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Bilateral Acquired Progressive Retinal Nerve Fiber Layer Myelination

Abstract

We present the multimodal imaging findings of an unusual case of bilateral acquired progressive myelination of the optic disc over a 10-year follow up period, in a hyperopic adolescent patient in the absence of an underlying ocular or systemic abnormality. Myelination of the left optic disc was noted at age 7 and of the right optic disc at age 13 but no other ocular or systemic abnormalities were identified. Cross sectional OCT and en face OCT angiography confirmed the presence of myelination of the retinal nerve fiber layer and excluded other etiologic possibilities including an astrocytic hamartoma.

Keywords: Optic Nerve; Optic Fiber Myelination, Acquired, Hyperopia

INTRODUCTION

Myelination of the retinal nerve fiber layer occurs in 1% of the population and is most frequently congenital and non-progressive. (1) Congenital cases are typically isolated but may be rarely associated with the syndrome of ipsilateral high myopia, strabismus and amblyopia (2,3), although cases have been reported in hyperopic eyes. Reports of acquired and progressive myelination associated with congenital optic nerve disorders such as Arnold-Chiari malformation, hydrocephalus, optic disc drusen, and iatrogenic causes such as optic nerve sheath fenestration, are relatively rare.(4-8) PLEASE ADJUST THE REFERENCE CITATIONS

While affected patients are typically asymptomatic, retinal nerve fiber layer myelination can be rarely associated with visual loss. Even more unusual are reports of acquired and progressive myelination of the nerve fiber layer. We present the multimodal imaging findings of such a case in a healthy young patient with a 10-year follow up.

CASE REPORT

A healthy 15-year-old male presented for routine ophthalmologic evaluation, his best corrected visual acuity (BCVA) was 20/20 in each eye with a manifest refraction of +5.00 +1.75 x 90 in his right eye and +4.75 +1.75 x 90 in his left eye. Dilated retinal examination demonstrated bilateral asymmetric myelination of the retinal nerve fiber layer starting from the optic disc, more prominent in the left eye. Prior color fundus photography was available dating back 8 years. Myelination was first noted in the superotemporal sector of the optic nerve of the left eye when the patient was 7 years old. Subsequent photography illustrated progressive myelination of the optic disc in the left eye and the development of acquired and progressive myelination of the superior disc border in the right eye when the patient was 13 years old. BCVA remained normal throughout the follow up. Family ocular and medical history was unremarkable. Automated perimetry displayed a full field

in both eyes (OU) without enlargement of the blind spot. Spectral domain optical coherence tomography (SD-OCT, (OCT Spectralis Heidelberg Engineering, CITY) illustrated a normal anatomy of the central macula and a thickened retinal nerve fiber layer (RNFL) corresponding to the myelination.

OCT angiography (RTVue XR Avanti; Optovue, Inc., CA, USA) with AngioVue software was performed using scan area protocols of 3x3 and 6x6 MM centered on the disc and the fovea and including segmentation through the nerve fiber layer. The superficial and deep retinal capillary plexus were within normal limits in the fovea OU and the radial peripapillary vascular plexus was also within normal limits with normal morphology and density even in the areas that co-localized with the myelinated nerve fiber layer excluding other etiologic possibilities such as an astrocytic hamartoma. The presence of optic nerve drusen was excluded by B scan ocular sonography.

DISCUSSION

Myelination of the retinal nerve fiber layer or the optic disc is frequently an incidental and isolated finding in patients without other ocular problems. Congenital nonprogressive forms are the rule and the development of acquired progressive myelination is very rare and the pathogenesis is unclear. Although optic disk drusen was ruled out, these unusual cases are typically associated with it as an evidence of constricted scleral canal. The formation of disk drusen has been attributed to a crowded disk anatomy causing axoplasmic stasis and subsequent axonal degeneration(9). Other optic nerve and systemic associations including abnormalities of the central nervous system and optic nerve procedures such as fenestration were also excluded.

Previous reports have described retinal vascular complications associated with myelination, including retinal capillary congestion, telangiectasia, neovascularization, and even recurrent vitreous hemorrhage(10,11). The high lipid content of myelin is primarily responsible for the blocking effect that is noted with fluorescein angiography. With the advent of OCT angiography, detailed analysis of the radial peripapillary plexus can be performed to ensure normal integrity and morphology of this vascular layer, and this non-invasive test can be repeated periodically to detect the development of any acquired vascular alterations. OCT angiography in our case failed to detect any vascular abnormalities associated with nerve fiber layer myelination.

The etiology of bilateral progressive myelination of the optic disc in our case is unclear, especially as local optic nerve and systemic associations were excluded. Anatomical considerations related to the optic canal and the optic ring causing axoplasmic stasis require more robust future study. We present however a very unique case of progressive bilateral myelination documented with detailed color fundus photography for 8 years and evaluated by OCT angiography.

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FIGURE LENGENDS

Figure 1. Sequential color fundus photography of the optic disc in each eye illustrating acquired progressive myelination of the superior disc margin. A. Note the normal color fundus photographs in 2007 with no evidence of optic disc myelination when the patient was 6 years old. B. Approximately 1 year later in 2008, myelination of the superior disc margin is noted OS (black arrowhead). Progressive myelination of the superior disc margin OS is noted in 2010 (C), 2011 (D), and 2013 (E). In 2014 myelination of the superior disc margin of the right eye (double black arrowhead) is noted (F). The progression continues bilaterally in 2015 (G) and 2017 (H).

Figure 2. Bilateral cross sectional spectral domain optical coherence tomography (OCT) images of the macula are within normal limits in each eye (A, B). Cross sectional OCT images through the nerve fiber layer around the disc illustrate a remarkably thickened nerve fiber layer corresponding to the myelination in each eye (C, D). En face structural OCT with segmentation at the level of the nerve head (upper and lower internal limiting membrane offset of 9 μ m and 150 μ m respectively) illustrate areas of hyper reflectivity co-localizing with the optic disc myelination at the superior disc margin in each eye (E, F). En face OCT angiography through the myelinated nerve fibers illustrates a normal radial peripapillary plexus in each eye without any abnormality in the density or morphology of this vascular layer (G, H).