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Case report

Absence of the foveal avascular zone in a nanophthalmic child revealed by optical coherence tomography angiography

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ABSTRACT

Purpose: Optical coherence tomography angiography (OCTA) is a new non-invasive imaging technique that does not require the use of contrast agents and that allows the visualization of the retinal microvasculature in a layer-by-layer manner without bright light. This merit allows us to obtain the fundus image in children. Retinal vessels are typically absent from the center of the fovea, an area known as the foveal avascular zone (FAZ). The purpose of the present case study was to evaluate the FAZ in a nanophthalmic pediatric patient with OCTA.

Obsevations: A 6-year-old girl was referred to the Hiroshima University Hospital because of her poor vision. She had a best-corrected visual acuity of 20/125 in the right eye and of 20/100 in the left eye. The refractive errors after the administration of atropine sulfate eye drops were +13.00D in the right eye and +14.00D in the left eye. The axial lengths were 17.03 mm in the right eye and 16.90 mm in the left eye. At 9 years of age, the patient was diagnosed with nanophthalmos and OCTA was used to investigate the superficial and deep retinal layers. We demonstrated that the FAZ could not be observed in either eye, whereas the FAZ was readily observed in both eyes of a control subject of similar age.

Conclusion and Importance: OCTA is a useful technique to reveal the absence of the FAZ in cases of nanophthalmos. Because OCTA is a non-invasive and rapid procedure that is ideal for use with children.

1. Introduction

Nanophthalmos, in the absence of other congenital malformations, is a bilateral and often familial form of microphthalmos.[1] It is characterized by hyperopia, a small cornea, thick sclera, and narrow iridocorneal angles.[2,3] Although by definition, nanophthalmos refers to eyes with an axial length of less than 20 mm in adults, there is a gradient of hyperopic refraction where the degree of hyperopia is inversely correlated with the axial length of the eye.[1] Nanophthalmos is also reported to have characteristic structural features when observed with optical coherence tomography (OCT) such as a thickening and hypoplasia of the macula.[4]

Macular blood vessels are primarily capillaries that are located in the ganglion cell layer of the foveal edge and along the foveal slope. However, there is small area approximately 0.4 mm in diameter near the center of the macula, known as the foveal avascular zone (FAZ) which is devoid of retinal vessels.[5] OCT angiography (OCTA) a recently developed non-invasive technique to visualize the retinal microvasculature, is an optimal tool to observe the retinal layers without the use of a contrast agent.[6,7] There is, however, currently little information available regarding the foveal vascular structure of the developing nanophthalmic eye. To address this knowledge gap, we used OCTA to examine the retinal microvasculature of a pediatric patient with nanophthalmos and revealed that the FAZ was absent in the superficial and deep retinal layers.

2. Case report

[Patient] A six-year-old girl.

[Main complaint] Bilateral poor visual acuity.

[Medical history] The patient consulted her local ophthalmologist complaining of poor vision in both eyes. She was found to have extreme hyperopia and was suspected of having ametropic amblyopia. She was therefore referred to the Hiroshima University Hospital.

[Personal and family history] There was no remarkable family history.

[Findings] The patient's best-corrected visual acuity at the first visit was 20/125 with a refractive correction of +13.00 spherical diopters (D) for the right eye and 20/200 with +14.00 spherical D for the left eye. Her eye positions were judged orthophoric, both before and after the application of atropine sulfate eye drops. The cornea was clear, and the corneal diameter was 10 mm in both eyes. Using a slit lamp

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Fig. 1. Fundus photography. This figure illustrates a reduction in the foveal light reflex as evidenced by fundus photography.

