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Brief report

Multimodal imaging in posterior microphthalmos

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Abstract

Purpose: To evaluate the multimodal imaging including optical coherence tomography angiography (OCTA) findings in patients with posterior microphthalmos (PM).

Methods: In an observational case series, four eyes of two patients, eight and twenty-three years old, with clinical proven PM underwent complete ophthalmic examination, including refraction, fluorescein angiography, optical coherence tomography (OCT), OCTA, B-scan ultrasonography, axial length measurement using IOL Master optical measuring, and Pentacam evaluation.

Results: Both patients were high hyperopic with partial thickness retinal fold in macula, retinoschisis, and foveal hypoplasia. Axial length was less than 17 mm with scleral thickening in all eyes. OCTA showed absence of foveal avascular zone (FAZ) in both superficial and deep capillary plexuses. Pentacam showed corneal steepness, shallow anterior chamber, and low anterior chamber volume.

Conclusion: OCTA findings showed absence of avascular zone in both superficial and deep capillary plexuses, while OCT shows partial thickness retinal fold and retinoschisis.

Keywords: Posterior microphthalmos; Optical coherence tomography angiography; Retinal fold; Macula

Introduction

Microphthalmos is a rare developmental disorder of the eye which is defined by shortness of axial length at least 2 SD below the mean of the normal age group.[1] Total microphthalmos was categorized into anophthalmos (extremely small eyes), simple (or pure) type, and a complex (or complicated with other malformations) type.[2] Nanophthalmos is a variant of this anomaly which is characterized by a short total axial length but no other ocular malformations. Posterior microphthalmos (PM) is a subclass of microphthalmos that specifically affects the axial length of the posterior segment of the eye, and posterior pole changes were found, like macular fold or retinoschisis. In a series of PM and nanophthalmos patients reported by Relhan et al. macular structure abnormalities have been reported to be solely associated with PM whereas pigmentary retinopathies were predominantly seen in nanophthalmos patients.[1] The main findings in patients with PM are high hyperopia and an elevated papillomacular retinal fold. Papillomacular fold usually involves retina, mostly inner retina, but can be full thickness retinal folds. Chorioretinal folds can be seen occasionally. Other posterior segment abnormalities include absence or marked reduction of the foveal avascular zone (FAZ), sclerochoroidal thickening, crowded optic discs, pseudopapilledema, uveal effusion syndrome, serous retinal detachment, retinoschisis, macular hole,

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pigmentary retinopathy, and avascular zone in the extreme periphery of retina.[3,4]

Optical coherence tomography angiography (OCTA) is a new non-invasive imaging modality that visualizes vasculature by detecting motion without the need for injecting dye. Additionally, it provides three dimensional volumetric enface images of the retina and choroid vessels using structural optical coherence tomography (OCT) information. OCTA enables visualization of deep capillary plexus of retina which is not possible with fluorescein angiography.[5]

The purpose of this report is describing the findings of two patients with PM using multimodal imaging. Due to the presence of reduced FAZ, we employed OCTA to evaluate the vasculature of macular area at different levels. To the best of our knowledge, it is the first report of OCTA findings in PM patients.

Methods

An eight-year-old male patient was presented to our clinic with reduced vision in both eyes. Refraction was +11.75 in the right eye and +11.75−1.00 × 160 in the left eye. Best corrected visual acuity (BCVA) was 20/60 in both eyes. Cornea and lens were clear and intraocular pressures (IOPs) were within the normal range. In funduscopy, there was a horizontal fold in the papillomacular area. Optic disc was small and crowded. Axial length was measured 16.86 mm and 16.93 mm by IOL Master (Carl Zeiss Meditec, Jena, Germany), in right and left eye, respectively. B-scan showed thickening of sclera. OCT (HRA, Heidelberg Engineering, Heidelberg, Germany) demonstrated an elevated papillomacular fold involving only inner part of sensory retina (anterior to outer nuclear layer) with sparing outer retina and underlying retinal pigment epithelium (RPE) and choroid. All the retina layers could be appreciated in the fovea, and there was also a macular retinoschisis. OCTA using the AngioVue XR Avanti device (RTVue XR; Optovue Inc) revealed absence of FAZ with demonstrating vascularized superficial and deep capillary plexus (Fig. 1). In anterior segment, Pentacam (Oculus, Wetzlar, Germany, Scheimpflug camera) showed steep corneas with K1 48.5 D and K2 of 49.7 D for the right eye and K1 49.1 D and K2 of 50.0 D for the left eye. Anterior chamber depth was 2.70 mm and 2.64 mm and anterior chamber angle was 25.2° and 32.5° in the right and left eyes, respectively. Corneal horizontal white to white diameter was 11.6 mm in the right eye and 11.7 mm in the left eyes.

