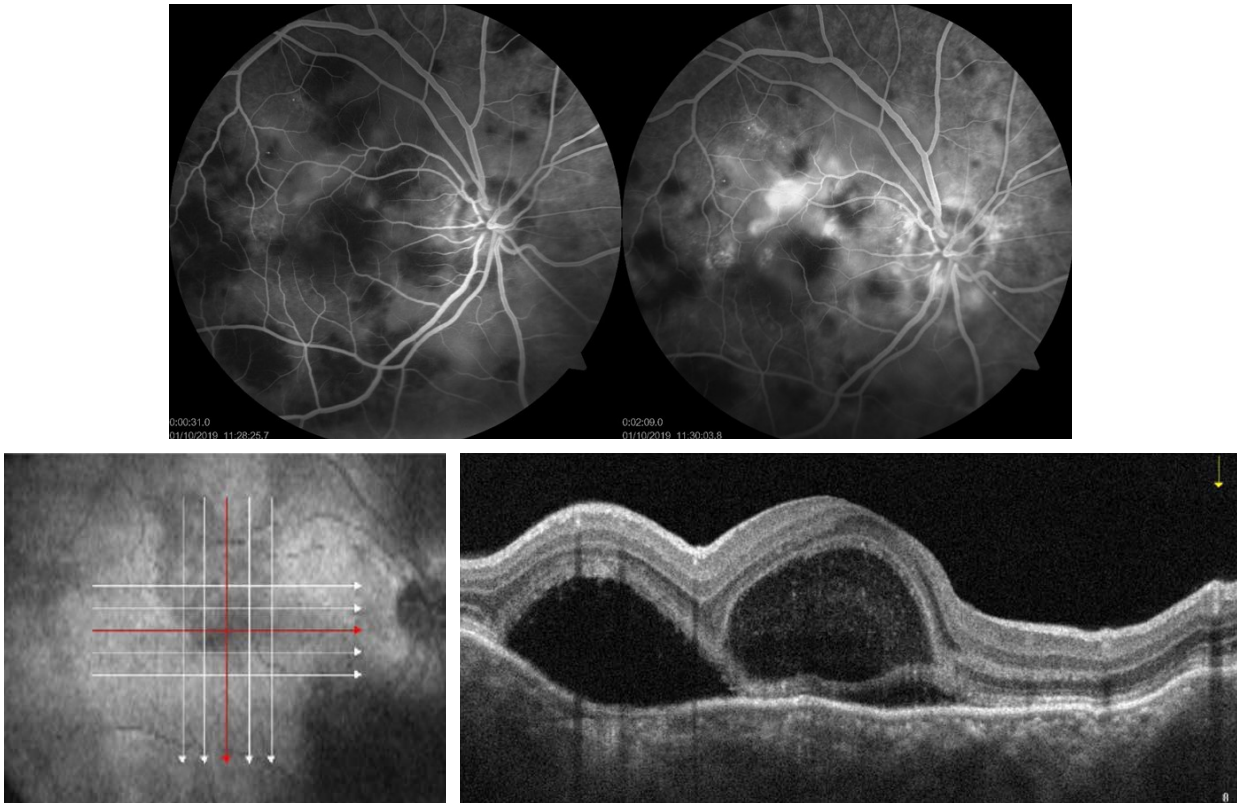

Figure 2A and 2B: FFA of the right eye showing multiple pinpoint leaks (Black arrows) with late pooling of the dye in the right eye (white arrows).

Figure 2C: OCT showing multiple pockets of SRF (white stars) with septae (white arrow) and an undulating RPE (black arrow).

