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

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Abstract: We present a case of adult-onset foveomacular vitelliform dystrophy (AFMVD) in a 62-year-old man. Fundus examination revealed subretinal depositions of yellowish material within themacula. During the 4-year follow-up period, the best corrected visual acuity (BCVA) in the right eye did not change; the BCVA in the left eye gradually deteriorated from 0.5 to 0.08. Ophthalmoscopic findings showed no change in the subretinal yellowish deposit in the right eye. Atrophic changes gradually developed in the left eye; vitelliruptive degeneration was observed 3.5 years after the initial visit. Optical coherence tomography (OCT) revealed progressive accumulation of hyper-reflective material in the subretinal space and vitelliruptive degeneration was observed in the left eye. The left central macular thickness decreased between the initial and last visits. The subretinal hyper-reflective materials were reabsorbed in both eyes during the natural course of the disease. Thus, OCT was useful in visualizing the subretinal lesions in a patient with AFMVD.
Keywords: Adult-onset foveomacular vitelliform dystrophy, Optical coherence tomography, Natural course.

INTRODUCTION

Adult-onset foveomacular vitelliform dystrophy (AFMVD) is a relatively uncommon macular disease that shares phenotypic features with Best vitelliform macular dystrophy [1]. The clinical onset is typically between the fourth and sixth decade of life [1]. AFMVD is generally diagnosed based on the observation of subretinal deposition of yellowish material within the macula during fundus examination [1]. Several recent reports have described the use of optical coherence tomography (OCT) to examine cases of AFMVD [1-7]. However, few reports have focused on the natural course of patients with AFMVD using OCT [1, 2].We describe OCT changes in a patient with AFMVD.

CASE REPORT

A 62-year-old Japanese man was referred to our 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 0.5 in the left eye. Anterior segments and ocular pressures were normal in both the eyes. Ophthalmoscopy revealed bilateral subretinal deposition of yellowish material within themacula (Fig. 1).

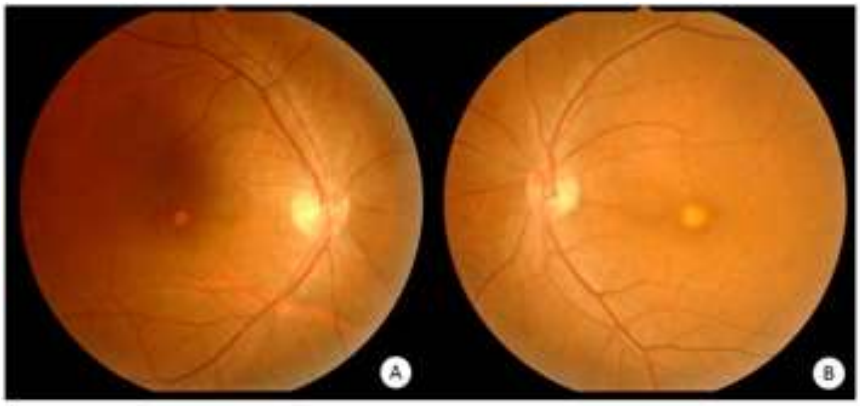

Fig.1: Fundus photographs of the (A) right and (B) left eyes at the initial visit.

Yellowish deposits were observed within the macula.

Fundus autofluorescence imaging showed clearly defined bilateral hyperfluorescent lesions corresponding to subretinal deposits. OCT revealed hyper-reflective subretinal deposits at the level of the retinal pigment epithelium (RPE)/Bruch membrane within the lesion area (Fig. 2). The inner segment/outer segment (IS/OS) interface was disrupted in the left eye.

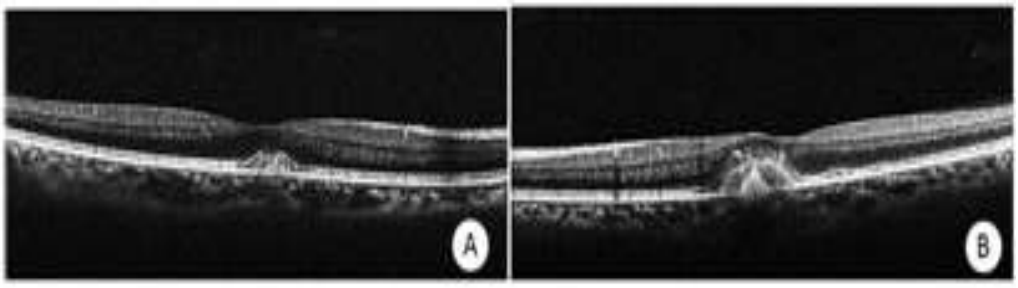

Fig. 2: Optical coherence tomography (OCT) images of the (A) right and (B) left eyes in the horizontal direction at the initial visit.

Hyper-reflective subretinal deposits were observed within the macula. Based on these typical findings, we diagnosed our patient with AFMVD. The patient was followed without treatment.

During the 4-year follow-up period, the BCVA score in the right eye did not change; the BCVA score in the left eye gradually deteriorated from 0.5 to 0.08. The subretinal yellowish deposit did not change in the right eye; gradual atrophic changes developed in the left eye (Fig. 3 A–J). Vitelliruptive degeneration was observed in the left eye 3.5 years after the initial visit (Fig. 3H).

