Optical Coherence Tomography Angiography in the Diagnosis and Follow-up of Vogt-Koyanagi-Harada Syndrome

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ABSTRACT

An 18-year-old female patient applied to our clinic owing to sudden visual loss which started 3 days ago on the right eye. There were +3 cells in the anterior chamber, +3 cells in the vitreous and multiple serous retinal detachment in the right eye. The anterior segment and fundus were normal in the left eye. Optic disc leakage in both eyes and hyperfluorescent areas due to subretinal fluid pooling in the phase in the right eye were detected in fundus fluoresceine angiography. Hypofluorescent foci were detected in the mid-late phase in both eyes in indocyanine green angiography. A marked thick choroid (690 µm) in optical coherence tomography and decreased choriocapillaris flow rate in right eye compared to left eye in optical coherence tomografi angiography were detected. Pleocytosis was detected in the lumbar puncture. Based on these findings, the patient was diagnosed as bilateral asymmetric onset acute Vogt-Koyanagi-Harada and treated with 1 g intravenous pulse steroid treatment. On the third day of the pulse steroid treatment, the amount of subretinal fluid significantly decreased and choriocapillaris flow rate significantly increased in the right eye and slightly increased in the left eye. On the 7th day, 1 mg / kg / day oral steroid and 100 mg azothiopurine treatment were started after complete resolution of subretinal fluid and increase of visual acuity up to 0.5.

Key Word: Vogt-Koyanagi-Harada syndrome, Optical coherence tomography angiography.

INTRODUCTION

Vogt-Koyanagi-Harada (VKH is a disorders resulting from autoimmunity development against melanocyte in choroid, meninx, inner ear and skin and diagnosis is made with clinical findings.[1] Ocular findings include diffuse

choroiditis at acute phase and secondary exudative retinal detachment, disc hyperemia, papillary edema and vitritis (ref 8). Findings are bilateral with simultaneous onset in 70% of cases; however, asymmetric involvement can also be observed rarely. As disease is a diffuse choroiditis, indocyanine green angiography (ICGA) is gold standard in the diagnosis[2] In the mid-phase of ICGA (on minutes 8-12), multi-focal hypo-florescent spots are diagnostics.[2] However, it is reproducibility is limited as it is an invasive procedure.

The optical coherence tomography angiography (OCTA) is a non-invasive imaging modality that enables detailed evaluation of retinal and choroidal vascularization without use of contrast material.[3] In OCTA, the motion contrast of intravascular erythrocytes are processed using consecutive optical coherence tomography B-scans of a certain retinal area and detailed images of vascular network is obtained. Since no intravenous contrast material is used in OCTA, no adverse reaction occurs. Another advantage is that it can be repeated numerously in the same day.

Here we aimed to emphasize imaging modalities used in the diagnosis and follow-up of an active VKH patient with asymmetrical onset and changes in OCTA findings during treatment.

CASE REPORT

A 18-years old woman presented to our clinic with acute onset of vision loss occurred 3 days ago. In the history, there was no previous trauma and detailed anamnesis revealed that patient had headache over a week. In the examination, visual acuity was at level of finger counting

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at 5 meters in right eye and full vision was detected in left eye. There was +3 cell in the anterior chamber, +3 cells in the vitreous and serous retinal detachment in the right eye. Anterior segment and fundus examination was normal in the left eye. On optic coherence tomography (OCT), there was septal serous retinal detachment in the right eye and choroidal thickness was measured as 690 µm. (Picture 1A-B). On fundus fluorescein angiography (FFA), leakage in optic disc in both eyes, hypo-florescent areas in the right eye and enlargement in hyper-florescent areas due to accumulation of subretinal fluid at late phase were observed (Picture 2A-B). IN ICGA, hypo-florescent foci were observed in mid-late phase in both eyes (Picture 3A-B). It was observed that these foci became isoflorescent on late phase after minute 20.

