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Ellipsoid zone changes in a patient with adult-onset foveomacularvitelliform dystrophy

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Abstract: We present a case of adult-onset foveomacular vitelliform dystrophy (AFMVD) in a 73-year-old man. Fundus examination revealed subretinal depositions of yellowish material within themacula in both eyes. During the 6-month follow-up period, the best corrected visual acuity (BCVA) in the right eye gradually deteriorated from 0.9 to 0.5. On optical coherence tomography (OCT), vitelliruptive degeneration and disruption of the photoreceptor inner segment/outer segment interface (ellipsoid zone) gradually developed in the right eye. We speculate that progression in the ellipsoid zone status and changes in lesion reflectivity were accompanied by a significant BCVA reduction between the initial and last visits.
Keywords: adult-onset foveomacularvitelliform dystrophy, optical coherence tomography, ellipsoid zone, natural course

INTRODUCTION

Adult-onset foveomacularvitelliform dystrophy (AFMVD) is a relatively uncommon macular disease that shares phenotypic features with Best vitelliform macular dystrophy (BVMD) [1]. Several reports have described the use of optical coherence tomography (OCT)toexaminecases of AFMVD and BVMD [1, 2, 3, 4, 5, 6, 7, 8, 9]. However, few reports have focused on the natural course of patients with AFMVD [1,2, 8].Here, we describe OCT changes in a patient with AFMVD.

CASE REPORT

A 73-year-old man was referred to our clinic forblurry vision. Hehad no significant medical history. His best corrected visual acuity (BCVA) was 1.2 in the right eye and 0.5 in the left eye. Slit-lamp examination showed cortical opacities in both lenses. Ophthalmoscopy revealed bilateral subretinal depositions of yellowish material within themacula(Figure 1a and b). FAF imaging showed clearly defined bilateral hyperfluorescent lesions corresponding to subretinal deposits surrounding hypofluorescent dark areas(Figure 1c and d).

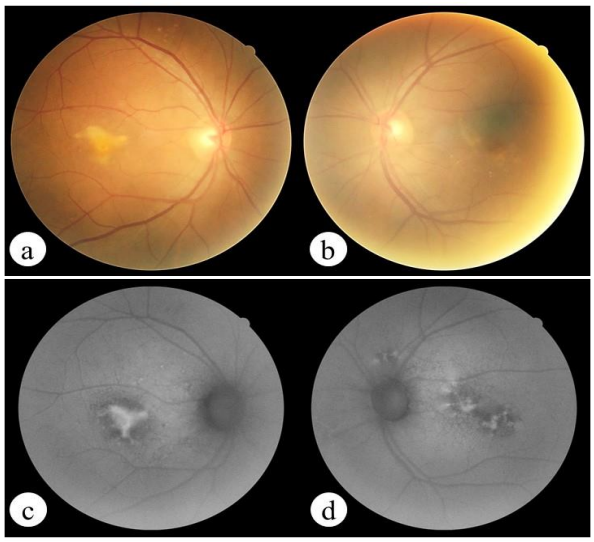

Fig.1:Fundus photographs (a, b)and fundus autofluorescent (FAF) imaging (c, d) of the (a, c) right and (b, d) left eyes at the initial visit

Yellowish deposits were observed within the macula.FAF imaging shows clearly defined hyperfluorescent lesions corresponding to subretinaldeposits surrounding hypofluorescentdark areas.

OCT revealed hyper-reflective subretinal deposits at the level of the RPE/Bruch membrane within the lesion area(Figure 2b).The inner segment/outer segment (IS/OS) interface (ellipsoid zone) was not disrupted in the righteye(Figure 2b arrows). The patient was followed without treatment. During the 6-month follow-up period, the BCVA in the left eye did not change; the BCVA in the right eye gradually deteriorated from 0.9 to 0.5. Vitelliruptive degeneration was observed 6months after the initial visit (Figure2e and f). In addition, the ellipsoid zone was disrupted (Figure2 f, flamed arrows).