A twenty-three-year-old hyperopic man was referred for refractive surgery. Refraction was +15.50 D in right eye and +16.00 D in the left eye. BCVA was 20/60 and 20/100 in the right and left eyes, respectively. Cornea and lens were clear, and IOPs were within normal limits. Pentacam showed superior corneal steepening with K1 48.6 D and K2 of 50.3 D for the right eye and K1 48.7 D and K2 of 50.9 D for the left eye. Anterior chamber depth was 2.61 mm and 2.59 mm in the right and left eyes, respectively. Anterior chamber angle was measured 33.7° in the right eye and 30.3° in the left eye. Corneal horizontal white-to-white diameter was measured 11.8 in both eyes. In funduscopy, horizontal superficial papillomacular retinal fold was observed. Small crowded optic disc was seen. Axial length was measured 16.43 mm and 16.37 mm, in right and left eyes, respectively. OCT demonstrated papillomacular fold of partial thickness sensorial retina without involvement of outer retina and underlying RPE and choroid. All the retinal layers were represented throughout the

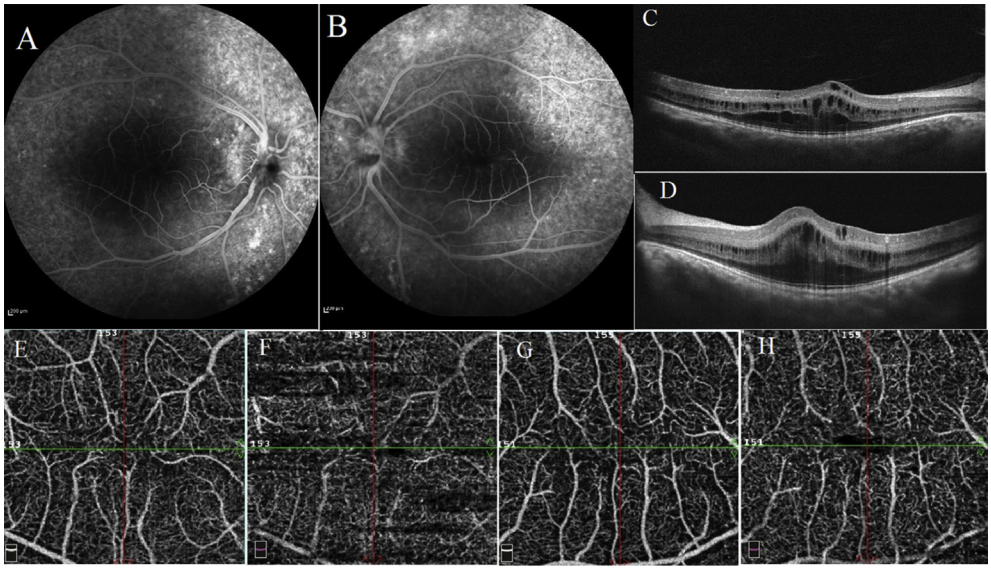

Fig. 1. Multimodal imaging of the first case. A and B show the fundus photo. Horizontal elevated papillomacular retinal fold is seen in both eyes. C and D show elevated retinal fold and retinoschisis with intraretinal fluid. No foveal depression is seen, and all layers are preserved all over the retina. E and G superficial plexus of optical coherence tomography angiography (OCTA), and F and H showing deep capillary plexus of OCTA, showing complete foveal vascularization. I shows thickened sclera and shortened eye in B scan ultrasonography. J shows corneal superior steepening in pentacam topography.

macular area. Macular retinoschisis was obvious. Fluorescein angiography showed reduction in size of the FAZ without leakage into the macular microcystoid spaces. OCTA showed vascularization of the fovea and absence of FAZ. B-scan showed thick sclera and small eye, even in axial length or in equatorial circumference (Fig. 2).