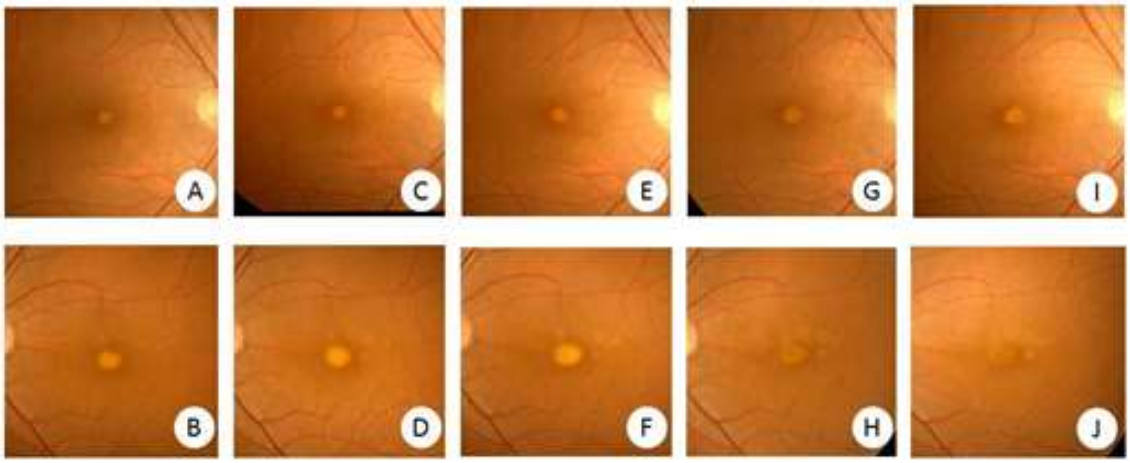

Fig. 3: Fundus photographs of the right (top) and left (bottom) fundus over a 4-year period. The subretinal yellowish deposit did not change in the right eye; atrophic changes gradually developed in the left eye. A, B: 6 months after initial visit; C, D: 1yearafter initial visit; E, F: 2years after initial visit; G, H: 3.5 years after initial visit; I, J: 4years after initial visit.

In contrast, OCT revealed progressive accumulation of hyper-reflective material in the subretinal space (Fig. 4 A-H) and vitelliruptive degeneration was observed in the left eye (Fig. 4 J, L).The central macular thickness decreased in the left eye between the initial and last visits. The subretinal hyper-reflective materials were reabsorbed in both eyes during the natural course of the disease.

High-magnification OCT images clearly demonstrated disruption of the IS/OS interface in the left eye during the last visit (Fig. 5).

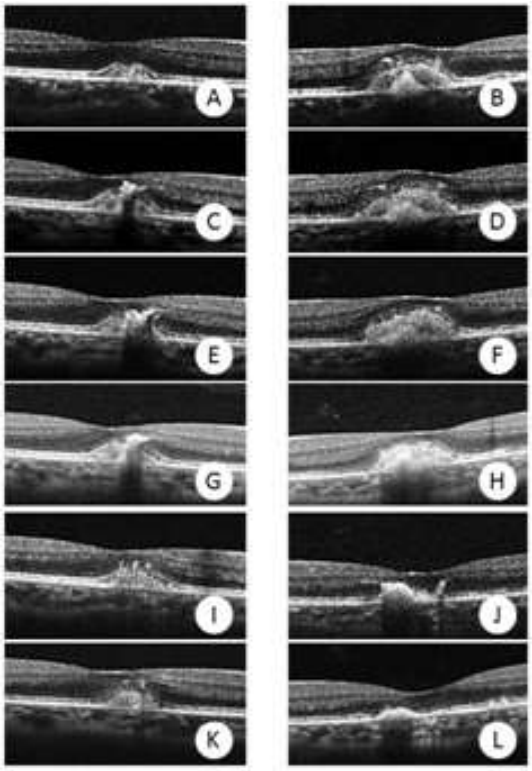

Fig. 4: Optical coherence tomography (OCT) images of the right (left) and left (right) fundi in the horizontal direction over a 4-year period. Progressive accumulation of hyper-reflective material in the subretinal space (A-H) and vitelliruptive degeneration in the left eye (J, L) were observed. A, B: initial visit; C, D: 2 years after initial visit; E, F: 2.5years after initial visit; G, H: 3years after initial visit; I, J: 3.5years after initial visit; K, L: 4years after initial visit.

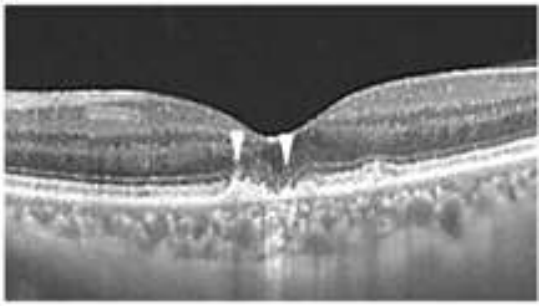

Fig. 5: High-magnification optical coherence tomography (OCT) images of the left eye in the horizontal direction at the last visit. OCT shows disruption of the inner segment/outer segment (IS/OS) interface (arrowheads).

DISCUSSION

Previously, the BCVA score was observed to changewith the age of the patient and the stage of Best vitelliform macular dystrophy [8]. Recently, several reports have described cases of AFMVD examined using OCT [1-7]. 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, IS/OS 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 IS/OS interface, as well as with a more advanced stage of the disease. In our patient also, a progression in the central photoreceptor IS/OS interface status and changes in lesion reflectivity were accompanied by a significant BCVA reduction between the initial and last visits.

CONCLUSION

Although our findings were based on a single case, retinal changes were more easily defined on OCT images than on color photographs.OCT was useful in visualizing the unusual subretinal lesions in a patient with AFMVD. Our findings may contribute to a better understanding of the natural course of this disease.

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