Optical Coherence Tomographic Findings of Optic Nerve Head and Retina in Infants and Young Children with Familial Exudative Vitreoretinopathy

Jonghyun Lee1,2, Maysontoine El-Dairi1, Du Tran-Viet1, Shwetha Mangalesh1, Malini Veerappan1, Alexandria Dandridge1, Kim Jiramongkolchai1, Cynthia A. Toth1,2

1Department of Ophthalmology, Duke University Medical Center, Durham, NC; 2Department of Ophthalmology, Inje University, Ilsan Paik Hospital, Goyang, Gyung-gi-do, Korea (The Republic of)

PURPOSE

- Knowledge regarding familial exudative vitreoretinopathy (FEVR) optical coherence tomographic findings, especially in infants and young children, is limited because imaging can be difficult due to inability to cooperate.
- To explore hand-held spectral domain optical coherence tomography (HH-SDOCT) findings in optic nerve head and retina in young FEVR patients.
- To explore the range of vitreo-retinal pathologies visible on HH-SDOCT images in young FEVR patients.
- To investigate those changes during follow-up period as the disease evolves.

METHODS

- Single center, retrospective case series of patients with a clinical diagnosis of FEVR from January 1, 2009, to December 31, 2015.
- Fundusphotographs and fluorescein angiography in children were obtained during examination under anesthesia using the RetCam II Wide-Field Digital Imaging System (Clarity Medical System Inc., Pleasanton, CA) and the 130° widefield D1300 lens.
- All SDOCT images were obtained with the portable HH-SDOCT system centered at 840nm wavelength (Biopixgen Inc., Research Triangle Park, NC) with prior consent, under an IRB-approved research protocol.
- A total 26 eyes from 16 patients were included in this study. (12 male and 4 female)

RESULTS

- In fundus photography, optic disc draging was obvious in 8 eyes and suspicious in 2 eyes. (left to right from suspicious to severe disc dragging)
- Those 10 eyes had retinal draging as a movement of neurosensory retina (green arrows) toward the lesion side in OCT. (mainly temporally) All eyes except one, which already underwent pars plana vitrectomy, had strong vitreo-papillary adhesion or prominent vitreo-papillary traction in OCT. (yellow arrows)
- We found same OCT findings in 2 eyes that developed the disc draging later during follow up.
- In addition to the temporal movement of nasal neurosensory retina (nasal optic disc), we observed pulling-up of optic nerve fiber as a finding of retinal nerve fiber layer(RNFL) thickening at temporal, superior and inferior optic disc margin. (green arrows)
- In 10 eyes among 12 eyes with optic disc draging (including 2 eyes that developed disc draging later), OCT revealed retinal elevation (arrows) temporal to optic disc, which was greatest at optic disc area and became lower as it extended farther away from the optic disc. Retinal elevation was predominantly due to the thickening of nerve fiber layer in all cases, and the apex of the elevation was centered over large retinal vessels. (posterior shadowing)
- At the first examination session, 16 eyes had no noticeable abnormal finding in optic disc area in fundus photography but, in OCT, 12 eyes had optic disc elevation. Among those 12 eyes, 7 eyes had vitreo-papillary traction or pre-papillary membrane, 1 eye had mild optic disc edema and 4 eyes had protrusion of retinal vessel at optic disc.

FINANCIAL DISCLOSURES

Cynthia A. Toth; Alcon (F), Biopixgen (F), Genentech (F), NIH R01 EY025009 (F), NIH P30 EY01583 (F), NCCR. IULIKR024128-01 (F). All other authors: None.

CONCLUSIONS

- OCT revealed many abnormalities in various stages of FEVR which were not apparent in photography. Also, OCT helped to detect early changes in optic disc, e.g. edema, pre-papillary membrane, retinal draging and vitreous organization.
- When peripheral retinal fold by fibrovascular membrane combines with optic disc draging (with retinal elevation), a radial retinal fold is generated.
- The vitreous has an important role in pathogenesis and progression of FEVR. Pathologically adherent and contractile nature of vitreous in young FEVR patients can cause many tractional complications in FEVR without PVD. This is different from vitreous tractional complications in the adult, which are almost always associated with a partial PVD or during PVD progression.
- Not only retinal draging (conventional concept) but also strong vitreous traction at the disc and retina contribute to the formation of a radial retinal fold in young children (in addition to disease progression).

REFERENCES


ACKNOWLEDGEMENTS

The Andrew Family Charitable Foundation

Correspondence: ophth@naver.com or lee.jonghyun@duke.edu