Graefes Arch Clin Exp Ophthalmol
DOI 10.1007/s00417-015-3058-3

LETTER TO THE EDITOR

Spectral domain optical coherence tomography imaging of mucopolysaccharidoses I, II, IV A, and VI

Yuan-Chieh Lee [1,2,3,4]

Received: 21 April 2015 / Accepted: 14 May 2015
© Springer-Verlag Berlin Heidelberg 2015

Dear editor,

We read with great interest the article entitled "Spectral-domain optical coherence tomography imaging of mucopolysaccharidoses I, II, and VI A" by Seok et al. [1]. Although the study provides important data, we have several comments. First, there is a typing error in the title. It should be "…mucopolysaccharidoses I, II, and IV A" instead of "VI A". Second, as for mucopolysaccharidoses (MPS) type VI, we do have some spectral-domain optical coherence tomography (SD-OCT) images of MPS type VI, which show depigmented retinopathy and thinning of the choroid [2].

Third, thinning of the choroid might play a role in the development of pigmentary retinopathy in MPS I, II, and VI, and we suggested that the SD-OCT with enhanced depth imaging might help evaluate the chorioretinopathy of MPS patients [2].

Fourth, Seok et al. described cystoid macular edema in case 7 (MPS II). We also observed prominent cystic spaces in the inner nuclear layer and outer nuclear layer of a MPS II patient, but those cystic spaces extend far beyond macular area (beyond 6,000 μm diameter). The character of the contents of the cystic spaces is still unknown and needs further study. However, those cystic spaces resolved with time in our patient, who did not receive enzyme replacement therapy. No loss of ellipsoid zone was noted 3 years after resolution of those cystic spaces. Therefore, those cystic spaces or fluid accumulation might not be directly related to the loss of ellipsoid zone or degeneration of photoreceptor cells.

Fifth, their study showed central foveal external limiting membrane (ELM) thickening and inner segment/outer segment (IS/OS) thinning in MPS I and II. The ages of their patients were between 9 and 20. We also observed thickened ELM in the central foveal area and loss of ellipsoid zone in the extrafoveal area. The thickened ELM at central foveal area was noted in MPS I and MPS II, but not so evident in MPS VI. The ELM is formed by the attachment site of the Müller cells and the adjacent photoreceptor cells. It is possible that accumulation of mucopolysaccharides might lead to the activation of the Müller cells and hence thickened ELM [2]. Although IS/OS thinning at fovea was demonstrated in their study, IS/OS thinning was more marked in the perifoveal (case 6, 7, 8, 10, and 11) and parafoveal areas (case 10). In our study, loss of ellipsoid zone was mainly noted in extra foveal area, and the remaining ellipsoid zone became smaller with advancing age. In two of our MPS II patients, aged 46 and 49 years old respectively, the fundus examination revealed pigmentary retinopathy mimicking retinitis pigmentosa. The SD-OCT showed that the ellipsoid zone existed only in the very central fovea. However, both patients still kept a visual acuity of 20/40 in each eye and intact color discrimination. Therefore, whether rod cells are involved more than cone cells and the significance of IS/OS thinning at fovea need further investigation.

Lastly, in their study, five eyes of three MPS IV A patients showed normal OCT results. Our study also demonstrated similar results in 16 eyes of eight MPS IV A patients.

✉ Yuan-Chieh Lee
yuanchieh.lee@gmail.com

[1] Department of Ophthalmology, Buddhist Tzu Chi General Hospital, 707, Section 3, Chung-Yang Road, Hualien, Taiwan

[2] Department of Ophthalmology and Visual Science, Tzu Chi University, Hualien, Taiwan

[3] Institute of Medical Science, Tzu Chi University, Hualien, Taiwan

[4] Department of Ophthalmology, National Taiwan University Hospital, Taipei, Taiwan

Published online: 27 May 2015

Conflict of interest The author certifies that he has NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Informed patient consent and IRB approval (IRB103-162-A) were obtained for the study.

References

1. Seok S, Lyu IJ, Park KA, Oh SY (2015) Spectral domain optical coherence tomography imaging of mucopolysaccharidoses I, II, and VI A. Graefes Arch Clin Exp Ophthalmol. doi:10.1007/s00417-015-2953-y/fulltext.html
2. Huang CT, Chu SY, Lee YC (2015) Optical coherence tomography of chorioretinopathy caused by mucopolysaccharidoses. Ophthalmology. doi:10.1016/j.ophtha.2015.01.015