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Fovea Plana in a 9-Year-Old Boy Presenting with Decreased Vision in the Left Eye

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 9
Final Diagnosis: Fovea plana
Symptoms: Visual acuity loss
Medication: —
Clinical Procedure: —
Specialty: Ophthalmology

Objective: Congenital defects/diseases
Background: Foveal hypoplasia (FH) is a congenital disorder, generally associated with other conditions.
Case Report: A 9-year-old boy presented with moderately decreased vision in the left eye. Fundus examination showed an absence of macular reflection and no foveal pit was seen on optical coherence tomography. Fluorescein angiography demonstrated the absence of a foveal avascular zone.
Conclusions: This is a rare case of a unilateral fovea plana associated with a visual impairment.

MeSH Keywords: Angiography • Capillaries • Fovea Centralis • Tomography, Optical Coherence

Full-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/912539>

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Background

Foveal hypoplasia (FH) occurs secondary to underdevelopment of the foveal pit. Foveal hypoplasia results in absence of foveal pigmentation, lack of the foveal avascular zone, and persistence of inner retinal layers in the foveola area. It is usually associated with other conditions such as aniridia, oculocutaneous albinism, microphthalmos, achromatopsia, congenital nystagmus, or retinopathy of prematurity [1]. When none of these conditions are found, FH is referred to as fovea plana (FP). Several studies reported cases of FP and FH. Patients have visual acuity between 20/20 and 20/200. A grading system allows classification of FH regarding morphologic findings obtained by optical coherence tomography (OCT) [2]. The majority of reported cases are bilateral. We describe the case of a young patient with unilateral FP and visual impairment.

Case Report

A 9-year-old boy reported moderately decreased vision in the left eye for a few years. He had no significant medical history and no ocular disease. There was no family history of genetic eye disease or systemic pathology. His best corrected visual acuity was 20/20 in the right eye and 20/32 in the left eye. Slit-lamp examination was normal. There was no iris transillumination defects suggestive of ocular albinism. He had no nystagmus. Pupils were round, symmetric, and reactive. Dilated fundus examination showed ill-defined maculofoveal area in the left eye. Optic nerves and retinal periphery were normal, with no uveitis signs or degeneration (Figure 1). Spectral-domain OCT of the left retina revealed absence of the foveal pit, and persistence of inner retinal layers in the foveolar area.

Foveal morphology in the right eye appeared normal (Figure 2). Fluorescein angiography showed a normal foveal avascular zone (FAZ) in the right eye. However, FAZ in the left eye was ill-defined and significantly smaller than normal (Figure 3). Optical coherence tomography angiography (OCT-A) showed persistence of superficial and deep capillary plexus in the left eye. In the right eye, retinal vascularization was normal, with the presence of a central dark area corresponding to the FAZ. There were no vessels abnormalities of the outer retinal and choriocapillary in either eye (Figure 4).

Discussion

Foveal development begins at fetal week 25. It follows a centripetal displacement of cone photoreceptors and centrifugal displacement of inner retinal layers to form a complete foveal pit at between 15 and 45 months after birth [3]. Foveal hypoplasia results from a developmental arrest during the steps of foveal formation. It can be isolated or associated with severe conditions such as albinism, aniridia, congenital nystagmus [4], microphthalmos, or others syndromes. Our patient did not show any symptoms of these diseases.

Several cases of fovea plana and foveal hypoplasia have been reported. One study reported that visual acuity can be between 20/200 and 20/20. Marmor et al. [1] showed that foveal depression is not a pre-requisite for foveal cone specialization. It is therefore possible to diagnose FP in a patient with good visual acuity.

Thomas et al. [2] developed a structural grading system for foveal hypoplasia based on the morphologic findings obtained by

Figure 1. Right and left eye retinographies. (A) Right eye: fundus normal. (B) Left eye: Ill-defined maculofoveal area and less delimited foveal reflex.

Figure 2. Optical coherence tomography of both eyes. **(A)** Right eye: foveal depression and absence of the inner retinal layers. **(B)** Left eye: absence of foveal pit with persistence of all the inner retinal layers and outer plexiform layer, outer nuclear layer widening, and outer segment lengthening in the foveolar area.

Figure 3. Fluorescein angiography of right and left eye. **(A)** Right eye: normal fovea avascular zone. **(B)** Left eye: ill-defined and significantly smaller fovea avascular zone.

Figure 4. Optical coherence tomography angiographies of the right and left eye. (**A, C**) Right eye: normal with the presence of a dark gap corresponding to the FAZ. (**B, D**) Left eye: lack of the FAZ and persistence of the superficial and the deep capillary plexi.

spectral-domain OCT. They also found a significant correlation between visual acuity and each grade. Our patient had no foveal pit, with persistence of all the inner retinal layers and outer plexiform layer, outer nuclear layer widening, and outer segment lengthening in the foveolar area, which makes it a grade 2 with a visual acuity of 20/32 according to Thomas et al. [2]. FH and FH are commonly bilateral [4–6]. In our case, the differential diagnosis includes an epiretinal membrane, which may simulate the absence of the foveal pit [7]. However, epiretinal membrane is rare in children with no retina-vitreous interface anomaly. Moreover, epiretinal membrane can be seen on OCT examination as a hyperreflective layer over the inner retina.

Finally, OCT angiography showed foveal capillaries across the fovea, which is consistent with the diagnosis of FP.

Only 3 cases of unilateral FH have been described and all were associated with varying degrees of vision loss [8–10].

Conclusions

We present a rare case of unilateral fovea plana. Angio optical coherence tomography is particularly important, as it provides definitive diagnosis by revealing unilateral affection.

Conflict of interests

None.

References:

1. Marmor MF, Choi SS, Zawadzki RJ, Werner JS: Visual insignificance of the foveal pit: Reassessment of foveal hypoplasia as fovea plana. *Arch Ophthalmol*, 2008; 126: 907–13
2. Thomas MG, Kumar A, Mohammad S et al: Structural grading of foveal hypoplasia using spectral-domain optical coherence tomography. A predictor of visual acuity? *Ophthalmology*, 2011; 118: 1653–60
3. Provis JM, Hendrickson AE: The foveal avascular region of developing human retina. *Arch Ophthalmol*, 2008; 126: 507–11
4. Giocanti-Aurégan A, Witmer MT, Radcliffe NM, D'Amico DJ: Isolated foveal hypoplasia without nystagmus. *Int Ophthalmol*, 2014; 34: 877–80
5. Karaca EE, Çubuk MÖ, Ekici F et al: Isolated foveal hypoplasia: Clinical presentation and imaging findings. *Optom Vis Sci*, 2014; 91: 61–65
6. Sanchez-Vicente JL, Contreras-Diaz M, Llerena-Manzorro L et al: Foveal hypoplasia: Diagnosis using optical coherence tomography angiography. *Retin Cases Brief Rep*, 2018; 12(2): 122–26
7. Govetto A, Lalane RA, Sarraf D et al: Insights into epiretinal membranes: Presence of ectopic inner foveal layers and a new optical coherence tomography staging scheme. *Am J Ophthalmol*, 2016; 177: 227–29
8. Asakawa K, Ishikawa H: Electroretinography and pupillography in unilateral foveal hypoplasia. *J Pediatr ophthalmol Strabismus*, 2016; 53: e26–28
9. Iqbal MM, Makar I: Unilateral foveal hypoplasia. *Can J Ophthalmol*, 2018; 53(3): e107–9
10. Villegas VM, Schwartz SG, Hamet TD et al: Variable clinical profile of fovea plana in normal children. *Ophthalmic Surg Lasers Imaging Retina*, 2018; 49(4): 251–57