Case Report

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Multimodal imaging modalities in acute Vogt-Koyanagi-harada disease

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Introduction

Vogt-Koyanagi-Harada disease (VKH) is a severe bilateral granulomatous posterior or panuveitis. An acute case of VKH presents with gradual painful diminution of vision which may be associated with symptoms like tinnitus, hearing loss, vertigo, meningismus, poliosis, and vitiligo, although not all patients present with the complete constellation of these extra-ocular findings. Clinically these patients present with granulomatous anterior uveitis, vitritis, disc edema and serous retinal detachments.

The revised diagnostic criterion was given in 1999 by AAU according to which the acute presentation of VKH were described as complete, incomplete, and probable, depending on how many of the diagnostic criteria they fulfilled. However, these are based more on clinical findings and more useful for clinical trials and do not incorporate the various imaging modalities available which are now an indispensable part of the armamentarium of an ophthalmologist in order to establish a diagnosis as well as to monitor therapy. Fundus fluorescein angiography, SD-optical coherence tomography and indo-cyanine angiography have been well described and studied extensively. Recently, enhanced depth imaging has emerged as a potential and promising modality in order to monitor the choroidal thickness and hence aids in monitoring and follow-up of the patients. Also, Angio-OCT is a newer and promising modality that employs motion contrast imaging to high-resolution volumetric blood flow information generating angiographic images in a matter of seconds for the same. Extensive studies are in vogue to research and co-relate the various modalities in order to make the diagnosis simpler, to make the disease more lucid, and to consistently monitor therapy as non-invasively as possible.

Case Report

A 40-year-old female patient presented to us with diminution of vision in both eyes since 15 days associated with headache and meningismus. There was no history of trauma or any other systemic illness. Best corrected visual acuity in right eye was 6/9 and left eye was 6/18 with a normal IOP. Anterior segment has cells of 2+ and other findings were unremarkable.

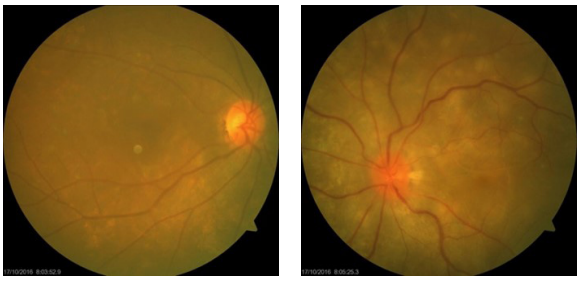

Figure 1 and Figure 2: *Posterior segment (Figures 1 and 2) showed vitritis, disc edema and deep yellowish lesions with pockets of sub-retinal fluids. B-scan showed an increased RCS of 1.83 and 2.63 mm in RE and LE respectively.*

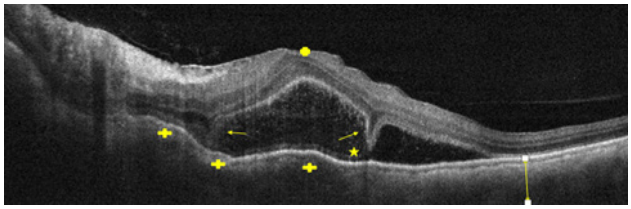

Figure 3: *Raster scan HD-OCT, showing an altered foveal contour (yellow cross) with hypo-reflectivity in the sub-retinal space suggestive of an exudative retinal detachment along with intramembranous structures (yellow star) which are hypothesized to be remnants of the degraded cone outer segments along with fibrinous septae (yellow arrows).*

The RPE has uneven bumpy appearance (yellow plus) due to the underlying choroidal granulomas showing RPE undulations. The choroidal thickness is increased (two headed yellow arrow) owing to the inflammation and is an important and useful parameter in order to confirm the diagnosis and monitor treatment and to detect exacerbations or recurrences in patients on follow-up.

These can be correlated well with the ICGA pictures which clearly demonstate the granulomas. Fuzzy and leaky choridal vessels are visualised the early phase along with areas of hypoflurosence which remain hypo in late phases in cases of full thickness granulomas or become iso or hyperfluroscent in partial thickness granulomas.

