American Journal of Ophthalmology Case Reports 26 (2022) 101542

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Multi-spectral imaging in adult-onset foveomacular vitelliform dystrophy: Report of two cases

Mingzhen Yuan [a], Feiyan Ma [b], Lulu Chen [c,d], Youxin Chen [c,d,*]

[a] Beijing Tongren Eye Center, Beijing Tongren Hospital, Capital Medical University, Beijing Ophthalmology and Visual Sciences Key Laboratory, Beijing, 100730, China
[b] Department of Ophthalmology, The Second Hospital of Hebei Medical University, Shijiazhuang, Hebei Province, China
[c] Department of Ophthalmology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China
[d] Key Laboratory of Ocular Fundus Diseases, Chinese Academy of Medical Sciences & Peking Union Medical College, China

ARTICLE INFO

Keywords:
Multi-spectral imaging
Adult-onset foveomacular vitelliform dystrophy
Optical coherence tomography

ABSTRACT

Purpose: To describe the characteristic findings of non-invasive multi-spectral imaging (MSI) for adult-onset foveomacular vitelliform dystrophy (AFVD).

Observations: On examination of MSI, the characteristic performances of AFVD include the nodule-like high-reflecting lesions, the line-like low-reflecting lesions in the high-reflecting lesion, and the scattered high-reflecting and low-reflecting lesions around the nodule-like lesion. MSI has an advantage over color fundus photography (CFP) and fundus autofluorescence (FAF) in finding tiny lesions, which corresponded to drusenoid structures on optical coherence tomography (OCT). MSI showed different characteristics at different stages of AFVD, which may be instructive to the pathogenesis and progression of AFVD.

Conclusions and Importance: MSI is a promising diagnostic and follow-up tool that will provide additional information in fundus imaging for AFVD, and the changes on MSI is partially instructive to the pathogenesis and progression of AFVD.

1. Introduction

Adult-onset foveomacular vitelliform dystrophy (AFVD) is recognized as a pleomorphic disease, with great diversification in measurement, appearance, as well as distribution of the vitelliform substance.[1] Different from normal eyes and other macular diseases, AFVD is characterized by subfoveal choroidal thickening, accumulation of vitelliform material above the retinal pigment epithelium (RPE) layer.[2] Studies suggest that the vitelliform substance may consist of photoreceptor outer segment debris, lipofuscin, melanin and melanolipofuscin-loaded macrophages, as well as RPE cells.[3] As a new noninvasive examination, multispectral imaging (MSI) could examine the full retina through to the choroid progressively by using different light wavelengths. Generally, the wavelengths range between 550nm and 850 nm. Examples of absorbing composition involve blood, heamoglobin, lipid and pigments such as melanin.[4] In addition, MSI has many advantages in observing retinal and choroidal lesions, and can provide clinicians with good methods to observe the early manifestations of some ocular diseases.[5] So far, MSI has been used in many ocular diseases, such as Vogt-Koyanagi-Harada diseases,[6] central serous chorioretinopathy,[7] age-related macular degeneration,[8] Stargardt disease,[9] as well as retinal vein occlusion and so on.[10] However, AFVD has not yet been reported. As the accumulation of vitelliform substance in retina is one of the major features in AFVD, as well as MSI has the ability to detect subtle RPE lesions, we speculated that the features of AFVD may be more easily characterized with MSI. Therefore, the purpose of our report is to reveal the new imaging findings of AFVD in MSI, which could provide the clues to pathogenesis of AFVD (see Fig. 4).

2. Case reports

Both cases underwent review of the clinical records, color fundus photography (CFP), fundus autofluorescence (FAF), optical coherence tomography (OCT), and MSI (Fig. 1 & Fig. 2). Interestingly, we found that there were some differences on manifestations in MSI among the 4 eyes of these two cases (Fig. 2). In the spectral images of case No.1, a homogeneous 1/5 papilla disc size nodule-like hyperreflectivity lesion with a clear boundary ranging from 550 nm to 850 nm could be observed on MSI for right eye, with the hyperreflectivity got intensified in longer wavelengths (Fig. 2 A). However, MSI in left eye showed

* Corresponding author. Peking Union Medical College Hospital, Shuaifu Garden No.1, Dongcheng District, Peking, China.
E-mail address: chenyx@pumch.cn (Y. Chen).