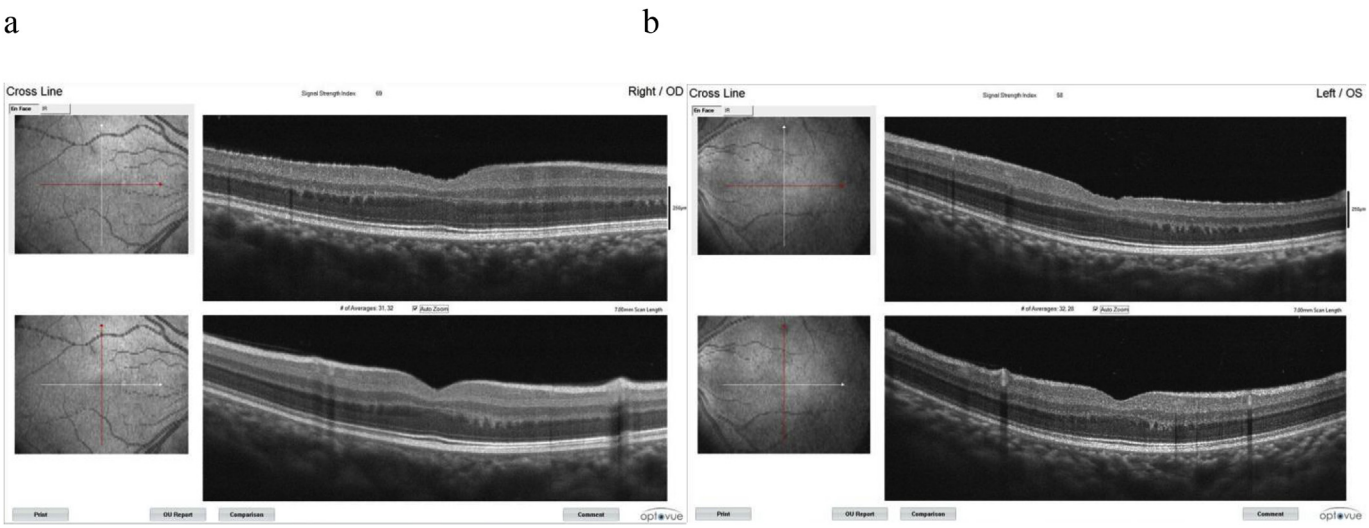


Fig. 2. Foveal ocular coherence tomography scans. a: right eye, b: left eye. The foveal thickness was 340 μm in the right eye and 352 μm in the left eye. The fovea centralis was small and poorly demarcated.

examination, no abnormalities were found in the anterior segment or in the media. A fundus photograph showed a reduction of the foveal light reflex in both eyes (Fig. 1). The axial lengths were short: 17.03 mm in the right eye and 16.90 mm in the left eye. The OCT findings showed that the foveal thickness was 340 μm in the right eye and 352 μm in the left eye and that the central fovea was poorly demarcated (Fig. 2).

[Clinical course] The patient was diagnosed with nanophthalmos and ametropic amblyopia and was prescribed complete corrective glasses to treat the ametropic amblyopia. After 2 years, the patient's best-corrected visual acuity had improved to 20/32 in the right eye and to 20/25 in the left eye.

Three years after the first visit, at age 9, we performed an OCTA (RTVue XR Avanti®, Optovue Co., Fremont, California, USA) to examine the foveal vasculature and compare it with that of a normal control subject (Fig. 3b and c). The control subject was a 10-year-old girl with strabismus that had normal visual acuity and normal refractive errors (< +2D) (Fig. 3, a). The FAZ was clearly observed in both eyes of the control subject, both in superficial and deep retinal layers. The FAZ observed in the deep layer was larger than that in the superficial layer. In contrast, no FAZ was found in the superficial or deep retinal layers of either eye in our patient with nanophthalmos.

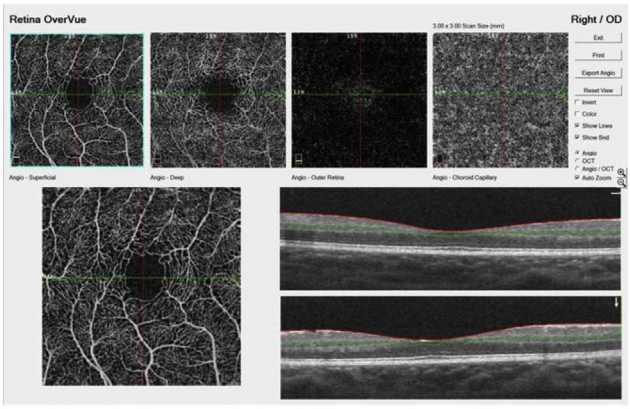
3. Discussion

We herein describe the vascular structure of the macula in a child with nanophthalmos in a layer-by-layer manner using OCTA. OCTA allows for the visualization of the superficial capillary plexus, the deep capillary plexus, the outer retinal zone, and the choriocapillaris. Using OCTA, we demonstrated the absence of the FAZ in the superficial and deep retinal layers in nanophthalmic eyes of 9 year old girl. We defined the absence of the FAZ there was a persistence of retinal vasculature in the fovea on OCTA. In contrast, the FAZ was readily observable in the eyes of a control patient. Although fluorescein angiography (FA) allows for the visualization of eye vascular information, it requires the injection of a fluorescein dye and subsequent exposure to bright light. OCTA, in contrast, is non-invasive, does not require a contrast agent, and takes less time to perform than FA, making it an ideal visualization technique to use with children.

Khariallah et al. reported that absent or marked reduction of the capillary-free zone on fluorescein angiography was a constant feature in nanophthaloms eyes, but they did not refer to the specific vascular structure.[8] On the other hand, OCTA revealed the presence of microvasculature in the superficial and deep foveal retinal layers of nanophtalmos eyes.