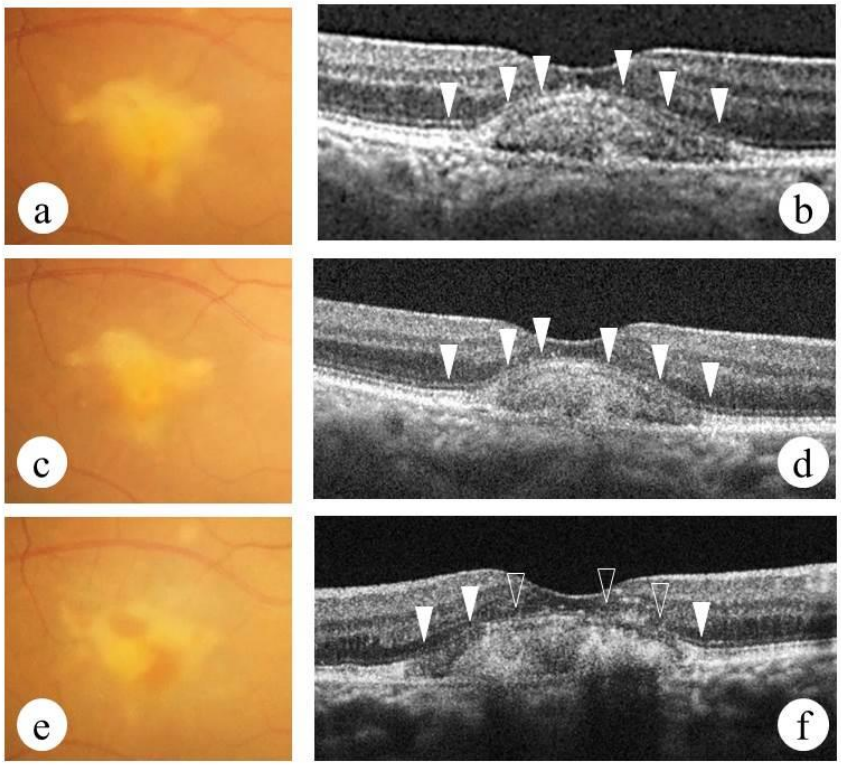

Fig.2: Fundus photographs (a, c and e)and optical coherence tomography (OCT) images(b, d and f) of the right eyeover a6-month period

Progressive accumulation of hyper-reflective material in the subretinal space (b, d and f) and vitelliruptive degeneration (f)were observed. The ellipsoid zone was disrupted(f, flamed arrows).a, b: initial visit; c, d: 4monthsafter initial visit; e, f: 6monthsafter initial visit.
Arrows indicate the ellipsoid zone.

DISCUSSION

In BVMD, five stages have been described, based on fundus examination findings: the previtelliform stage (normalmacula or subtle alteration of the retinal pigment epithelium (RPE)), the vitelliformstage (a well-circumscribed lesion resembling an egg yolk), the pseudohypopyon stage(yellow material accumulated inferiorly), the vitelliruptivestage (partial resorption of the material, scrambled-egg lesion), and the atrophic/fibroticstage (final macular atrophy or fibrosis) [1-4].Previously, the BCVA was observed to change with the age of the patient and the stage of BVMD [9]. Recently, several reports have described cases of AFMVD and BVMD examined using OCT[1, 2, 3, 4, 5, 6, 7, 8, 9].Querques et al. [2]suggested that it should be considered as a dynamic process involving alternating phases of material accumulation and reabsorption asit progresses in AFMVD. 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 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 this present case, we speculate that progressionin the central ellipsoid zone status and changes reflectivity was accompanied by a BCVA reduction between the initial and last visits.

Although our findings were based on a single case and relative short follow-up period, they may contribute to a better understanding of the natural course of this disease.

DISCLOSURE

The authors have no conflicts of interest to disclose.

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