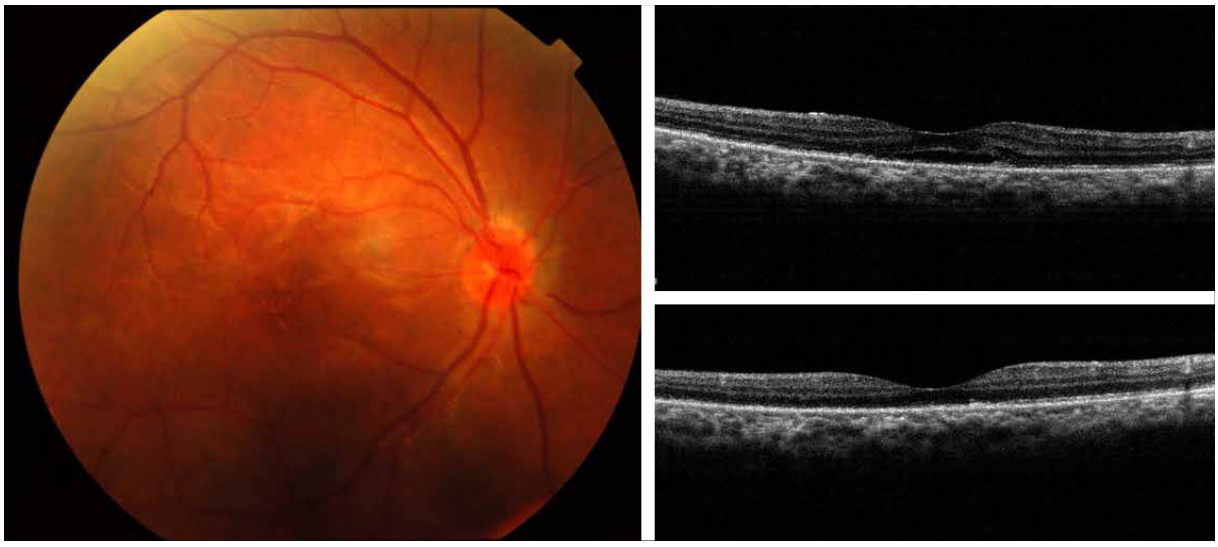

Figure 3A and 3B: Colour fundus and OCT picture of the right eye at the end of 1 month showing resolution of disc edema on colour fundus photograph and reduction in the srf pockets on OCT.

Figure 3C: OCT of the right eye at the end of 2 months showing complete resolution of the srf pocket.

He was a known partial hyperopic amblyopia in OD and claimed that he regained the vision he had before the episode. He was also evaluated systemically for any ENT, neurological and dermatological manifestations of VKH but none were found. Since this was the first episode, we did not start him on an immunosuppression and he has been kept on close follow up.

DISCUSSION

VKH is diagnosed as a bilateral disease with auditory, neurological and dermatological manifestations (Mota and Santos, 2010). However there are a few anecdotal case reports suggesting that VKH can present unilaterally with no extraocular manifestations. Our case adds to the anecdotal cases reported so far on unilateral VKH but differ from them in certain aspects.

The differential diagnosis of VKH includes other causes of posterior uveitis such as sympathetic ophthalmia (SO), syphilis, intraocular lymphoma, lymes disease and multifocal CSR. Absence of trauma and ocular surgery ruled out SO in our case. Intraocular lymphoma was ruled out as there was no history of weight loss, fever and the total, differential counts were normal. Our patient had disc hyperemia with multiple pockets of SRF along with multiple septae and RPE undulations on OCT. Bacillary layer detachments (BLD) which are typical of VKH were also noted in our patient on OCT. FFA also showed pinpoint areas of leak with pooling in late stage suggestive of VKH. The presence of these findings ruled out multifocal CSR.

Agarwal A et al presented a retrospective case series of 2 cases having unilateral VKH (Agarwal and Biswas, 2011). Their diagnosis of

VKH was clinical and they treated their cases with oral prednisolone alone. Another case report by Tsui et al reported VKH like features in only one eye and the patient responded to oral steroids (Tsui et al, 2018).

Our case differed from other cases reported in literature as he did not have any anterior or vitreous chamber reaction. This may be because VKH starts in the choroid and then progresses on to involve the vitreous and anterior segment. Since our patient presented early, he only had choroidal manifestations. It has been reported that properly treated VKH may not have dermatological involvement thereby explaining no dermatological findings in our case (Burkholder, 2015). Severe headache and eye injection could indicate the prodromal symptoms noted in VKH.

Our case highlights the fact that the bilaterality in VKH may be sequential and a prompt diagnosis and appropriate therapy might prevent the involvement of the other eye. We believe that unilateral VKH is an underreported entity which can be easily picked up by maintaining a high degree of suspicion.

CONCLUSION

This case adds to the few cases of unilateral VKH reported so far. Though a rare clinical presentation, VKH should be considered in the differential diagnosis of unilateral posterior uveitis as well.

REFERENCES

Agarwal A, Biswas J.(2011) Unilateral Vogt-Koyanagi-Harada disease: report of 2 cases in Middle east African journal of ophthalmology,18,82-84. doi: 10.4103/0974-9233.75898

Beniz J,Forster DJ,Lean JS, Smith RE, Rao NAA.(1991) Variations in clinical features of the Vogt-Koyanagi-Harada syndrome in Retina, 11,275-280. doi: 10.1097/00006982-199111030-00001

Burkholder BM. (2015)Vogt-Koyanagi-Harada disease in Curr opin ophthalmol, 26, 506-511. doi: 10.1097/ICU.0000000000000206.

Lodhi SA, Reddy JM,Peram V.(2017) Clinical spectrum and management options in Vogt-Koyanagi-Harada disease in Clin Ophthal,11,1399-1406.doi: 10.2147/OPTH.S134977

Mota LA,Santos AB.(2010) Vogt-Koyanagi-Harada syndrome and its multisystem involvement in Rev Assoc Med Bras,;56, 590-595. doi: 10.1590/s0104-42302010000500023.

Narsing A Rao, Amod Gupta, Laurie Dustin, Soon Phaik Chee, Annabelle A Okada, Moncef Khairallah, et al.(2010). Frequency of distinguishing clinical features in Vogt-Koyanagi-Harada disease. Ophthalmology. ,117,591-599. doi: 10.1016/j.ophtha.2009.08.030

Tsui E et al.(2018) Unilateral ocular manifestations of Vogt-Koyanagi-Harada disease in Ocular immunology and inflammation,26,1297-3000. doi: 10.1080/09273948.2017.1353638.