Optical Coherence Tomography Angiography in Fovea Plana

Rosa Dolz-Marco, MD, PhD; Nopasak Phasukkijwatana, PhD, MD; David Sarraf, MD; K. Bailey Freund, MD

ABSTRACT: Fovea plana is characterized by the anatomical absence of the foveal pit in eyes with normal visual function. The authors have analyzed three cases of idiopathic fovea plana with optical coherence tomography angiography (OCTA). As previously reported, the authors found the absence of a foveal avascular zone in all cases with OCTA; however, a preserved fusion of both the superficial and the deep capillary plexuses was found around the foveal center. This novel observation cannot be detected with conventional dye-based angiography, in which the deep capillary plexus is not visualized.

[Introduction]

Foveal hypoplasia refers to the abnormal anatomical absence of the foveal pit.1 Various conditions have been associated with this disorder, including ocular albinism, retinopathy of prematurity and premature birth, aniridia, nanophthalmos, incontinentia pigmenti, and achromatopsia,1,4 whereas some cases are presumed to be idiopathic.1,3 The anomalous development of the foveal pit has been related to the absence or reduction of the foveal avascular zone (FAZ).2,5 Marmor et al. introduced the term “fovea plana” to describe eyes showing a flat fovea with normal visual function, thus reserving the term “foveal hypoplasia” for those cases with visual impairment. The presence of fovea plana has been reported in up to 3% of children with normal visual acuity.3 We report the abnormalities of the FAZ in patients with idiopathic fovea plana analyzed by optical coherence tomography angiography (OCTA) using the split-spectrum amplitude decorrelation angiography algorithm on scans obtained with the Optovue RTVue XR Avanti (Optovue, Fremont).

[CASE REPORTS]

Case 1

A 32-year-old woman who denied a history of prematurity presented with visual symptoms in her right eye due to choroidal neovascularization secondary to multifocal choroiditis. Following a single intravitreal injection of ranibizumab, visual acuity improved from 20/30 to 20/20.

Funduscopy examination showed bilateral mild atrophic round macular lesions. Fluorescein angiography (FA) showed a bilateral absence of the FAZ. The spectral-domain OCT (SD-OCT) images showed flattening of the foveal contour with loss of the foveal pit. En face OCTA demonstrated a continuous plane of vessels with an absent FAZ at the level of the superficial capillary plexus (SCP) and a barely discernible FAZ in the deep capillary plexus (DCP). Flow B-scans illustrated anastomosis of both the SCP and the DCP, merging into a single capillary layer at the center of the fovea (Figure 1).

Case 2

A 64-year-old woman who denied a history of prematurity presented with peripheral retinal...
breaks in both eyes that were treated with laser photocoagulation. Visual acuity was 20/20 in her right eye and 20/30 in her left eye.

Funduscopic examination was unremarkable. SD-OCT demonstrated bilateral flattening of the foveal contour. En face OCTA of the SCP showed a continuous plane of vessels without a FAZ. The DCP showed a small avascular area at the corresponding foveal center. Flow B-scans illustrated anastomosis of the DCP and SCP merging into a single vascular layer at the central fovea (Figure 2).

Case 3
A 72-year-old woman with bilateral pseudophakia denied a history of prematurity. Visual acuity was 20/30 in both eyes. Funduscopic examination was unremarkable. En face OCTA of the SCP demonstrated a continuous plane of vessels across the

Figure 1. Multimodal imaging of idiopathic fovea plana associated with multifocal choroiditis. Case 1: (A, C) Color photograph of the right eye illustrates an extrafoveal choroidal neovascular membrane associated with multiple small atrophic lesions. In the left eye, there are only subtle pigmentary changes. (B, D) Fluorescein angiography in both eyes shows the absence of a foveal avascular zone (FAZ). (E, F) Optical coherence tomography (OCT) scans corresponding to the yellow lines in Figures 1A and 1C illustrate absence of the foveal pit with preservation of inner retinal layers through the foveal center. The outer retinal layers are anatomically intact through the central fovea. (G, I) En face OCT angiography (OCTA) segmented at the level of the superficial capillary plexus (SCP) and corresponding to the yellow boxes in Figures 1B and 1D show complete vascularization of the fovea and loss of the FAZ, whereas segmentation of the deep capillary plexus (DCP) (H, J) shows an attenuated vascular density the central macula. Cross-sectional OCTA scans at the corresponding blue line in Figures 1A and 1C demonstrate flow of the SCP and the DCP throughout the macula with a merging of these plexuses at the central fovea, creating a single central flow signal (white lines).
fovea in the absence of a FAZ. The DCP demonstrated a reduction in vascular density at the central fovea. Central anastomosis of the DCP and SCP was shown in the flow B-scans (Figure 2).