Discussion

PM is a rare congenital anomaly that is known to disproportionately affect the posterior segment of the eye. We found subnormal dimensions of anterior chamber depth and angle in our cases as well, suggesting that this entity may affect anterior segment as well although with less severity compared to posterior segment. The four eyes reported here showed retinal papillomacular fold, absence of FAZ, retinoschisis, and crowded optic discs. Macular folds are more common in PM compared with nanophthalmos.[6]

There are different explanations for the absence of FAZ in PM patients. It is postulated that an abnormally thickened sclera during the organogenesis inhibits the growth of choroid and RPE; resulting in formation of a papillomacular fold which obscures the depression of fovea. Others believe in true foveal hypoplasia with continuity of inner retinal layers in these patients.[7,8]

OCT features of papillomacular fold of PM have been previously described.[9] These include intraretinal cystic cavities within the inner nuclear and ganglion cell layer. It has also been shown that despite the presence of inner retinal fold, photoreceptor outer segments, ellipsoid zone, external limiting membrane and outer nuclear layers are usually preserved.[10,11] Therefore, it can be concluded that the potential for vision is good, and the main reason for decreased visual acuity in these patients is high hyperopic amblyopia. However, this is in contrast to our findings in OCTA which showed complete vascularization of fovea and absence of FAZ and also macular hypoplasia and all layers saving in OCT findings. This finding suggests that fovea may have an arrest in development during embryogenesis similar to true foveal hypoplasia and propose that this may partially contribute to poor vision in these patients despite receiving good refractive correction. Previous studies that used time domain optical coherence tomography (TD-OCT)[4,8,12] and spectral domain optical coherence tomography (SD-OCT)[9] to evaluate the patients with PM also showed the absence of foveal depression and presence of all retinal layers in the macular area of eyes with this condition. Despite of presence of intraretinal cystic cavities in the macula of both patients on SD-OCT, fluorescein angiography of case number 2 was unable to show any leakage in this area. It is believed that the cystic cavities in the macula of patients with PM are inherently different from cystic changes observed in patients with diabetic retinopathy and retinal vascular abnormalities. It is speculated that these cystic changes are the result of structural abnormalities rather than the accumulation of extracellular fluid and breakdown of blood retinal barrier.[9] It has been shown that there is a reverse relationship between axial length and corneal power in PM patients.[7] Similarly, we found cornea steepening in our patients.

In conclusion, we described the multimodal imaging features of eyes with PM and the OCTA features for the first time. Moreover than OCT which shows us the retinal fold and the retinoschisis, OCTA shows absence of avascular zone in both superficial and deep capillary plexuses. This new imaging technique, combined with other imaging modalities can help us better understand this rare congenital anomaly. Further studies with a larger sample size are needed to better substantiate our findings.

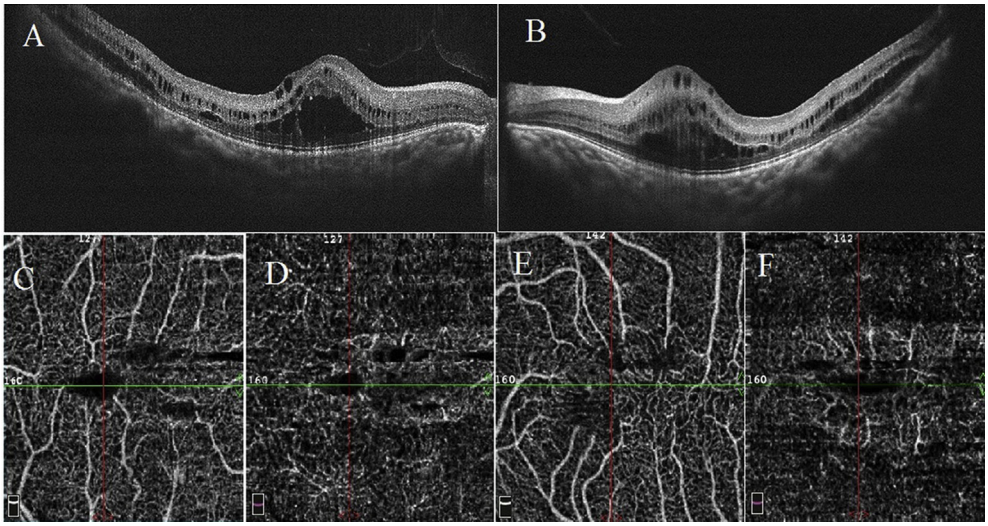

Fig. 2. Multimodal imaging of the second case. A and B show optical coherence tomography (OCT) of the patient showing retinal folds, schisis-like changes and all layers preservation in the macular. C and D show optical coherence tomography angiography (OCTA) of the superficial and deep layer of the right eye, and E and F show them for the left eye. It is obvious that the FAZ is absent and demonstrates vascularized superficial and deep capillary plexus.

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