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Case Report Open Access

Peripapillary Staphyloma in a Child with Intermittent Exotropia

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Introduction

Peripapillary staphyloma is an extremely rare non-hereditary congenital anomaly in which an excavation surrounds the relatively normal optic disk [1]. The aetiology of peripapillary staphyloma is unknown. It is thought to arise as a sequel to disturbance of scleral development at about 20 weeks gestation, perhaps arising as a consequence of the new development of intraocular pressure causing scleral herniation at this stage of development [2,3]. It is generally occurs in isolation [3,4]. Peripapillary staphyloma is generally unilateral, but a few cases of bilateral staphylomas have been reported [1,5-7]. Patients with peripapillary staphyloma are generally emmetropic or slightly myopic and their visual acuity usually is markedly reduced in the affected eye [5,8]. Occlusion therapy may rarely provides visual improvement [4]. Retinal detachment associated with peripapillary staphyloma has been published in a few reports [4,6,8,9]. We document the clinical features of peripapillary staphyloma in a 3-year-old girl with intermittent exotropia.

Case Report

A healthy 3-year-old girl was referred with a 3-month history of intermittent exotropia. The patient was born at 40 weeks of gestational age with a birth weight of 3200 g after a normal gestation period. Her development was normal. The girl was the first child of her parents and there was no history of significant ocular disease in her family. No ocular abnormalities were visible in either parent.

On examination, her corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. Cycloplegic refraction was +1.00 +0.50α30° OD and -3.00 -2.00α45° OS. The eyes were orthophoric (Figure 1A). Examination showed 30 prism diopters (PD) of intermittent exotropia at distance and near, and normal extraocular movements (Figure 1B). Both pupils were reactive with relative afferent pupillary defect in the left eye. She had no nystagmus, and the results of slit lamp biomicroscopy were normal. Dilated funduscopic examination of the left eye revealed a posterior peripapillary excavation with a nearly normal appearing optic nerve head at its base (Figure 2A).

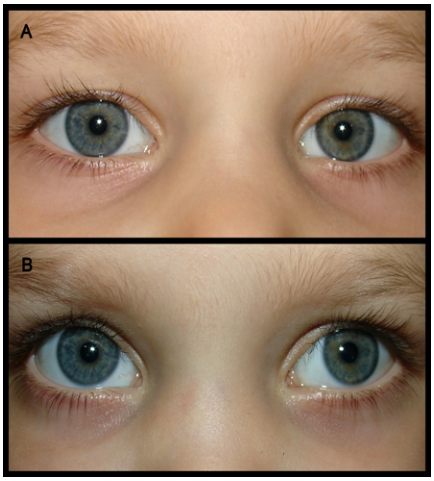

Figure 1: Orthophoria (A), exotropia in the left eye (B).

There was marked retinal pigment epithelium alteration surrounding the staphyloma. No glial tissue was present, and the retinal vasculature was normal. Ultrasonography of the left eye revealed an axial length of 23.45 mm with an excavation of the posterior wall of the globe (Figure 2B). Fundoscopic examination and ultrasonography of the right eye were normal with 21.63 mm axial length. Patch occlusion therapy with full refractive correction was given.

Discussion

The differential diagnosis of this patient's disorder includes the other excavated congenital optic disc anomalies. Morning glory disc anomaly is a congenital optic disc dysplasia in which the excavation may be shallower, cone-shaped and is filled with glial tissue. The optic disc may be anomalous and the vessels radiate more distinctly from the disc periphery [6]. Optic disc coloboma comprises a clearly demarcated bowl-shaped excavation of the optic disc, which is typically decentred and deeper inferiorly [3]. Iris or retina coloboma frequently accompanies optic disc coloboma [1]. Excavated optic disc anomalies are occasionally seen with other congenital defects or systemic diseases. But peripapillary staphyloma is known to be rarely associated with other congenital defects or systemic diseases [4,5].

Eyes with peripapillary staphyloma may rarely achieve visual improvement by occlusion therapy, but visual outcome is generally poor that reported in 2005 by Kim and colleagues in a series of patients with peripapillary staphyloma.

Although extremely rare, the peripapillary staphyloma may cause

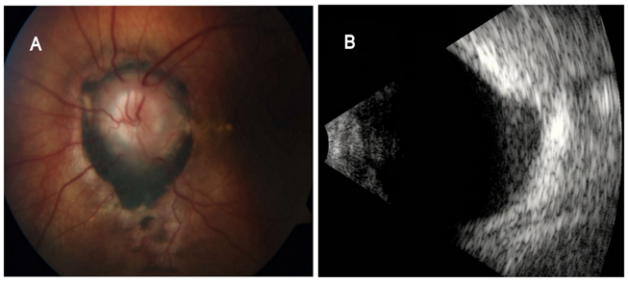

Figure 2: Fundus photograph (A) of the left eye shows a posterior peripapillary excavation with a nearly normal appearing optic nerve head surrounded by marked pigment epithelium alteration. Retinal vasculature is normal and no glial tissue is present. B-scan ultrasonography of the left eye (B) shows an excavation of the posterior wall of the globe.

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severe visual impairment and a complete ophthalmic examination and follow ups are necessary.

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