**DISCUSSION**

Clinically, the FAZ is often assessed with FA, which shows irregularly shaped variations and a mean area of $0.43 \text{ mm}^2 \pm 0.16 \text{ mm}^2$. On FA, the SCP is easily imaged, but the DCP is poorly visualized. The advent of OCTA has provided a unique window to separately evaluate the superficial and deep retinal capillary plexuses of the macula. Using this technology to evaluate the FAZ in healthy patients, Samara et al. demonstrated larger avascular areas at the level of the DCP compared with the SCP. They also found an inverse relationship between the central foveal thickness and the size of the FAZ. Our results correlate with this observation, as patients with fovea plana showed a greater central foveal thickness in the absence of a foveal pit, with an extreme decrease in the foveal avascular area and consequently an absence of the FAZ.

The development of the anatomy and function
of the fovea include three processes: the centrifugal
displacement of the inner retinal layers, the cone
photoreceptor specialization, and the centripetal
migration of the cones to the center of the fovea.
These processes can be observed on OCT as 1) a
foveal pit with an absence of the inner retinal layers,
2) outer segment lengthening, and 3) outer nuclear
layer widening.1 Our patients showed a normal ap-
ppearance of the outer retina on structural OCT with
thickening of the outer segments of the photorecep-
tors and the outer nuclear layer, but they lacked a
foveal pit due to failure of centrifugal migration of
the inner retinal layers. We found an incomplete
development of the foveal vascular pattern, show-
ing an absence of the FAZ with preservation of the
normal fusion of the SCP and DCP occurring in the
fovea. This novel observation cannot be detected
with conventional dye-based angiography in which
the DCP is poorly visualized. As the OCTA review
software shows the segmentation lines used for en
face analysis, we were able to determine that seg-
mentation errors did not account for our findings.
In conclusion, we report three cases of idiopath-
ic fovea plana with preserved visual function all
showing an absent FAZ in the SCP and an absent
or markedly reduced FAZ in the DCP with OCT an-
giography. As reported previously, eyes with fovea
plana lack a FAZ,2,5 but this report more clearly
eucidates the level and nature of this vascular ab-
normality. In addition, we illustrated merging of
the SCP and the DCP into a single foveal capillary
plexus at the center of the fovea in the absence of a
FAZ. Further analysis in a larger series of cases will
be required to confirm these findings and to deter-
mine if they occur with true foveal hypoplasia as-
associated with poor visual function and nystagmus.

REFERENCES

1. Thomas MG, Kumar A, Mohammad S, et al. Structural grad-
ing of foveal hypoplasia using spectral-domain optical coher-
ce tomography a predictor of visual acuity? Ophthalmology.
2011;118(8):1653-1660.
2012;96(7):961-966.
4. Marmor MF, Choi SS, Zawadzki RJ, Werner JS. Visual insignifi-
cance of the foveal pit: reassessment of foveal hypoplasia as fovea
5. Dubis AM, Hansen BR, Cooper RF, Beringer J, Dubra A, Carroll
J. Relationship between the foveal avascular zone and foveal pit
6. Wu LZ, Huang ZS, Wu DZ, Chan E. Characteristics of the cap-
7. Spaide RF, Klancnik JM, Cooney MJ. Retinal vascular layers imaged
by fluorescein angiography and optical coherence tomography an-
8. Samara WA, Say EA, Khoo CT, et al. Correlation of foveal avascu-
lar zone size with foveal morphology in normal eyes using optical
coherence tomography angiography. Retina. 2015;35(11):2188-
2195.