https://doi.org/10.1016/j.ajoc.2022.101542
Received 18 November 2021; Received in revised form 12 April 2022; Accepted 12 April 2022
Available online 21 April 2022

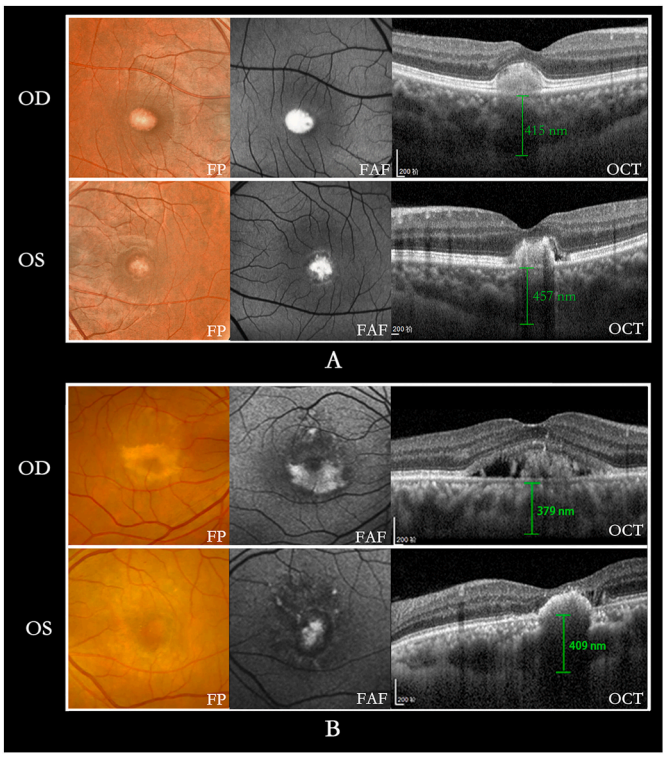

Fig. 1. Ocular examinations for both patients. A: patient No.1; B: patient No.2. FP: fundus photography; FAF: fundus autofluorescence; OCT: optical coherence tomography; OD: right eye; OS left eye; SFCT: subfoveal central thickness.

intertwined linear hyporeflective lesions in the high-reflective lesion from 780 nm to 850 nm wavelength (Fig. 2 B). In case No.2, MSI in right eye showed a nodule-like lesion in macular area, which had low reflectivity in the center part and high reflectivity in the peripheral part from 550 nm to 850 nm wavelength, and the lesion extended and the reflectivity gradually enhanced as the wavelength of MSI became longer. Besides, it appeared some scattered hyper-reflective and hypo-reflective lesions around the nodule-like lesion from 680 nm to 850 nm, and the number of the scattered lesions increased as the wavelength of MSI became longer (Fig. 2 C). As for left eye, MSI showed a nodule-like hyper-reflective lesion in macular area from 780 nm to 850 nm wavelength with some scattered hyper-reflective and hypo-reflective lesions around, and the lesions extended and the reflectivity gradually enhanced as the wavelength of MSI became longer (Fig. 2 D) (see Fig. 3 and Fig. 4).

3. Discussion

AFVD is clinically rare and often misdiagnosed as best vitelliform macular dystrophy and age-related macular degeneration. which was first described by Gass in 1974.[11] Many test methods were used in the diagnosis of AFVD, including FAF, OCT, optical coherence tomography angiography, and electro-oculogram, and electroretinography.[3,12–14] According to OCT, some researchers suggested a four-stage classification for AFVD, including vitelliform phase, pseudohypopyon phase, vitelliruptive phase and atrophic phase, in progressive order.[15]

As for our cases, the nodule-like high-reflecting lesions on MSI that most prominently demonstrated starting from 680nm (mid-level retina and RPE) corresponded with the round cake-like bulge beneath the macular photoreceptor on OCT. And the linear low-reflecting lesions in the high-reflecting lesion from 780 nm to 850 nm wavelength on MSI were quite like RPE disruption on OCT. Moreover, we also found that MSI had an advantage over color fundus photography and FAF in finding