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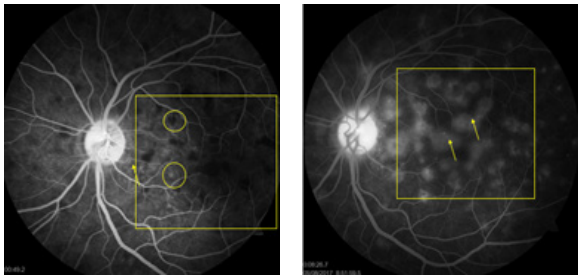

Figure 4 and Figure 5: *The classical lesions seen on fundus fluorescein angiography (Figure 4) are the well defined areas of hyper and hypoflurensence (yellow circles)in the early phase suggestive of the underlying choroidal granulomas along with the pin point areas of hyperflurensce(yellow arrow) which leak in the late phase followed by pooling of the dye (yellow arrows in Figure 5) in areas of exudative retinal detachment.*

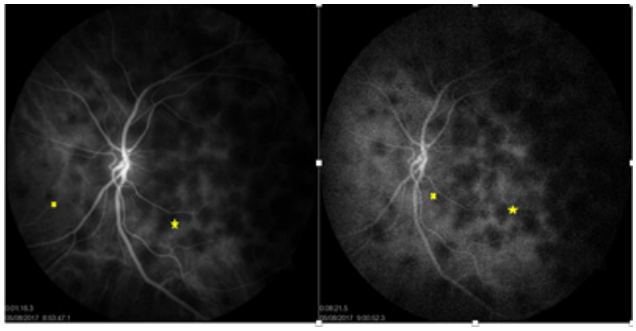

Figure 6 and Figure 7: *Observe the yellow star in Figure 6(early phase) which remians hypo while the pentagon which becomes hyperfloursent in Figure 7 (late phase).*

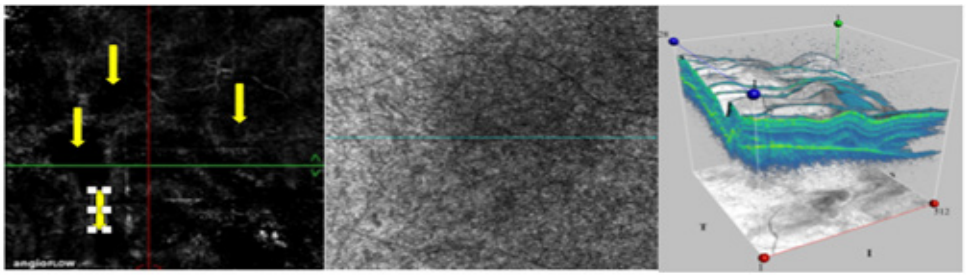

Figure 8, Figure 9, Figure 10: *OCT angiography en face image (scanned area represented by yellow square on FFA) at the level of the choriocapillaris shows multiple hypo-reflective round-to-oval lesions, which represent areas of flow void or choriocapillaris hypo-perfusion corresponding to the hypofluorescent lesions seen on FA and ICGA (yellow arrows in Figure 8). Figure 9 is an En-Face image demonstrating capillary hypo-perfusion with Figure 10 showing a 3-D demonstration of the increased choroidal thickness and RPE undulations obtained on an Enhanced depth OCT.*