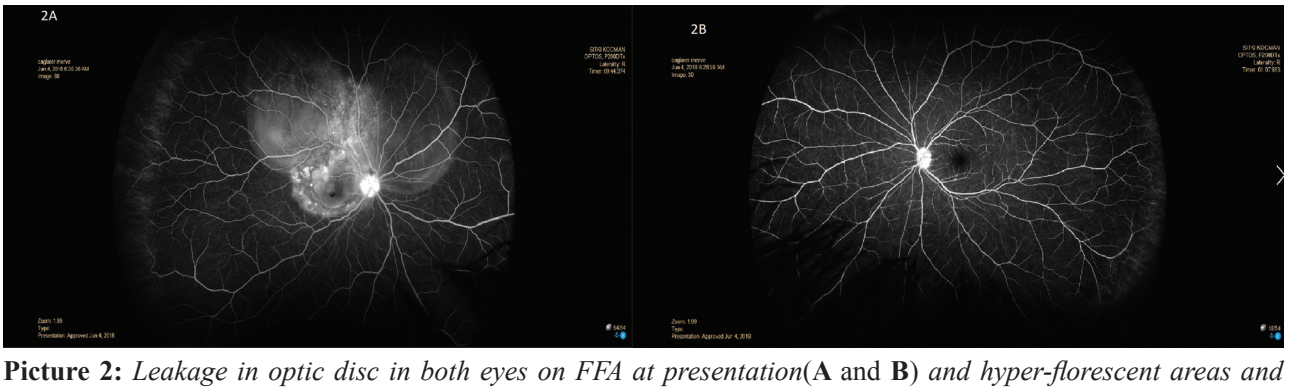
On OCTA, choriocapillaris flow rate was found to be markedly lower in right eye when compared to left eye KTA (Picture 4A-B). The patient was consulted to ETN, neurology and dermatology departments. On audiogram, no abnormal finding was detected. Lymphocytic pleocytosis was detected in cytological evaluation of lumber puncture performed by neurology department. Based on these findings, the patient

was diagnosed as acute VKH with asymmetrical onset and intravenous pulse steroid therapy (1 g) was initiated. The patient was monitored by daily OCT and OCTA scans. On day 3 of pulse steroid therapy, it was detected that subretinal fluid was markedly decreased in right eye and that there a marked increase choriocapillaris flow rate in right eye and slight increase in the left eye was detected on OCTA (Picture 5A-B).

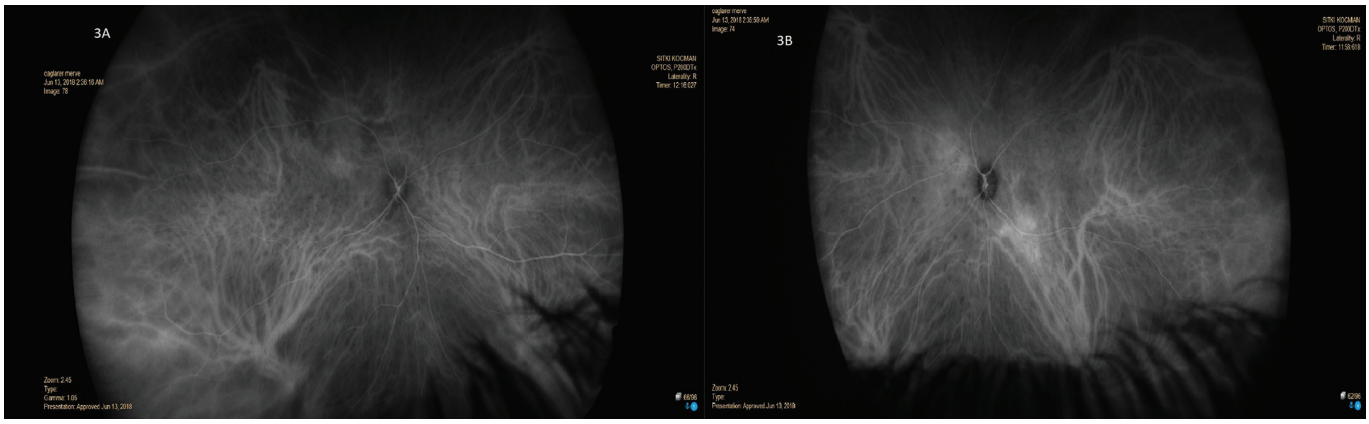
Thus, parenteral steroid dose was gradually reduced and it was discontinued as subretinal fluid was completely resolved on day 7. The patient was given oral steroid (1 mg/kg/day) and azathioprine (100 mg).