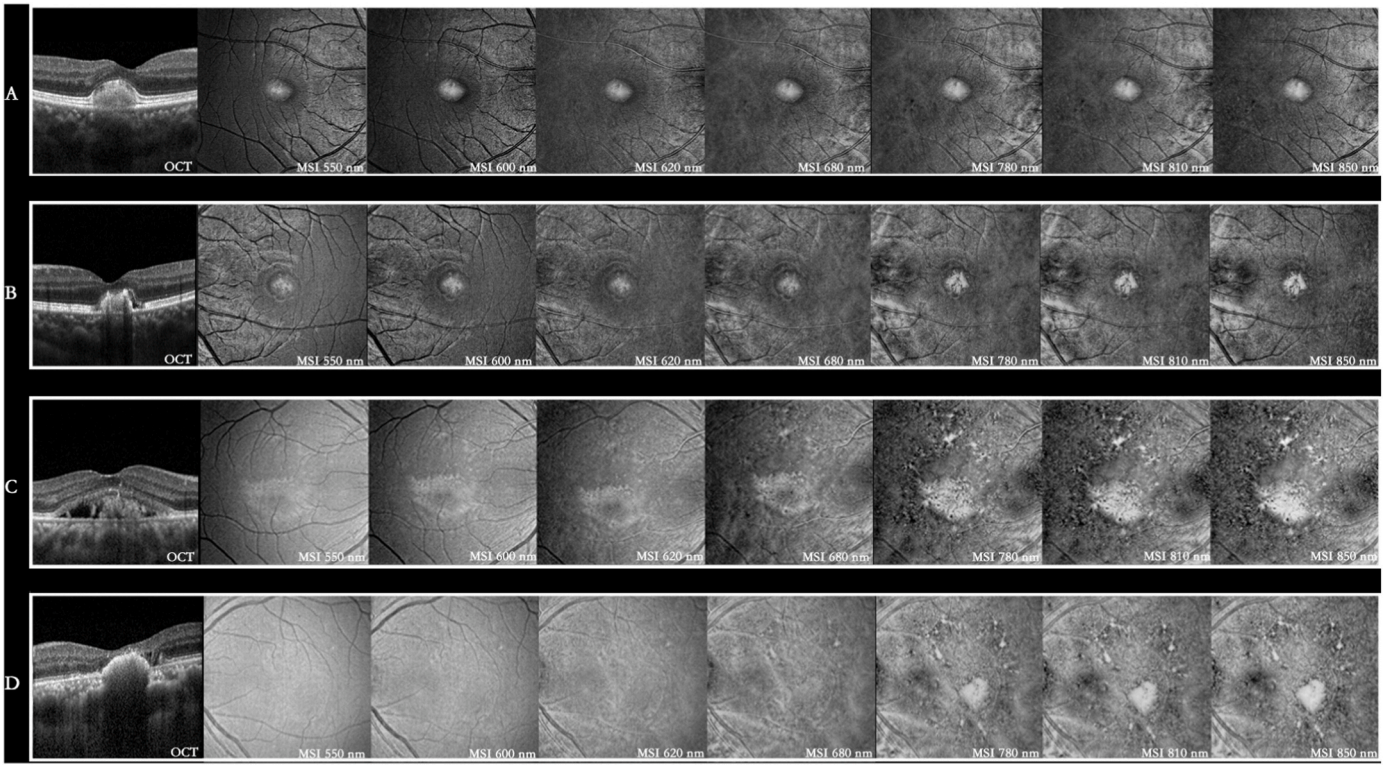

Fig. 2. MSI images of both patients. A: MSI images of right eye in patient No. 1; B: MSI images of lest eye in patient No. 1; C: MSI images of right eye in patient. No. 2; D: MSI images of left eye in patient No. 2. The wavelengths in the figure are 550nm, 600nm, 620nm, 680nm, 780nm, 810nm and 850nm, respectively. OCT: optical coherence tomography; MSI: multispectral imaging.

