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Optical Coherence Tomography Findings in a Patient with Adult-Onset Foveomacular Vitelliform Dystrophy

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Abstract | **Case Report**

We present a case of adult-onset foveomacular vitelliform dystrophy (AFMVD) in a 73-year-old man. Ophthalmoscopy revealed discoloration in both eyes, but typical subretinal depositions of yellowish material were not observed. OCT revealed epiretinal membrane in the right eye. Hyper-reflective subretinal deposits at the level of the retinal pigment epithelium in the left eye. Moreover, hyper-reflective deposits were also detected at the sub-ellipsoid zone and intraretinal layers. Based on these findings, we diagnosed our patient with AFMVD in vitelliruptive phase. OCT was useful in visualizing the location of the vitelliform material in a patient with AFMVD.

Keywords: Adult-onset foveomacular vitelliform dystrophy, optical coherence tomography.

INTRODUCTION

Adult-onset foveomacular vitelliform dystrophy (AFMVD) is a relatively uncommon macular disease that shares phenotypic features with Best vitelliform macular dystrophy [1-5]. AFMVD is generally diagnosed based on the observation of subretinal deposition of yellowish material within the macula [1-5]. According to optical coherence tomography (OCT), some researchers suggested a four-stage classification for AFVD, including vitelliform phase, pseudohypopyon phase, vitelliruptive phase and atrophic phase, in progressive order [1-5]. However, few reports have focused on the exact location of the vitelliform material of AFMVD using OCT [6]. We describe OCT findings in a patient with AFMVD.

CASE REPORT

A 73-year-old Japanese man presented at Inoda Eye Clinic for bilateral fundus discoloration. He had no significant medical history. His best corrected visual acuity (BCVA) was 1.2 in the right eye and 1.0 in the left eye. Ophthalmoscopy revealed discoloration in both eyes, but typical subretinal deposition of yellowish material were not observed (Figure 1A, B).

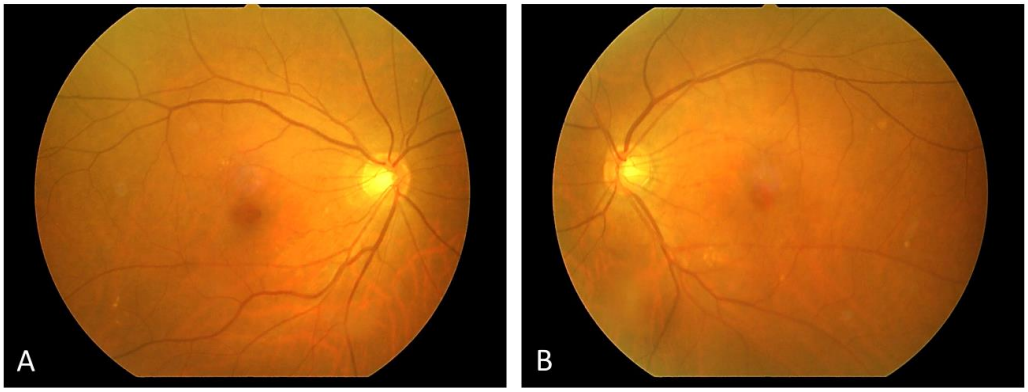

Fig 1: Fundus photographs of the (A) right and (B) left eyes
Note discoloration within the macula.

OCT revealed epiretinal membrane in the right eye (Figure 2A, B). Hyper-reflective subretinal deposits at the level of the retinal pigment epithelium (RPE) in the left eye (Figure 3A, B). Moreover, hyper-reflective deposits were also detected at the sub-ellipsoid zone (EZ) (framed arrowhead) and intraretinal layers (arrowheads) (Figure 3). The EZ was not disrupted in the left eye.