On week 1, visual acuity was improved to 0.5 in right eye. There was increase in choriocapillaris flow rate in right eye

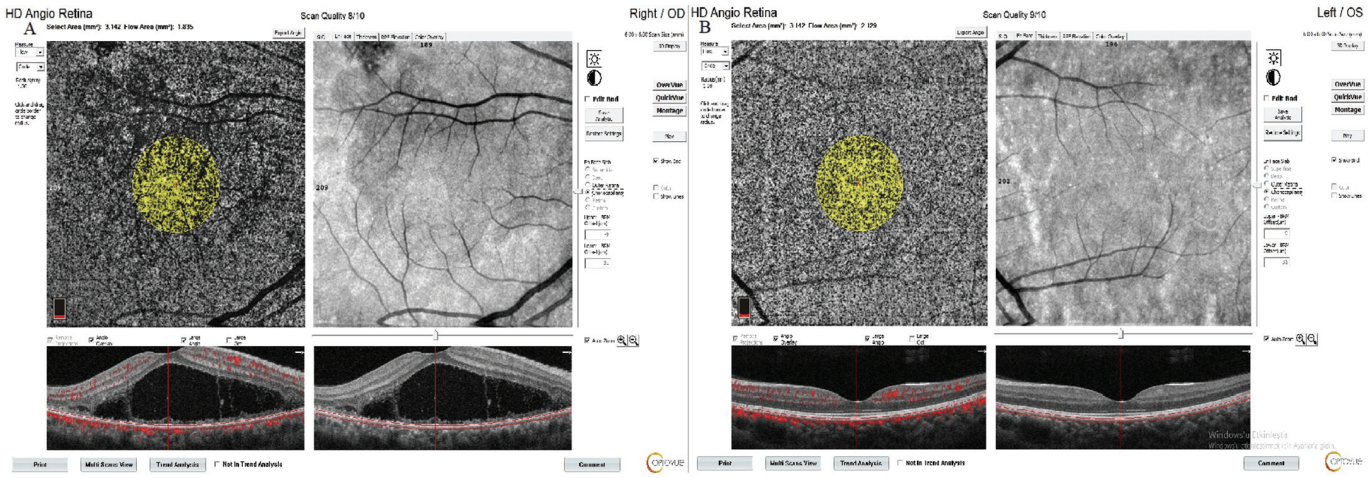
Picture 1. A) *Above, septal retinal detachment and markedly thick choroid (690 micron) in right eye. Below, very thin subretinal fluid and choroid thinning after treatment.* **(B)** *It was seen that choroid was thick (473 micron despite normal retina in left eye. Below, choroid thinning after treatment*



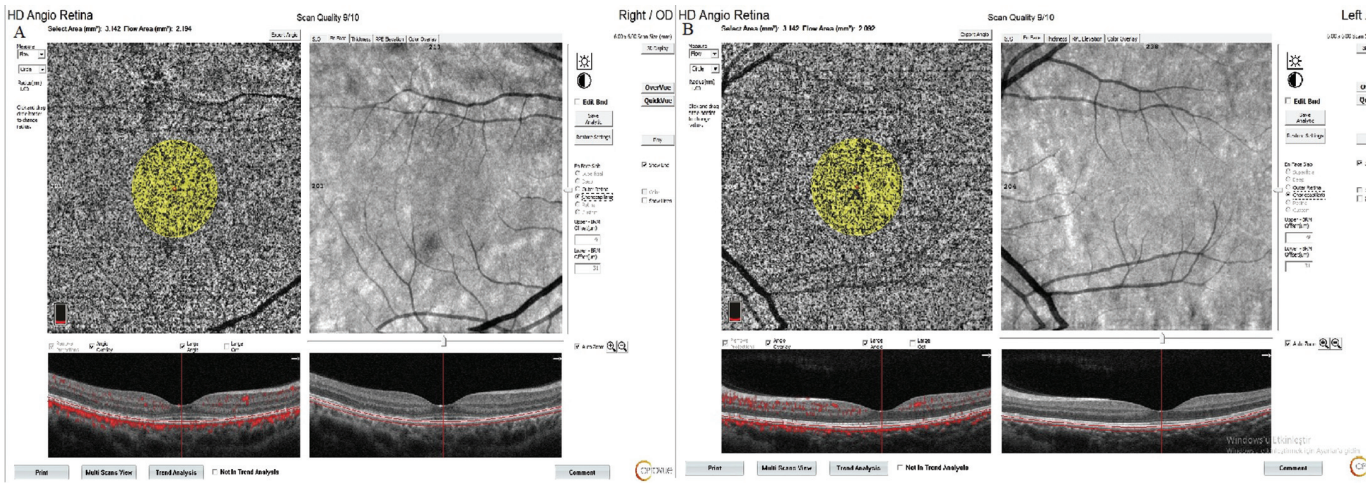
Picture 2: *Leakage in optic disc in both eyes on FFA at presentation*(**A** and **B**) *and hyper-florescent areas and enlargement in hyper-florescent areas due to subretinal fluid accumulation at late phase in right eye* (**A**).