Majima et al. defined nanophthalmos in adults as eyes presenting with an axial length less than 20.4 mm in males and 20.1 mm in females.[9] Majima et al. also reported that the average axial length for normal 6- to 7-year-old girls was 21.68 mm, whereas it was less than 18.86 mm for those presenting with nanophthalmos. The findings from our case study are therefore consistent with the established diagnostic

a



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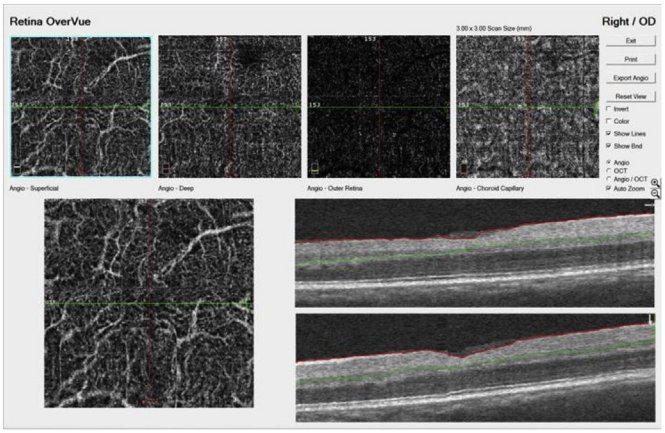

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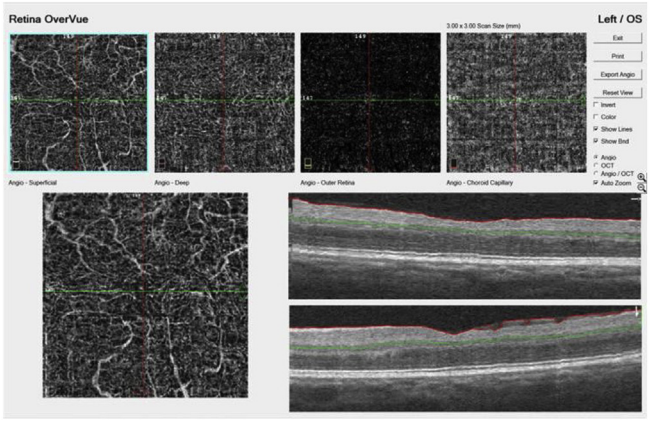

Fig. 3. OCT angiography scans. a: 10-year old normal right eye, b: the right eye of the subject with nanophthalmos, c: left eyes of the subject with nanophthalmos. The top row of each panel (from left to right) shows the superficial capillary plexus, the deep capillary plexus, the outer retinal zone, and the choriocapillaris. The bottom row of each panel (from left to right) shows the superficial capillary plexus and the OCT scan, where the part in between the red and green line indicates the superficial capillary plexus. Although the FAZ was clearly observable in both eyes of the control subject, no FAZ was found in the superficial or deep retinal layers in either of the nanophthalmic eyes. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

criteria of nanophthalmos. Sampolesi et al. proposed a method to calculate the expected eye axial length of normal children as follows: 18.7 + 2.245*(logarithm x), where x corresponds to the child's age (in months).[10] Based on this formula, the average axial length for a child of 6 years and 4 months of age should be approximately 22.9 mm, which is again consistent with our findings.

Chen et al. reported that the average thickness of the macula in children (194 eyes from 194 children, mean age 10.15 ± 2.61 years old, average refractive power −2.25 ± 2.47D) was 240.25 ± 19.85 μm and that this measure was not affected by myopia.[11] Read et al. examined the macular retinal thickness using a Spectral-domain Optical Coherence Tomography instrument (Copernicus SOCT-HR; Optopol Technology SA, Zawiercie, Poland) in 196 children aged between 4 and 12 years of age (mean age: 8 ± 2 years old, mean spherical equivalent refraction of +0.06 ± 0.21D, range: +1.25 to −0.50D). The mean total retinal thickness in the central 1-mm foveal zone was 255 ± 16 mm and increased significantly with age (mean increase of 1.8 mm per year).[12] In our case subject, the macular thickness was 340 μm in the right eye and 352 μm in the left eye. Therefore, the macula was thicker in both eyes of our patient compared with normally developing eyes. Furthermore, our data support the report published by Bijlsma et al.[4] that short axial length is correlated with the retinal thickness.

OCTA showed there is no FAZ in children with nanophthalmos and ametropic amblyopia. We also examined the macula OCTA in children with another ametropic amblyopia, 4 eyes of 2 control patients with an average age of 7 years old. Children with hypermetropic ametropic amblyopia and normal axial length also had FAZ, therefore the presence of FAZ may not be the essential factor to hypermetropia and amblyopia.

The fovea is a specialized retinal area that is responsible for sharp central vision. The normal fovea does not have an inner retinal layer and contains a large number of cone cells. In the center of the fovea is localized an avascular area, the FAZ, which allows light to traverse the retina without any dispersion or loss. Walsh et al. reported that this central portion of the fovea is important for obtaining a best-corrected visual acuity above a certain threshold, whereas an absent or rudimentary FAZ may limit the visual potential.[6] The exact relationship between the size of the FAZ and visual acuity, however, remains unclear. Longitudinal studies will be useful for evaluating the relationship between the surface area of the FAZ and visual acuity by determining whether the size of the FAZ changes with age in the same patient. Because OCTA is a non-invasive and rapid procedure that is ideal for use with children, it will allow researchers to more easily address this important research question.

We could not correct for the axial length of the nanophthalmic eye by offsetting the depth of the OCTA slabs. This is one of limitations of our study.

Patient consent

The patient's legal guardian consented to publication of the case in orally. This report does not contain any personal information that could lead to the identification of the patient.

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Conflicts of interest

All authors have no financial disclosures.

Authorship

All authors attest that they meet the current ICMJE criteria for

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