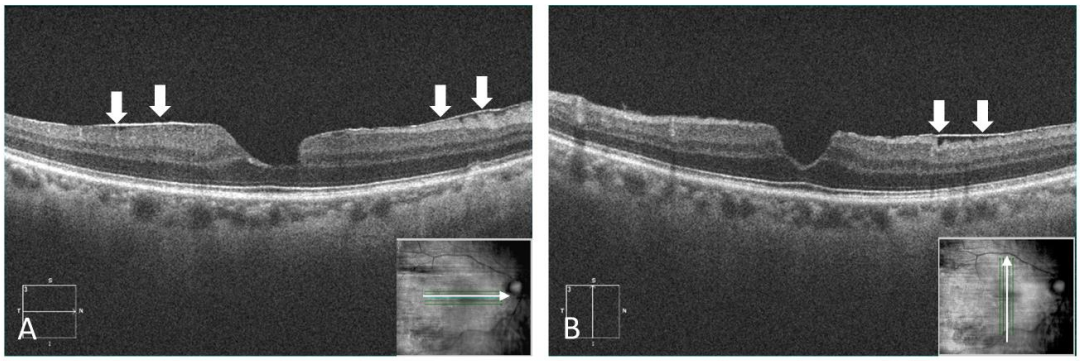

Fig 2: Optical coherence tomography images of the right eye in the horizontal (A) and vertical (B) direction
Note epiretinal membrane within the macula (arrows).

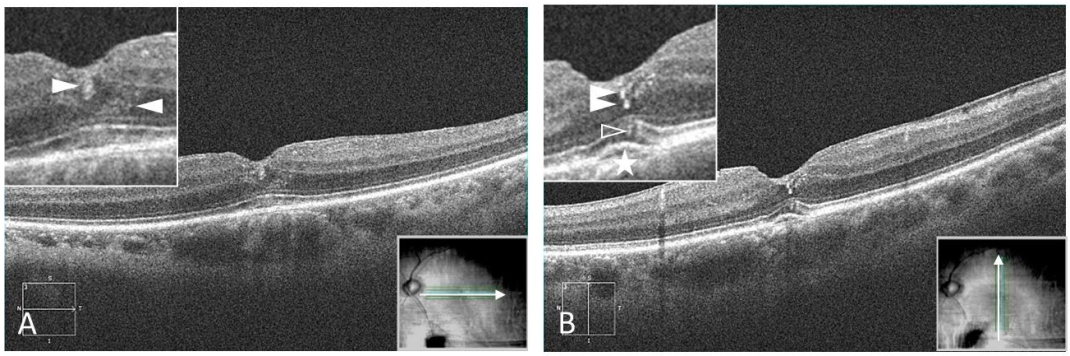

Fig 3: Optical coherence tomography images of the left eye in the horizontal (A) and vertical (B) direction
Hyper-reflective subretinal (star) and intraretinal (arrowheads) deposits within the macula.
Note hyper-reflective deposits at the sub-EZ (framed arrowhead) and intraretinal (arrowheads) layers.

Based on these findings, we diagnosed our patient with AFMVD in vitelliruptive phase. The patient was followed without treatment.

DISCUSSION

Querques *et al*. [2] described AFMVD and suggested that it should be considered as a dynamic process involving alternating phases of material accumulation and reabsorption as it progresses. Querques *et al.*, [3] also described the correlation between BCVA, EZ integrity, and stage of the disease. According to their report, BCVA loss has a strong, statistically significant correlation with the presence of focal disruption or diffuse loss of the EZ, as well as with a more advanced stage of the disease. Because the EZ was not discontinuous, the present case presented relatively good vision.

The exact location of the vitelliform material in AFMVD has not been fully elucidated. Puche *et al.*, [6] described the different morphological features in AFMVD using high-resolution spectral domain OCT. According to their report, hyper-reflective clumps within the outer plexiform and outer nuclear layers in 28/60 eyes. In 9/60 eyes, the EZ appeared highly reflective like a shell all around the vitelliform material, and appeared irregular and discontinued in 27/60 eyes. The vitelliform material appeared as a highly reflective dome-shaped lesion located between the photoreceptor layer and the RPE. They hypothesized that early changes involve the layer between the RPE and the EZ, first with vitelliform material accumulation beneath the sensory retina, and then with the EZ alterations, "pigments migration" towards inner layers and fluid accumulation. In our present case, hyper-reflective deposits were detected at the subretinal, sub- EZ and intraretinal layers. Therefore, we consider these deposits have probably migrated from the outer retinal layers.

CONCLUSION

Although our findings were based on a single case, OCT was useful in visualizing the location of the vitelliform material in a patient with AFMVD.

Disclosure: The authors have no conflicts of interest to disclose.

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