Picture 3: *Hypo-florescent foci at mid-late phase in both eyes on indocyanine green angiography* (**A** *and* **B**).



Picture 4. *Baseline choriocapillaris flow rate in right and left eye on optic coherence tomography.*



Picture 5. *Post-treatment choriocapillaris flow rate in right and left eye on optic coherence tomography.*

on OCTA. On the control visit at month 1, visual acuity was 0.8 in right and 1.0 in the left eye. No subretinal fluid was detected in either eye on OCT. an increase was recorded in choriocapillaris flow rate in right eye on OCTA.

DISCUSSION

The VKH is an autoimmune disorders involving many systems such as eye, skin, meninx and auditory system.[1] Particularly, in patients with genetic predisposition, choroiditis is observed due to involvement of stromal choroid. Punctuate hypo-florescent areas on ICGA are compatible with granulomatous infiltration observed in histopathological evaluation.[2] Although ICGA has an important role in the assessment of treatment, it has some disadvantages such as invasiveness and cost.

In this case report, although choroidal thickness was increased markedly on EDI-OCT, considerable reduction was observed in choriocapillaris flow rate when compared to contralateral eye. Choroidal vascular layer is formed by choriocapilalris/Sattler and Haller layer beneath retinal pigment epithelium. In central serous chorioretinopathy (CSCR), there was enlargement in Haller layer while focal hypo-florescent in choriocapillaris/Sattler later on ICGA.[4] The amount of non-vascular smooth muscle cells (NVSMC) is substantial in Haller layer.[5] In the pathogenesis of CSCR, it was reported that relaxation of NVSMCs via sympathetic innervation or K^+ activation can cause increased vascular filling and interstitial edema[6,7] The increased overall choroidal thickness on EDI-OCT was attributed to increased thickness of Haller layer.[8] In VKH, the increased thickness of Haller layer caused by inflammation may be cause of increased overall choroidal thickness due to higher vascular filling and interstitial edema on EDI-OCT. however, focal hypo-perfusion may be attributed to less NVSMC in choriocapillaris/Sattler layer[9] and failure to enlarge due to compression caused by Haller layer. In fact, Aggarwal et al. found focal hypo-perfusion in choriocapillaris on OCTA which was correlated to ICGA in 10 patients with acute VKH. Authors emphasized that change in choriocapillaris flow rate as rated by OCTA can be a helpful method in monitoring treatment response.[10] In a patient with acute VKH, Wintergerst et al. showed decrease in flow rate in both choriocapillaris and Sattler layer by OCTA and that there was an increase in flow rate on OCTA afteer 4-weeks treatment.[11] In our patient, it was seen that there was a marked increase in the choroidal thickness in involved eye on baseline EDI-OCT and that, when assessed by OCTA, choriocapillaris flow rate was lower in involved eye compared to contralateral eye. After treatment, it was found that choriocapillaris flow rate was increased in both eyes. Due to increased flow rate, exudation in involved eye was disappeared on OCT and visual acuity was improved. In addition, the increased choriocapillaris in clinically intact eye suggest that involvement may be bilateral but asymmetrical in VKH which is generally suggested as bilateral and rarely unilateral.

In conclusion, it was seen that acute VKH showed good response to high dose corticosteroid treatment at early period. Oral steroid and immunosuppressive therapy is required to control disease activity. Although clinical findings are observed in one eye, involvement in contralateral eye without visual involvement can be detected by ICGA and OCTA. In patients with VKH, the success in early diagnosis and treatment is highly effective in preventing chronic changes that severely worsen vision. Thus, we think that assessment of flow rate alterations in choriocapillaris by OCTA may be helpful in both diagnosis and monitoring treatment response.

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