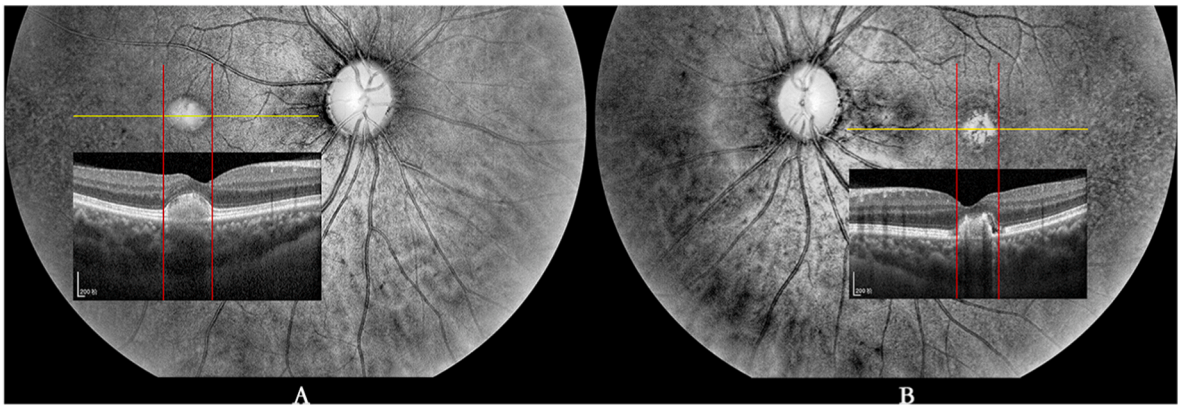

Fig. 3. A The nodule-like high-reflecting lesions on MSI were associated with the round cake-like bulge beneath the inner and outer segments of the macular photoreceptor on OCT. B. The line-like low-reflecting lesions in the high-reflecting lesion on MSI corresponded to RPE disruption on OCT.

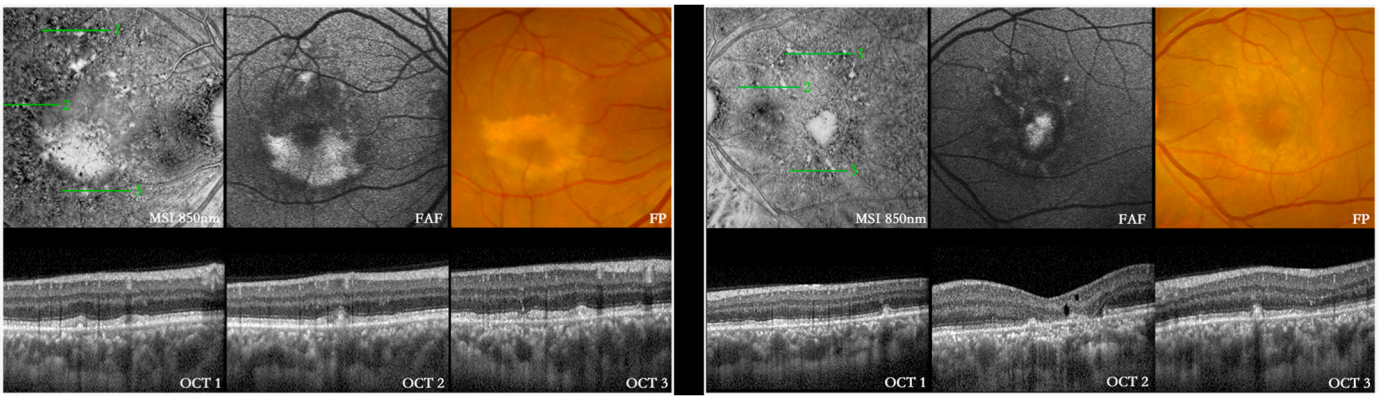

Fig. 4. The lesions on the green line on MSI were not observed on FAF or color fundus photography, but could well-illustrated in MSI long-wavelength slabs which corresponded to drusenoid structures on OCT. MSI: multispectral imaging; FAF: fundus autofluorescence; FP: fundus photography; OCT: optical coherence tomography. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

tiny lesions, which corresponded to drusenoid structures on OCT. We suspected that these structures were the residual components of subretinal deposits. Therefore, MSI, especially in higher wavelength, provides clinicians with the conditions to observe tiny lesions in AFVD through the particular spectral slices of the retina, as well as do regular follow-ups with MSI.