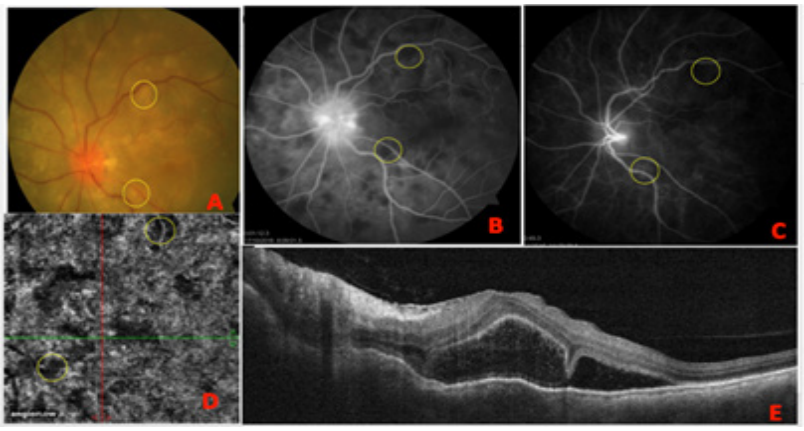

Figure 11: *Demonstrates multimodal imaging showing choroidal granulomas and associated changes in the choriocapillaries. Part A is a fundus picture showing multiple deep yellowish lesions which on Fundus fluorescein angiography (B) show hypo- fluorescent lesions which on Indocyanine green angiography (ICGA) in the early shows hypofluorescent lesions that continue to remain hypofluorescent in the late phase suggestive of choriocapillaris ischemia (C). The study lesions (yellow circles) on ICGA can be seen as areas of flow void on OCTA en face image at the level of the choriocapillaris (D). Enhanced depth imaging optical coherence tomography scan (E) passing through the lower choroidal lesion shows localized thickening of the choriocapillaris with loss of typical pattern along with RPE undulations suggestive of choroidal granulomas.*

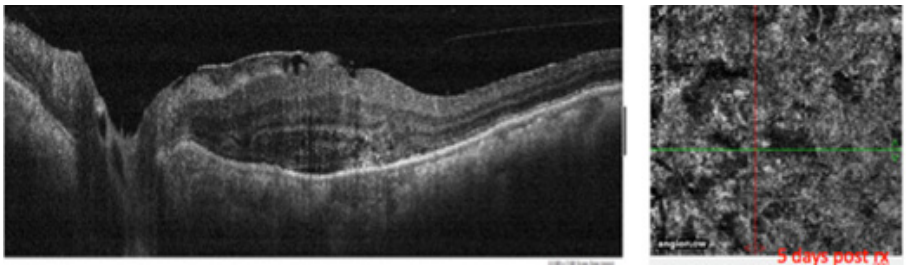

Figure 12: *The patient was started on Intravenous steriods for 5 days following which her subretinal fluid started resolving, and choroidal thickness reduced along with reduction in the flow void areas on the angio-scan and discharged on oral steroids and oral immunosupressnats (Figure 12).*

The patient was followed up on using Angio OCT-scans and Enhanced depth imaging scans to monitor the disease activity via periodically measuring the central macular thickness, choroidal thickness and flow-void areas.Picture 13 shows the serial OCT scans of the LE of the patient uptil 6 months till when the patient was on oral Immunosupressant tab Azathioprine 50mg BD and the oral steriods tapered by 3months. The patient is followed up using the enhanced depth OCT scan which show the resolution of the sub-retinal fluid and the fibrin along with decrease in the choroidal thickness. These are well co-related with the Angio-OCT scans at the level of choriocapilaris where the areas of flow void decrease as the gramulomas and the inflammation resolved.The patient is stable 6 months with a BCVA of 6/6 in RE and 6/12 in LE and is on oral immunosuprresant and is on regular follow-up.

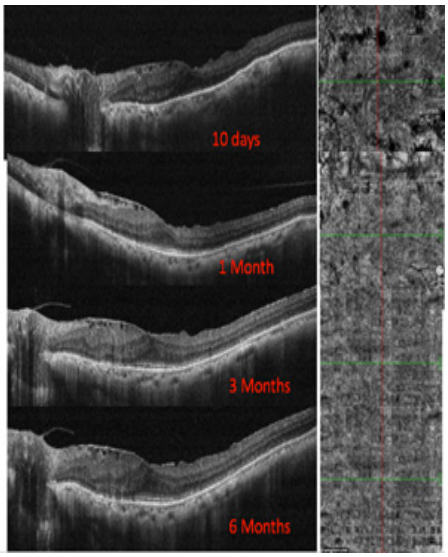

Figure 13: *In conclusion, multimodal imaging is not the future, it's the present standard of care for these eyes not only in the diagnosis, but also for non-invasive follow-up and early recognition of recurrences. Early diagnosis, early intravenous and oral steroids and long term immunosuppression along with multimodal imaging with pertinent follow-up of these patients is an indispensable part for the proper management and desired visual rehabilitation of these patients.*

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