Myelin Fiber Syndrome, Strabismus and Amblyopia: About A Case

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ABSTRACT

We report the case of a 2.5-year-old female patient referred for exotropia of the left eye evolving since birth. He had no particular history. Refraction under cycloplegics found strong anisometropia with strong myopia of the left eye: -1.50 (-1.00 to 100°) in the DO and -16.25 (-1.25 to 50°) in the OG. Corrected visual acuity was unquantifiable given the child's age. Anterior segment examination was normal as well as ocular tone. Fundus examination showed diffuse chorioretinal atrophy in the posterior pole of the left eye, peripapillary myelin fibers and a normal macula. The fundus examination of the right eye was normal.

The clinical picture and the imagery together lead us to suggest a myelin fiber - myopia - amblyopia syndrome. The patient was put on amblyopia treatment with regular follow-up, with no notable improvement to date.

Keywords: Myelin fiber syndrome, retina, strabismus.

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I. INTRODUCTION

Central Serous Chorioretinopathy (CSCR) is a rare retinal pathology that affects the young adult usually male which is characterized by the occurrence of a vanishing point resulting in an accumulation of sub-retinal fluid associated with a focal detachment of the retina. The pathophysiology has unfortunately not been clearly elucidated, several risk factors have been found (stress, disruption of the adrenergic system, anxiety, HTA, genetic predisposition, corticosteroids).

The CSCR is a rare complication of systemic corticosteroids, the dilated fundus examination finds a central serous retinal detachment (DSR). The diagnosis is confirmed by fluorescein angiography which reveals one or more fluorescent “pinpoints” indicating fluid leakage.

II. CASE REPORT

We report the case of a 2.5-year-old female patient referred for exotropia of the left eye evolving since birth. He had no particular history. Refraction under cycloplegics found strong anisometropia with strong myopia of the left eye: -1.50 (-1.00 to 100°) in the DO and -16.25 (-1.25 to 50°) in the OG. Corrected visual acuity was unquantifiable given the child's age. Anterior segment examination was normal as well as ocular tone. Fundus examination showed diffuse chorioretinal atrophy in the posterior pole of the left eye, peripapillary myelin fibers and a normal macula (Fig. 1). The fundus examination of the right eye was normal (Fig. 2). A cerebral MRI was requested and returned normal. The clinical picture and the imagery together lead us to suggest a myelin fiber - myopia - amblyopia syndrome. The patient was put on amblyopia treatment with regular follow-up, with no notable improvement to date.
III. DISCUSSION

Myelin fibers are present in 0.5 to 1% of the general population, and are bilateral in 7.7% of cases [1], [2]. They are sometimes inherited, in an autosomal dominant mode of transmission [3] without any particular mutation being incriminated to date. The pathophysiology is not well understood. Abnormal retinal myelination by ectopic oligodendrocytes and dysfunction of the cribiform lamina, developmental or acquired, are thought to be responsible for the development of myelin fibers [2]. The visual prognosis is poor in bilateral cases.

Patients with normal retina has nonvisible retinal nerve fibre layer, unmyelinated, which allows us to see retinal vessels. In abnormal patients with myelin fibers we can visualize the retinal nerve fibres. Recognized in the fundus exam as white or gray, opaque plaques on the retina, hiding the visualization of retinal blood vessels and the optic disc. Rarely some patients have different ocular anomalies associated, like, myopia, strabismus which can causes amblyopia if the diagnosis is pronounced lately.

The exact pathophysiology of myelin fiber syndrome is currently not known, glial cells are responsible for myelination in the central nervous system. Normal myelination of the optic nerve begins in the lateral geniculate ganglion and ends in the lamina cribrosa.

Indeed, the astrocytes concentrated in the lamina cribrosa a barrier to oligodendrocyte migration. Oligodendrocytes surrounding the syndrome have been observed in histological and electron microscopic studies in patients with myelin fiber syndrome [4], [5].

IV. CONCLUSION

Myelin fibers are benign and often asymptomatic lesions however they can be associated with other ocular lesions including myopia that may be the cause of amblyopia poor.

The screening is very important for refractive abnormality in unilateral myelin fibers, and fairly aggressive occlusive therapy.

CONFLICT OF INTEREST

The authors declare no competing interests. All the authors have read and agreed to the final manuscript.

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