Gass first described the pathological changes in this entity as the photoreceptors lost partially and the pigment migrating to the central macula in AFVD cases. With further research, some experts discovered other characteristics of AFVD, including choroiretinal adhesions and drusens in central macula.[11] Later, some researchers pointed out that the vitelliform materials in the subretinal region were mainly composed of extracellular photoreceptor fragment and RPE-derived substance. In addition, the spectral changes of the vitelliform lesion in our cases revealed some of its pathological components and stages. The right eye of case No.1 was consistent with the initial stage which was characterized by a homogeneous hyperreflectivity lesion from 550 nm to 850 nm on MSI, which was associated with extracellular photoreceptor fragment and RPE-derived substance; while the linear hyporeflective lesions in the hyperreflective lesion in the left eye on MSI were consistent with the presentations in the following stage featured by RPE attenuation, loss of microvilli on top of RPE.[2,3] With the progress of the disease, the lesions were not obvious from 550 nm to 620 nm, but were clearly visible from 780 nm to 850 nm. And the scattered high-reflecting and low-reflecting lesions around the nodule-like lesion in the both eyes of case No.2 were associated with the release of pigment as subretinal deposits. It inferred that early lesions were relatively limited and contained more solid components which is located mostly in the outer retina. These results are consistent with the conclusion put forward by Freund et al. that both dysfunctional RPE and loss of apposition between the photoreceptor tips and the RPE can interfere with the phagocytosis of shed outer segments.[14] However, as the disease progressed, the lesions gradually became liquefied and the range expanded, and eventually formed the residual components of subretinal deposits around the original lesions.

4. Conclusion

All in all, MSI is a promising diagnostic and regular follow-up method that will provide additional information for AFVD, and MSI in higher wavelength might show more detailed information of RPE and outer retinal lesions in AFVD, which may be instructive to the pathogenesis and progression of AFVD. We need a larger case series study to reveal more potential value of MSI in AFVD.

Patient consent

Written consent from the patient to publish this report was obtained.

Funding

No funding or grant support.

Authorship

All authors attest that we meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The following authors have no financial disclosures: M.Y., F.M., L.C., Y.C.

Acknowledgements

Mingzhen Yuan and Feiyan Ma are co-first authors; they contributed equally to the work.

References

1. Hannan SR, et al. Common spectral domain OCT and electrophysiological findings in different pattern dystrophies. *Br J Ophthalmol*. 2013;97(5):605–610.
2. Arnold JJ, et al. Adult vitelliform macular degeneration: a clinicopathological study. *Eye (Lond)*. 2003;17(6):717–726.
3. Chowers I, et al. Adult-onset foveomacular vitelliform dystrophy: a fresh perspective. *Prog Retin Eye Res*. 2015;47:64–85.
4. Everdell NL, et al. Multispectral imaging of the ocular fundus using light emitting diode illumination. *Rev Sci Instrum*. 2010;81(9), 093706.
5. Levenson RM, Fornari A, Loda M. Multispectral imaging and pathology: seeing and doing more. *Expert Opin Med Diagn*. 2008;2(9):1067–1081.
6. Huang G, et al. Multispectral image analysis in Vogt-Koyanagi-Harada disease. *Acta Ophthalmol*. 2018;96(4):411–419.
7. Zhu X, et al. Sensitivity and specificity of multispectral imaging in detecting central serous chorioretinopathy. *Laser Surg Med*. 2017;49(5):498–505.
8. Dugel PU, Zimmer CN. Imaging of melanin disruption in age-related macular degeneration using multispectral imaging. *Ophthalmic Surg Lasers Imag Retina*. 2016;47(2):134–141.
9. Pang CE, et al. New insights into Stargardt disease with multimodal imaging. *Ophthalmic Surg Lasers Imag Retina*. 2015;46(2):257–261.
10. Xu Y, et al. A light-emitting diode (LED)-based multispectral imaging system in evaluating retinal vein occlusion. *Laser Surg Med*. 2015;47(7):549–558.
11. Gass JD. A clinicopathologic study of a peculiar foveomacular dystrophy. *Trans Am Ophthalmol Soc*. 1974;72:139–156.
12. Makati R, et al. Electrooculography and optical coherence tomography reveal late-onset Best disease. *Optom Vis Sci*. 2014;91(11):e274–e277.
13. Querques G, et al. Optical coherence tomography angiography in adult-onset foveomacular vitelliform dystrophy. *Br J Ophthalmol*. 2016;100(12):1724–1730.
14. Freund KB, et al. Acquired Vitelliform Lesions: correlation of clinical findings and multiple imaging analyses. *Retina*. 2011;31(1):13–25.
15. Querques G, et al. High-definition optical coherence tomography features in vitelliform macular dystrophy. *Am J Ophthalmol*. 2008;146(4):501–507.